Antonello Trecca *Editor*

Atlas of Ileoscopy

A Collection of Clinical Cases

Springer

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Preface

Last year I was very proud to edit my first book dedicated to the diagnosis and treatment of ileal diseases, published under the simple title *Ileoscopy*.

The book's success derived from the participation of so many colleagues from several countries, who demonstrated their passion and competence in their contributions on this new topic of research in the field of modern gastroenterology and endoscopy.

Nonetheless, I never imagined that Springer would return with an even greater challenge: an atlas focused on this same topic, with brilliant images and concise but insightful discussions on the most important issues stemming from clinical practice!

But once again I was fortunate in being able to call upon my skilled colleagues, who, as before, did not disappoint in that they have been invaluable in sharing their knowledge. The result is a very interesting and user-friendly book that will not fail to attract the close attention of its readers, whether they are simply curious, wish to compare notes on a similar experience, consider a rare disease, view a particular image, update their skills, or, of course, share their passion for the world's most beautiful profession: medicine.

So please enjoy our book and let's hope that we meet again!

Rome, December 2012 Antonello Trecca

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The Asymptomatic Patient

Case 1
Ileoscopy in the Asymptomatic Patient

Antonello Trecca, Giuseppe Cerno, Pasquale Trecca,
Fabio Gaj and Gabriele Marinozzi

Background

In the field of modern gastroenterology, ileoscopy has opened up new frontiers in the diagnosis of intestinal diseases [1]. Exploration of the terminal ileum during total colonoscopy has gained much greater acceptance among endoscopists, based on its diagnostic accuracy in patients with intestinal diseases, including neoplasms [2]. Whether a routine terminal ileoscopy should be mandatory when performing a screening colonoscopy remains to be assessed. However, many authors seem to agree that, at the very least, an ileoscopy should always be attempted, because it allows an accurate diagnosis even in asymptomatic patients [3]. Here we present a clinical case in which ileoscopy enabled the definitive treatment of a rare tumor located in the terminal ileum.

Clinical Presentation

A 53-year-old woman underwent a screening colonoscopy as an outpatient procedure at our department. The terminal ileum was intubated while a total colonoscopy was performed. A sessile, (Paris classification 0-Is), submucosal lesion 5 mm in diameter was found (Fig. 1.1), located 5 cm from the Bauhin valve.

Virtual chromoendoscopy using the FICE (Fujinon Intelligent Chromoendoscopy) technique showed the round pitted pattern of the lesion and its regular vascular morphology (Fig. 1.2). Palpation with biopsy confirmed its elastic consistency and its mobility over the submucosal plane.

The lesion was completely excised with an endoscopic mucosal resection and the iatrogenic ulcer was closed with one resolution endoclip (Figs. 1.3, 1.4). The specimen was fixed and sent to the pathologist. Histology showed a well-differentiated neuroendocrine neoplasia, immunohistochemically positive for chromogranin A, with lateral and vertical free margins (Fig. 1.5), consistent with a diagnosis of grade 1 neuroendocrine tumor. The patient was discharged uneventfully.

After the initial diagnosis, a capsule endoscopy of the small bowel, a CT total body scan, and an octeotride scan were performed to stage the disease, all with negative findings. One year later, a second CT scan confirmed the absence of residual or recurrent disease. The patient is in good general conditions after 18 months of clinical follow-up.

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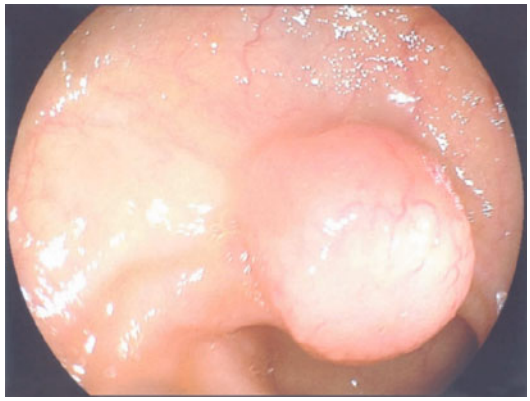

Fig. 1.1 Endoscopic view of a submucosal tumor of the terminal ileum

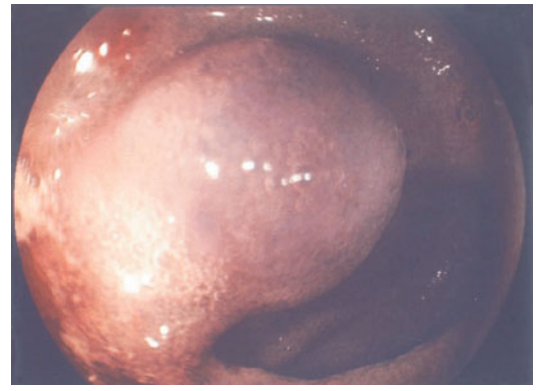

Fig. 1.2 Virtual chromoendoscpy (FICE) shows the normal vascular pattern of the tumor

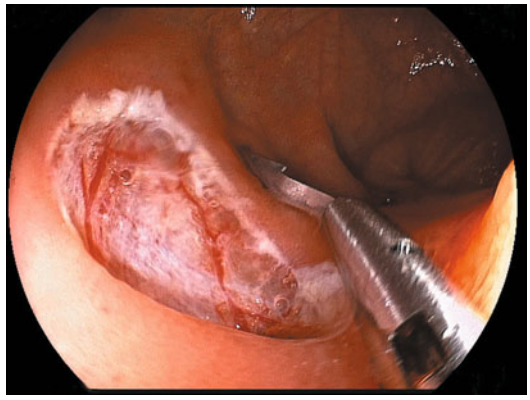

Fig. 1.3 Ulcer scare after endoscopic mucosal resection

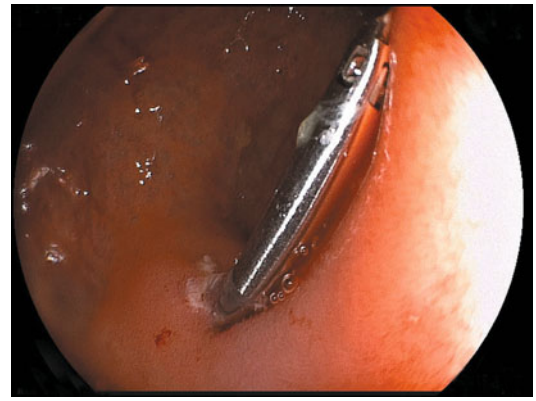

Fig. 1.4 Closure of the defect with a resolution clip

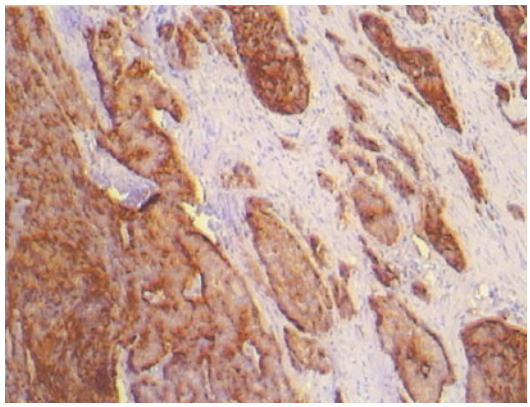

Fig. 1.5 Well differentiated carcinoid tumor, positive for chromogranin, ×200

Open Issues

Our experience demonstrates that a carcinoid of the terminal ileum can be detected even during a screening colonoscopy, underlining the importance of at least attempting an ileoscopy, even in asymptomatic patients.

Endoscopic treatment was carried out during the same session as the routine scheduled colonoscopy. The decision for immediate treatment was based on the small size of the lesion, such that a biopsy might have been insufficient for the definitive diagnosis, mainly due to the submucosal origin of the lesion

[4, 5]. Tissue sampling of the tumor can be performed with different techniques, such as the bite-on-bite (or stacked) biopsy, endoscopic-ultrasound-guided fine-needle aspiration (EUS-FNA) and EUS-guided tru-cut biopsy, but all of them have a low diagnostic accuracy. The elastic consistency of the lesion and its wide mobility over the submucosal plane convinced us to simultaneously treat the lesion, because only the resected specimen allows a definitive histological diagnosis.

In this patient, endoscopic treatment can be considered as definitive, confirmed at histological examination by the clear and free lateral and vertical margins and by the other imaging modalities, which excluded distant disease [6, 7]. Follow-up of the patient was negative and she was disease free at 18 months [8, 9].

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Multiple Choice Questionnaire

1) Which is/are the absolute indication(s) to ileoscopy?

a. chronic diarrhea
b. right lower quadrant abdominal pain
c. abnormal imaging finding
d. all of the above

2) Which of the following serological tests helps to diagnose a carcinoid syndrome?

a. raised transaminases
b. leukocytosis
c. high levels of chromogranin A and 5-hydroxyindoleacetic acid
d. raised lipase and amylase
e. raised γ-glutamyl transpeptidase and alkaline phosphatase

3) What is the 5-year survival percentage for patients with all forms of gastrointestinal carcinoids?

a. 20%
b. 40%
c. 50%
d. 70%
e. 90%

1.d – 2.c – 3.d

The Symptomatic Patient

Case 2
Isolated Polypoid Primary Lymphangiectasia of the Terminal Ileum

Federico Iacopini, Patrizia Rigato, Emma Calabrese and Agostino Scozzarro

Background

Intestinal lymphangiectasia is a rare condition of impaired lymphatic flow. Several forms are recognized: primary or congenital malformation (Waldmann's disease) [1], or secondary to a related localized obstructing pathology [2, 3]. The elevated pressure of the lymphatic drainage in the intestinal wall results in the leakage of lymphatic fluid and the manifestation of an exudative enteropathy with hypoproteinemia, peripheral edema, low serum gammaglobulin and/or lymphocytopenia. However, asymptomatic cases also have been described [4].

Clinical Presentation

A 67-year-old man underwent colonoscopy due to a positive fecal occult blood test. He was asymptomatic, with arterial hypertension as his only comorbidity. The family history was unremarkable. The physical examination was negative and the laboratory examination showed a normal complete blood count and ferritin.

Colonoscopy findings were normal as well, but on ileoscopy a hard-elastic 5-mm pseudo-pedunculated polyp (Paris classification type 0-Isp) was seen located 3 cm above the ileo-cecal valve, with a small erosion (Fig. 2.1). Chromoendoscopy with indigo-carmine (0.4%) evidenced a roundish and small, tubular pit pattern (type IIIs by Kudo) (Fig. 2.2) and no adjacent subtle lesions (Fig. 2.3). No further lesions were found in the ileum explored for 30 cm proximally.

The ileal polyp was resected during the same procedure by endoscopic mucosal resection after it was elevated by the injection of normal saline into the submucosa (Fig. 2.4). Microscopic examination demonstrated dilated mucosal and submucosal lymphatic vessels with polyclonal normal plasma cells, shortened and widened villi, and marked edema of the lamina propria, compatible with intestinal lymphangiectasia (Fig. 2.5). No acute or chronic inflammation was observed in biopsy specimens from the colon.

Subsequent esophagogastroduodenoscopy (EGD), abdominal MRI, and small intestine contrast ultrasonography (SICUS) (Fig. 2.6) were negative. Total protein, albumin, gamma globulin, creatinin, electrolytes, and C-reactive protein were normal. A stool sample was negative for steatorrhea and α1-antitrypsin was normal.

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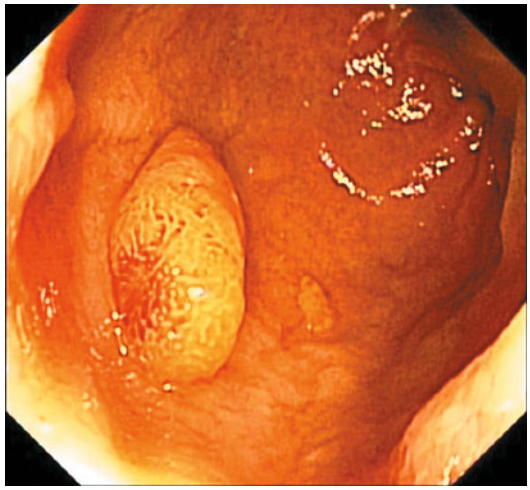

Fig. 2.1 Isolated polypoid primary lymphangiectasia of the terminal ileum

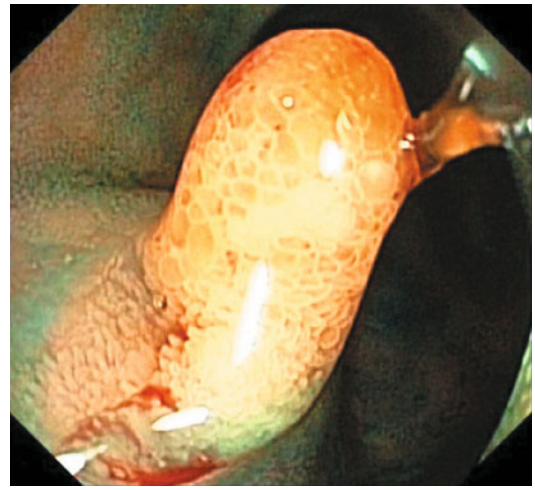

Fig. 2.2 Close-up view of the polypoid intestinal lymphangiectasia: roundish and small, tubular (type IIIs) pit pattern, and small mucosal erosion

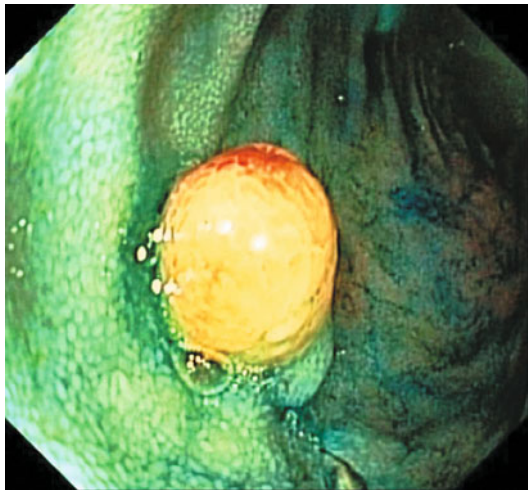

Fig. 2.3 Chromoendoscopy (indigo-carmine 0.4%) shows no subtle lesions adjacent to the polyp and normal villi

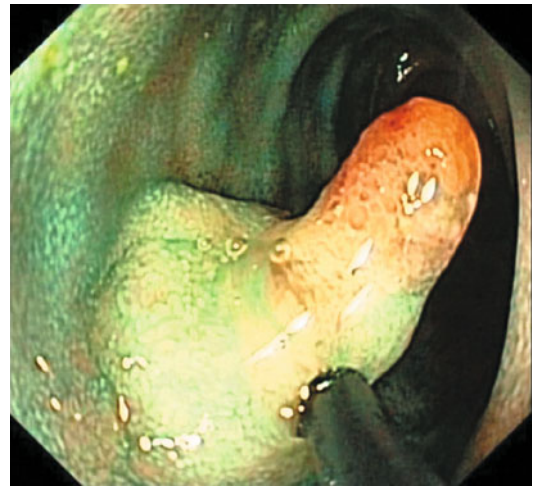

Fig. 2.4 Submucosal lifting of the polypoid intestinal lymphangiectasia before en bloc endoscopic mucosal resection

Isolated primary intestinal lymphangiectasia of the terminal ileum was diagnosed. During the following 36 months, the patient remained asymptomatic. The abdominal ultrasound was negative for lymphoadenopathy and/or masses.

Open Issues

The standard endoscopic features of intestinal lymphangiectasia are: (1) multiple scattered pinpoint white spots, (2) diffuse prominent villi plus whitish-discolored tips, and (3) focal small whitish macules or nodules [2, 5].

This is a rare case of an asymptomatic isolated polypoid primary intestinal lymphangiectasia of the terminal ileum. Thus far, it has been observed only in two previous cases, one primary [6] and the other secondary [7]. The pattern is considered as indicative of an advanced stage of the disorder, with an increased risk of lymphoma.

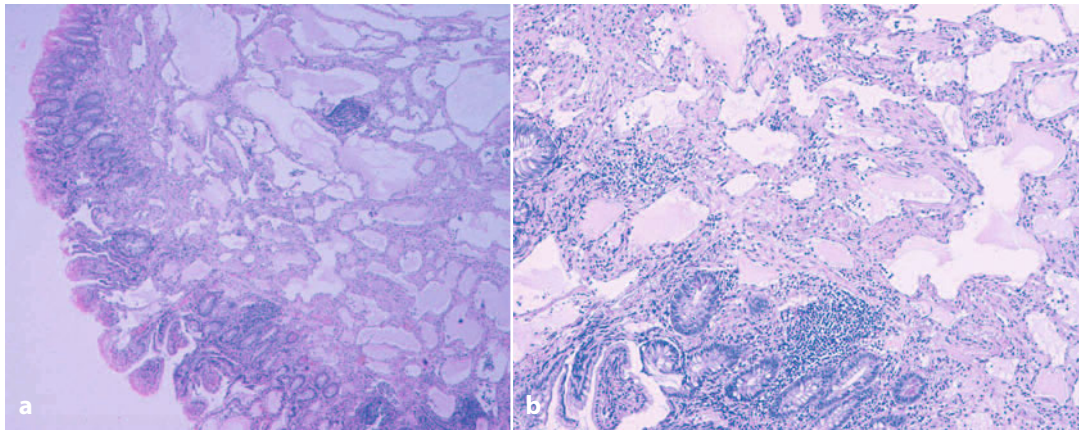

Fig. 2.5 Histological view of ileal lymphangiectasia: dilated mucosal and submucosal lymphatic vessels with cystic dilatations (lactocele); plump villi; surrounding lipid-rich macrophages (hematoxylin and eosin): **a** low-power view; **b** high-power view

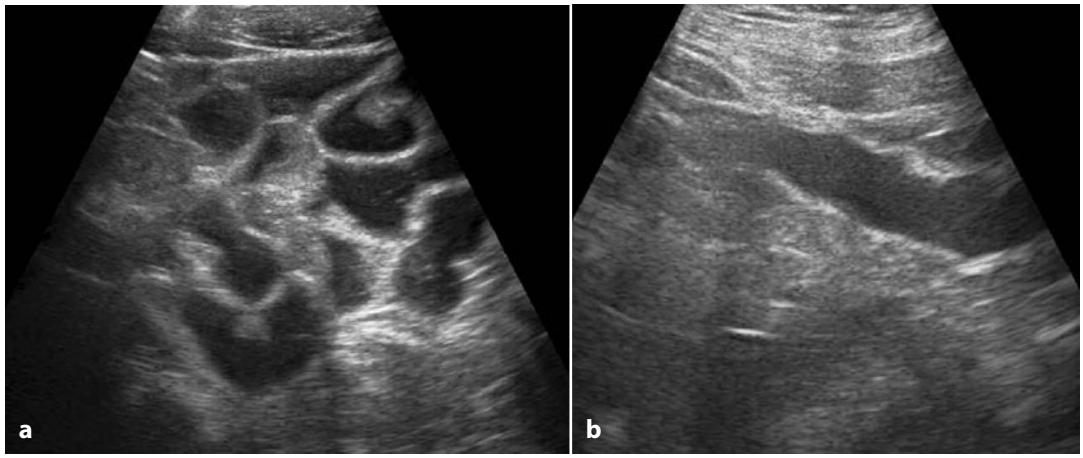

Fig. 2.6 Small intestine contrast ultrasonography (SICUS): **a** normal appearance of the ileal loops; **b** normal (2 mm) bowel wall at the terminal ileum

Features of intestinal lymphangiectasia are mainly visible in the duodenum. EGD, with the corresponding histology of intestinal biopsy specimens, is the main diagnostic approach. However, MRI, CT, or a dynamic examination by SICUS [8] and deep retrograde ileoscopy [9] are necessary to assess multifocality and to exclude secondary intestinal lymphangiectasia.

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Multiple Choice Questionnaire

1) Intestinal lymphangiectasia is
a. characterized by dilated lymphatic ducts in the bowel wall
b. associated with hypoproteinemia, edema, hypogammaglobulin, lymphocytopenia
c. primary or secondary
d. mainly diagnosed by endoscopy and histology
e. all of the above

2) Secondary forms of intestinal lymphangiectasia are
a. associated with systemic disorders
b. characterized by diffuse small bowel involvement
c. differentiated according to the endoscopic features
d. diagnosed by CT scan or MRI
e. related to advanced abdominal cancer

3) Endoscopic features of intestinal lymphangiectasia include
a. multiple scattered pinpoint white spots
b. diffuse prominent villi with whitish-discolored tips
c. focal small whitish macules or nodules
d. segmental or diffuse
e. all of the above

4) Intestinal lymphangiectasia is
a. characterized by exudative enteropathy but can be asymptomatic
b. treated with a low-fat diet with medium chain tryglicerides and vitamin supplementation
c. associated with a very low risk of lymphoma
d. all of the above
e. requires surgery for a definitive resolution

1.e – 2.d – 3.e – 4.d

Case 3
A Case of Unrecognized Complicated Celiac Disease

Riccardo Urgesi, Manuela Marzo, Cono Casale and Italo de Vitis

Background

Celiac disease (CD) is a genetic autoimmune disease in which affected individuals are unable to tolerate foods containing gluten, a specific protein of wheat, barley, and other cereals. The abnormal immune response in the intestine triggered by gluten generates a chronic inflammation and results in tissue damage to the small intestine, with the disappearance of the intestinal villi. If CD is not promptly diagnosed and treated, important and, in some cases, irreversible complications may occur [1]. In the most severe manifestations, CD can promote tumor development in the small bowel, such as intestinal lymphoma or adenocarcinoma. Especially in the initial phase of CD, the differential diagnosis will include lymphoma and other diseases [2]. Since the symptoms and endoscopic picture may be similar and non-specific, the diagnosis of CD is often challenging.

Clinical Presentation

In January 2007, a 53-year-old female without a previous medical history underwent a series of medical examinations to determine the cause of leg edema. The tests did not detect any evidence of cardiovascular disease but abdominal CT showed an "...important thickening of the third portion of the duodenum and the first jejunal loops with associated mesenteric lymphadenopathy (from 8 to 35 mm)...". She suffered weight loss (about 10 kg in 3 months), nausea, loss of appetite, and vomiting and was therefore admitted to the Department of Oncology. Blood tests including antibodies to CD and total immunoglobulins were normal. Ileocolonoscopy documented a diffusely edematous and fragile mucosa with large superficial ulcers covered with fibrin and involving only the terminal ileum. A diagnosis of Crohn's disease was then proposed. An entero-CT showed dilation of the proximal duodenum with obvious reduction of the caliber of the lumen at the level of the III duodenal segment. The involved duodenal walls were slightly thickened, irregular, and hyperdense until the duodenal-jejunal junction. The same pattern was evident at the first jejunal loops, where the wall thickness was about 7 mm, with mesenteric hyperdensity and multiple enlarged lymph nodes, (maximum diameter 3 cm) (Fig. 3.1). In January 2008, a push-and-pull enteroscopy confirmed these findings (Fig. 3.2). Histological examination was not definitive for Crohn's disease nor was it able to differentiate CD from autoimmune enteritis. Given the difficulty of

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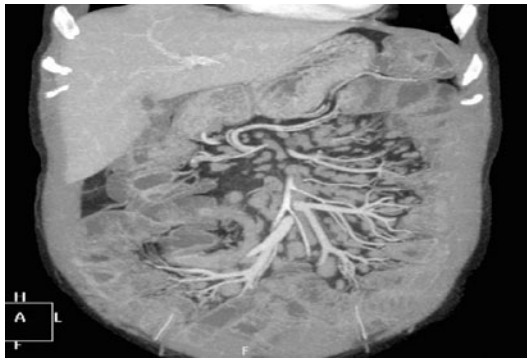

Fig. 3.1 Entero-CT showing dilation of the proximal duodenum with reduction of the caliber of the lumen at the level of the III duodenal segment

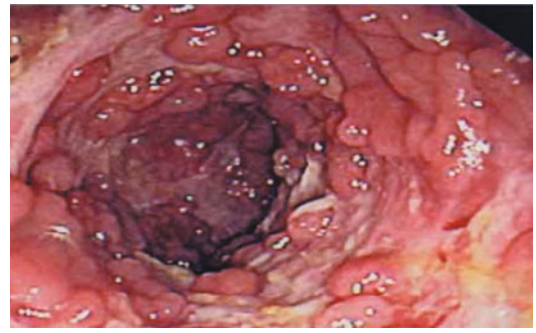

Fig. 3.2 Enteroscopic appearance of the first jejunal loop

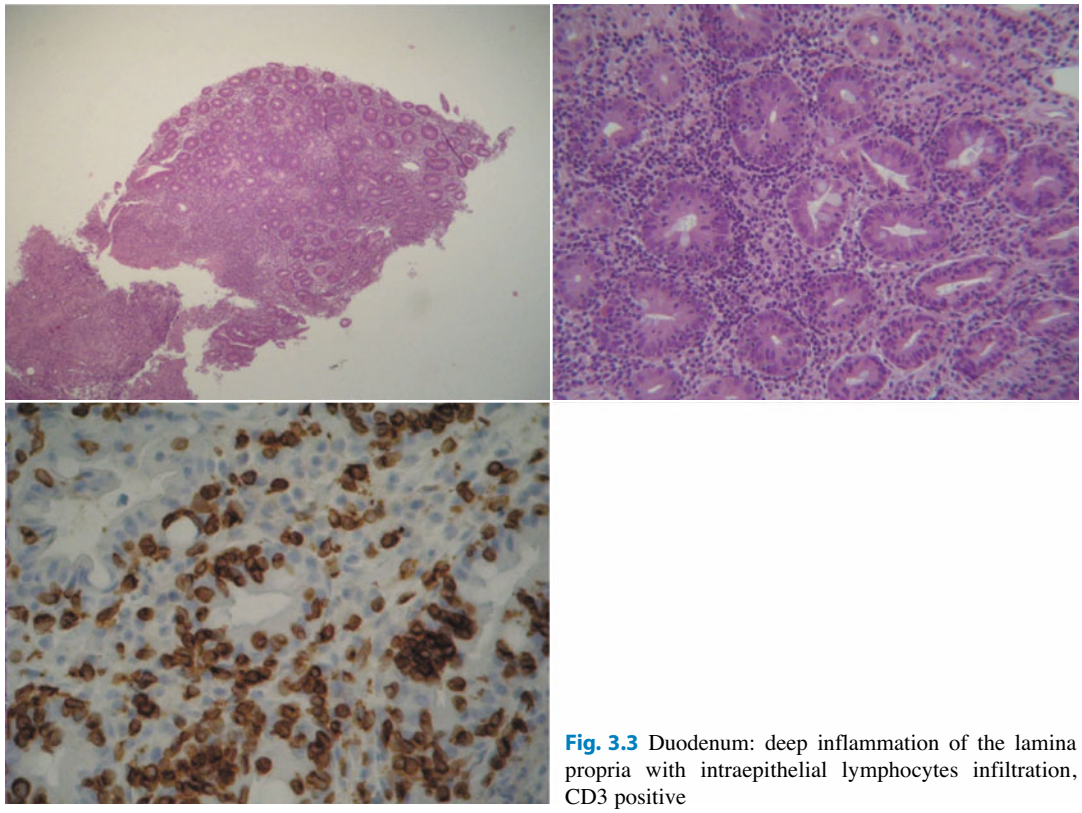

Fig. 3.3 Duodenum: deep inflammation of the lamina propria with intraepithelial lymphocytes infiltration, CD3 positive

this case the histological samples were sent to another reference center, which subsequently provided the following response: "Morphological aspect indicative of jejunal-ileitis with ulcerative negativity to CD8 and positivity for CD3 antigen, suggestive of a state of refractoriness strongly suspicious of a possible lymphomatous evolution" [3] (Figs. 3.3, 3.4). The patient also underwent DQ2/DQ8 haplotyping (DQ2 +), a bone marrow biopsy, lymphocyte subpopulation analysis, and an exploratory laparoscopy with lymph node biopsies, all not significant; and a CEUS (ultrasound with contrast medium) and diagnostic enteroclysis,

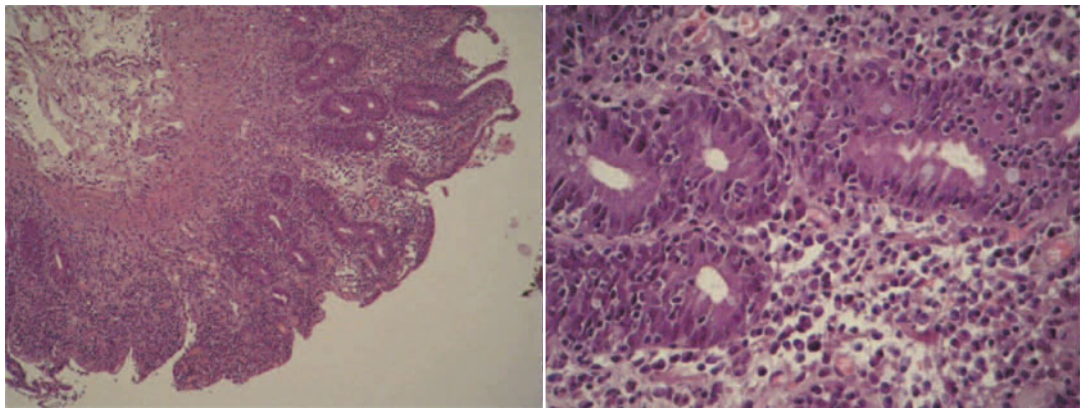

Fig. 3.4 Jejunum: histological view

both compatible with suspected ileal Crohn's disease with skip jejunal lesions; however "*…the appearance did not allow the exclusion of a possible jejunitis ulcerative.*" Based on the progressive deterioration of the woman's general condition and the inability to reach a final differential diagnosis between inflammatory bowel disease and suspected ulcerative jejunal ileitis in refractory celiac disease (or in lymphomatous transformation), the patient started steroid therapy and was placed on a gluten-free diet. After an initial clinical improvement followed by the discontinuation of steroid therapy while maintaining the strict gluten-free diet, there was a further significant worsening of symptoms. In June 2008, the patient was again hospitalized during which time her son was diagnosed with CD. She again underwent enteroscopy, with histology showing evidence of refractory CD with a lymphomatous evolution. Specific chemotherapy was then started, but her general condition worsened. The patient died due to gastrointestinal bleeding, impossible to treat.

Open Issues

The notable features of this case are that the clinical, radiological, and endoscopic features at the onset were all typical for Crohn's disease, particularly aphthoid ulcer, cobblestone pattern, and the deep mucosal lesions observed during push enteroscopy. Only the histological pattern was dubious for refractory CD. But the patient's non-responsiveness to steroid therapy and to a gluten-free diet should alert the physician to the likely evolution of the disease to intestinal lymphoma. This case shows that different diseases may have a common endoscopic picture and that T cell lymphoma can be synchronous with the onset of CD [4].

Sometimes the obvious diagnosis is not the correct one!

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Multiple Choice Questionnaire

1) Celiac disease is

a. an allergy
b. a temporary condition typical of childhood
c. a syndrome characterized by damage to the small intestinal mucosa caused by gliadin
d. an infectious disease
e. a malabsorption condition related to the pancreas

2) The most frequent complication of celiac disease is

a. enteropathy-associated T-cell lymphoma
b. refractory sprue
c. ulcerative jejunitis
d. all of the above
e. none of the above

3) The only current treatment for celiac disease is

a. immunosuppressant drugs
b. a vaccine
c. a lifelong, strict, gluten-free diet
d. a zonulin inhibitor
e. antibodies against IL-15

4) The enteropathy-associated T-cell lymphoma is

a. MALT lymphoma
b. burkitt-like lymphoma
c. T cell lymphoma
d. immunoproliferative small intestinal disease (IPSID)
e. gastrointestinal stromal tumor

1.c – 2.d – 3.c – 4.c

Case 4.1
Magnified Terminal Ileoscopy and Crohn's Disease: Added Value?

Antonello Trecca, Pasquale Trecca, Giuseppe Cerno, Fabio Gaj and Gabriele Marinozzi

Background

The endoscopic detection of Crohn's disease (CD) is generally difficult and confined to the late stage of the disease [1]. The importance of inspecting the terminal ileum during lower endoscopy is gaining increasing acceptance in the international literature based on its ability to provide an earlier diagnosis of this inflammatory bowel disease (IBD), especially in asymptomatic patients [2]. Over the last few decades, new endoscopic imaging modalities that offer enhanced early detection of neoplastic disease have been introduced into clinical practice, although their impact on the diagnosis of IBD is still under evaluation [3]. In the following, we present an unusual case of a patient with CD that was detected by terminal ileoscopy.

Clinical Presentation

In 2006, a 20-year-old man underwent lower endoscopy for the recent onset of abdominal pain and a small amount of rectal bleeding. Blood examinations and a fecal occult blood test showed unremarkable findings. Total colonoscopy was negative, but an attempted retrograde ileoscopy revealed multiple aphthoid lesions located in the last 5 cm of the ileum (Fig. 4.1). Histology performed on biopsy specimens showed severe inflammation of the terminal ileum, with a lymphoplasmacytic inflammatory infiltrate largely due to eosinophil granulocytes (Fig. 4.2). Capsule endoscopy of the small bowel confirmed the lesions in the terminal ileum and excluded further localization of the disease. C-reactive protein and mucoprotein were both negative. The patient was treated medically with 800 mg mesalazine b.i.d., which stabilized his condition at periodic follow-up, performed every 6 months. A second lower endoscopy performed 3 years after the initial diagnosis showed complete mucosal healing in the terminal ileum. Last year, the patient came to our attention for a reappraisal of his clinical symptoms, mainly abdominal pain. A third ileocolonscopy showed slight hyperemia of the villi, but a magnified view with the use of virtual chromoendoscopy showed enlarged lymphoid follicles with flat distorted domes surrounded by scattered hyperemic villi (Fig. 4.3). Histological examination of the biopsy specimen highlighted the presence of a non-caseous epithelioid granuloma, allowing a definitive diagnosis of CD (Fig. 4.4). After a negative radiologic examination of the gut the

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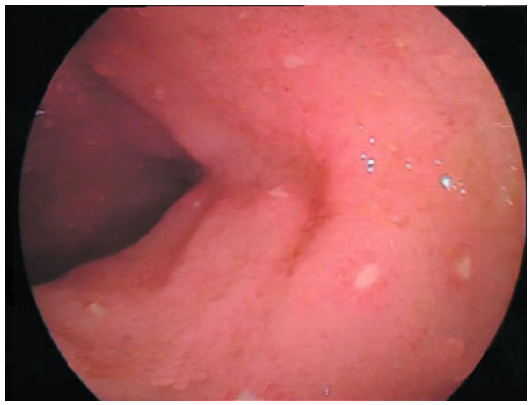

Fig. 4.1 Ileoscopy shows multiple aphthoid lesions located in the terminal ileum

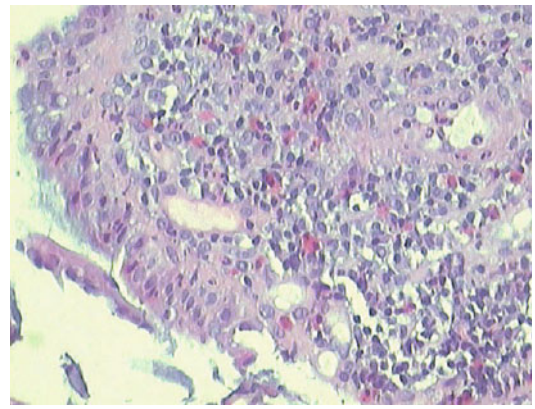

Fig. 4.2 Histological examination shows severe inflammation of the terminal ileum, characterized by a lymphoplasmacytic inflammatory infiltrate that is largely due to eosinophil granulocytes

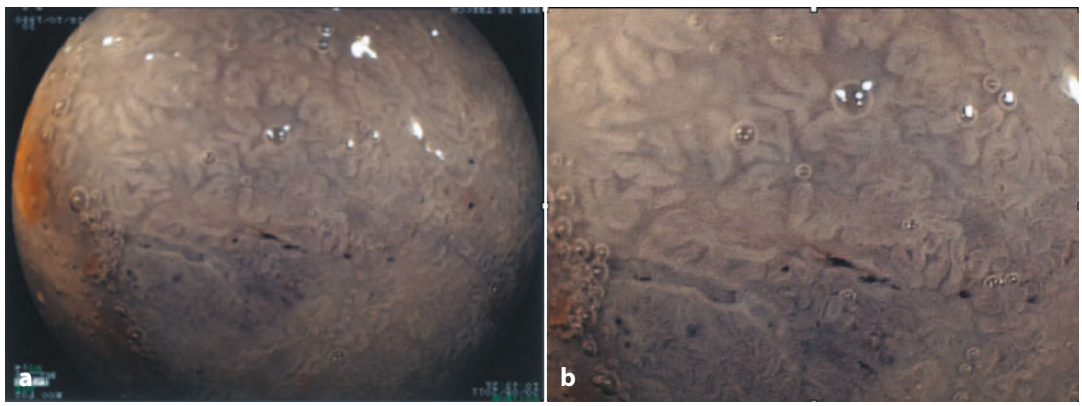

Fig. 4.3 **a** Conventional ileoscopy with slight hyperemia of the villi. **b** Magnified view on virtual chromoendoscopy shows enlarged lymphoid follicles with flat distorted domes

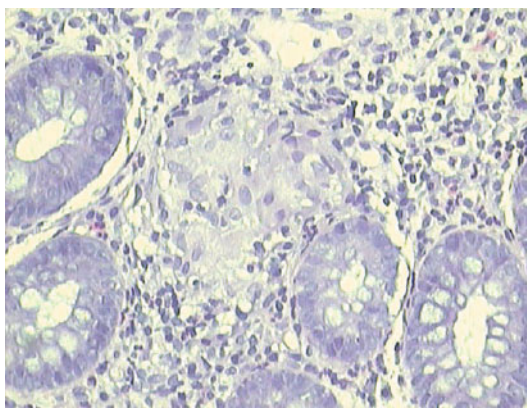

Fig. 4.4 Histologic examination on the biopsy specimen highlights the presence of a non-caseous epithelioid granuloma

patient continued medical treatment and was disease free at the last follow-up.

Open Issues

The absolute indications to perform terminal ileoscopy are well accepted and recognized. Thus, patients presenting with chronic diarrhea, right lower quadrant abdominal pain, abnormal imaging findings, suspicion of IBD, and a family history of CD must undergo complete exploration of the ileum [4]. In case of non-specific symptoms or in totally asymptomatic patients ileoscopy is still a matter of

debate, even if many authors agree on its importance also in this group of patients. This was the case in our patient, in whom early detection of the disease was possible by ileoscopy alone [5]. Another topic of discussion is the possible clinical impact of new imaging modalities applied to terminal ileoscopy. CD can appear with tiny aphthoid lesions located in the lymphoid follicles and originating from Peyer's patches along the gastrointestinal tract. In 1983, Fujikura [6] was the first to note that the follicle-associated epithelium in the terminal ileum can assume two main appearances: an elevated pattern (E type), corresponding to lymphoid follicles, and a flat pattern (F type), consisting of lymphocyte aggregations. In a series of seven patients with CD, magnified ileoscopy confirmed the presence of the F pattern in all of them, with flat distorted domes surrounded by few scattered villi. By contrast, in 11 of the 19 controls the E pattern was present. Shikuwa et al. [7, 8] postulated that the weak epithelium characteristic of the F type was likely to be more exposed to luminal antigens. The case of our patient with magnified-ileoscopy-confirmed disease underlines the pivotal importance of determining the pattern assumed by the Peyer's patches in patients with suspected IBD and to target these structures in the biopsy. As shown here, this approach resulted in the diagnosis of a non-caseous epithelioid granuloma, thus demonstrating the value of recognizing the F type in the diagnosis of CD.

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Multiple Choice Questionnaire

1) CD involves the terminal ileum in what percentage of cases?
a. 40–70%
b. 10–30%
c. 80–90%

2) How many CD patients will need surgery during the course of the disease?
a. up to 50%
b. up to 75%
c. up to 30%

3) Which of the following criteria describes the endoscopic severity of CD?
a. superficial ulcerations involving > 50% of the mucosal area
b. stenotic area or evidence of a fistulous tract
c. deep ulcerations eroding the muscle layer

1.a – 2.b – 3.c

Case 4.2
A Difficult Diagnosis of Crohn's Disease: Role of Ileoscopy with Biopsies

Roberto Lorenzetti, Angelo Zullo, Cesare Hassan, Francesca Stella and Vincenzo Bruzzese

Background

The diagnosis of inflammatory bowel disease (IBD) remains challenging even for the gastroenterologist who is specialized in its recognition and treatment. In IBD, each diagnostic modality has its specific role: laboratory data, radiologic imaging, capsule endoscopy, and endoscopy with histology. The correct diagnosis derives from an accurate consideration of the results of all the procedures undertaken and, of course, the clinical presentation of the patient. In the following, we present a clinical case in which all of these topics are well documented and discussed.

Clinical Presentation

A 26-year-old man presented with abdominal pain, reported daily, and nocturnal diarrhea (4-5 movements/day) and had a fever of 38–39°C. He was treated with ciprofloxacin 500 mg twice daily for 5 days, prescribed by his general practitioner, and his symptoms improved. However, 1 week following antibiotic discontinuation he was admitted to the Emergency Room for symptom recurrence. A CT scan showed a small fluid collection in the Douglas cavity and several enlarged mesenteric nodes. No increased intestinal wall thickness was detected. The patient was treated with a new course of antibiotics (metronidazole and gentamicin), which resulted in fever regression but persistence of both the diarrhea and the abdominal pain. Accordingly, he was admitted to our unit. At clinical history, he referred to a weight loss of about 11 kg in the last 3 months, and he currently smoked 20 cigarettes/day. Biochemistry showed mild anemia (hemoglobin 13.2 g/dL), iron deficiency (ferritin: 17 μg/dL), normal leukocyte count, but elevated C-reactive protein (5.31 mg/dL). Stool cultures were negative for *Campylobacter, Salmonella, Shighella,* and other common bacteria. An intestinal ultrasound and a small-bowel follow-through showed normal findings (Fig. 4.5). At ileocolonoscopy, normal colonic mucosa and mild erythema with focal granularity limited to 3 cm of the terminal ileum mucosa were seen (Fig. 4.6). Histopathological examination of several biopsies in the ileum revealed: (1) villous blunting, (2) thin superficial epithelium, (3) crypt distortion and enlarged spaces between crypts, (4) lymphoplasmacytic inflammatory infiltrate with eosinophil granulocytes, (5) lymphoid nodules, and (6) multiple epithelioid cells granulomas (Fig. 4.7). Together,

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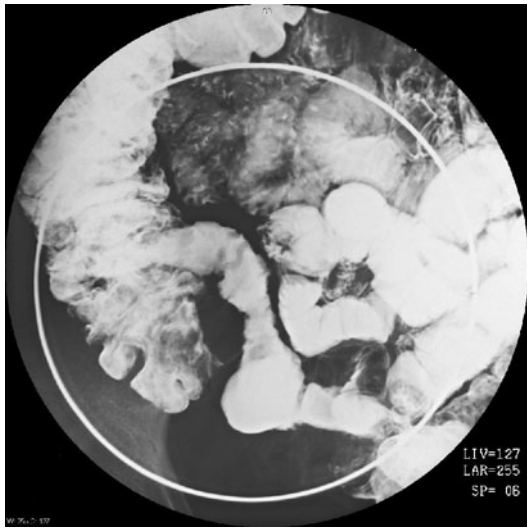

Fig. 4.5 Small-bowel follow-through showing normal distal ileum

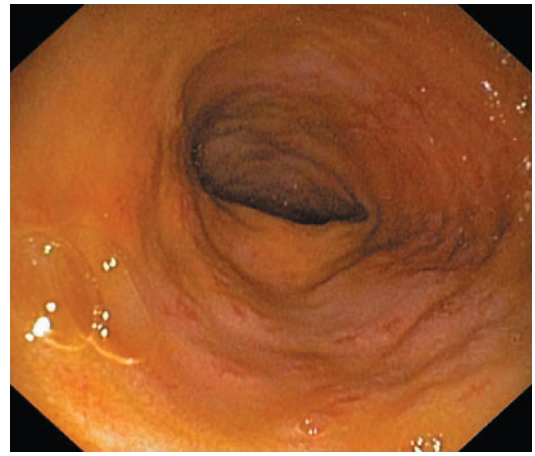

Fig. 4.6 Ileoscopy showing mild erythema and granularity in the distal ileum

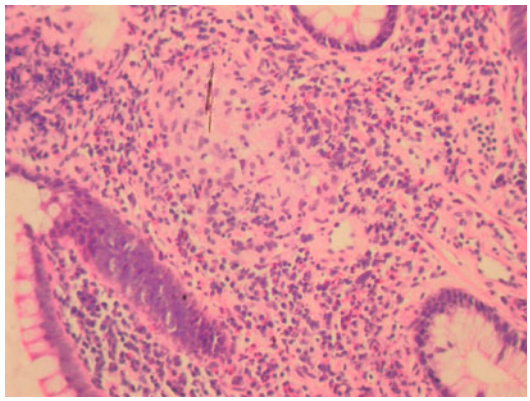

Fig. 4.7 Histopathologic examination showing epithelioid cells granuloma and other findings consistent with CD

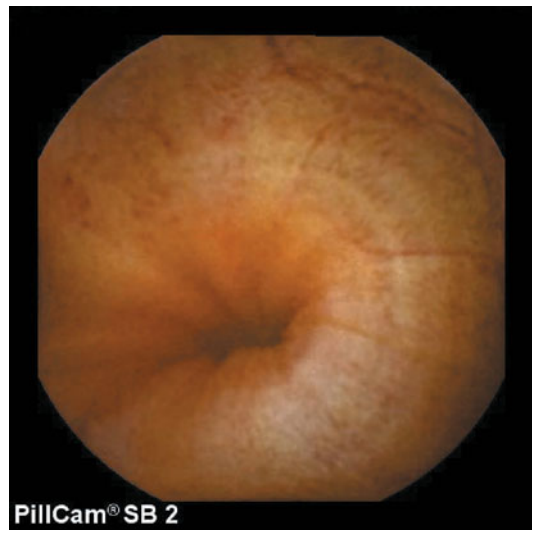

Fig. 4.8 Video capsule endoscopy with normal appearance of the ileal mucosa

these findings were consistent with a diagnosis of Crohn's disease (CD) ileitis. To rule out a jejunal or proximal ileal localization of the disease, a video capsule endoscopy was performed, but any pathologic findings were detected based on two consecutive interpretations by two different readers (Fig. 4.8). The patient was treated with 9 mg budesonide/daily for 6 months and then with mesalazine (3 g/daily), with prompt relief of symptoms.

Open Issues

This report describes a clinical case in which there was a discrepancy between a clinical picture highly suggestive of an IBD and the absence of relevant findings on all imaging procedures. Cross-sectional imaging techniques, i.e., ultrasonography and CT scan, were not diagnostically useful due to the absence of typical extraluminal manifesta-

tions of the disease, such as fistula or abscesses, or increased intestinal wall thickness. The endoscopic examination of the terminal ileum showed only mild and non-specific alterations limited to 3 cm of the mucosa. Moreover, video capsule endoscopy, despite its sensitivity in the diagnosis of CD of > 90% [1], missed the mucosal changes previously observed at ileocolonoscopy. CD was eventually confirmed only by histological examination of ileal specimens. This lack of significant findings at endoscopy could be explained by a spontaneous or drug-induced reduction of the inflammatory process between the onset of symptoms and the time of the procedures. The role of ileoscopy with biopsies in patients with chronic non-bloody diarrhea especially in those with clinically suspected IBD has been highlighted [2, 3]. Specifically, CD can be diagnosed based on the presence of mild endoscopic alterations together with chronic inflammation at histology when the clinical features are highly suggestive of IBD [4]. Finally, in accordance with ECCO guidelines [5], ileocolonoscopy and biopsies in the terminal ileum should be the first-line procedures, to be performed as soon as possible after the onset of symptoms.

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Multiple Choice Questionnaire

1) What is the first-line diagnostic procedure for suspected IBD?
a. ultrasonography
b. MRI enterography
c. CT enterography
d. ileocolonoscopy
e. ileocolonoscopy and ultrasonography

2) The sensitivity of video capsule endoscopy for the diagnosis of Crohn's disease is
a. 90-100%
b. 80-90%
c. 70-80%
d. 60-70%
e. 50-60%

3) Which is the histological finding highly suggestive of Crohn's disease?
a. criptic abscess
b. epithelioid granuloma
c. both of them
d. none of them
e. eosinophilic infiltrate in the submucosa

1.d – 2.a – 3.b

Neoplastic Disease

Case 5.1
A Small Non-Polypoid Advanced Colon Cancer 8mm in Size

Kuangi Fu, Hiroya Ueyama and Taiji Saga

Background

Small colorectal cancer, 10mm or smaller in size, is rarely encountered during colonoscopy. Almost all of the cases have been reported from Japan [1, 2]. The incidence has been estimated to be 0.05% of 5120 colonoscopies. Although the tumor is small in size, nodal involvement can be seen histologically, which suggests its high malignant potential compared to polypoid cancers. Considering its small size, non-polypoid shape, no coexistence of adenomatous component, a negative k-ras point mutation, and frequent positive p53 immunostaining in reported cases, this kind of small advanced colorectal cancer is likely to have progressed from the depressed type of early cancer rather than a polypoid adenomatous polyp.

Clinical Presentation

A 45-year-old man underwent total colonoscopy because of a positive fecal occult blood test (FOBT). His medical history included paroxysmal atrial fibrillation and he received anticoagulation therapy with warfarin. There was no family history of colorectal polyp or cancer. No remarkable findings were seen in both of his physical examinations and laboratory data except for a positive FOBT. The colon was very spastic during probe insertion and withdrawal, but we could only administer glucagon instead of buscopan because of his heart disease. We also sprayed peppermint oil to reduce the spasm, but it was ineffective [3]. In the ascending colon, colonoscopy almost missed a small depressed area, about 8mm in size, with a polypoid nodule in the center (Fig. 5.1). The lesion was reddish in color and was covered by dense mucus. Moreover, there was submucosal-mass-like lesion around the depressed area, which indicated that the lesion deeply extended into the submucosal layer. Chromoendoscopy using 0.4% indigo-carmine highlighted the well-demarcated depressed area (Fig. 5.2). Macroscopically, the lesion was classified as IIa+IIc in shape according to the Paris classification. Narrow-banding imaging (NBI) with magnification revealed prominent small capillaries on the surface, which was diagnosed as type IIIA capillary pattern (CP) according to Sano's classification (Figs. 5.3, 5.4) [4]. However, tiny avascular areas were also identified in the depressed area, suggestive of a type IIIB CP (Fig. 5.5). Magnifying chro-

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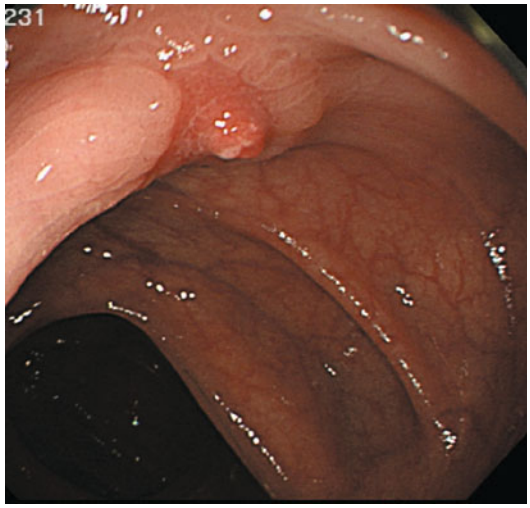

Fig. 5.1 In the ascending colon, colonoscopy almost missed a small depressed area, about 8mm in size, with a polypoid nodule in the center

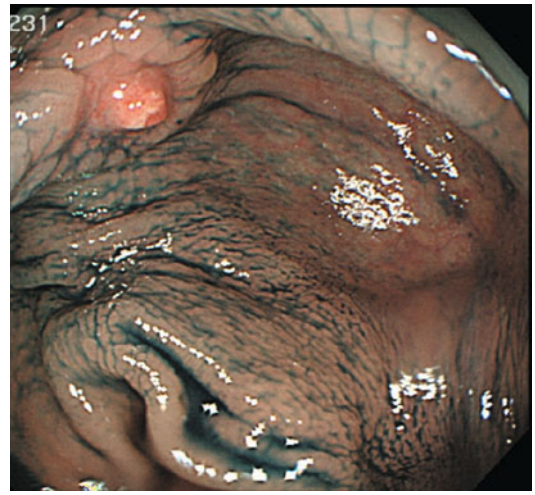

Fig. 5.2 Chromoendoscopy using 0.4% indigo-carmine highlighted the well-demarcated depressed area

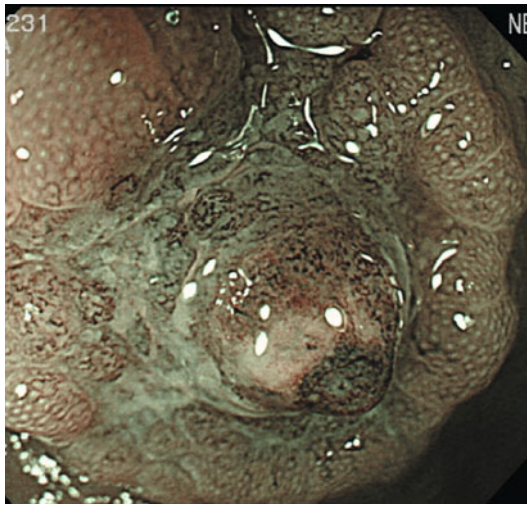

Fig. 5.3 Macroscopically, the lesion was classified as IIa+IIc in shape according to the Paris classification

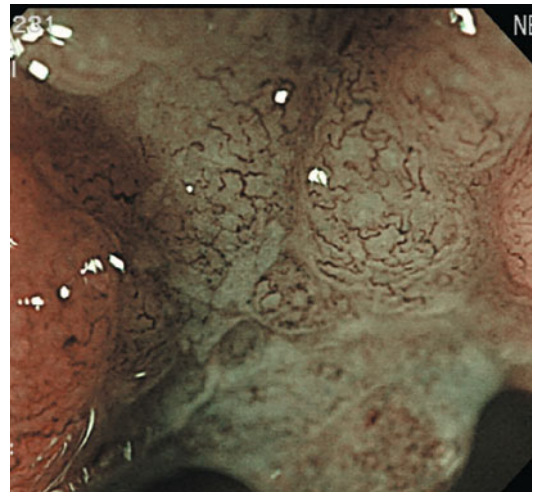

Fig. 5.4 Narrow-banding imaging (NBI) with magnification revealed prominent small capillaries on the surface, which was diagnosed as a type IIIA capillary pattern (CP) according to Sano's classification

moendoscopy (0.4% indigo-carmine and 0.05% crystal violet staining) could not clearly show the pit pattern for diagnosis, as dense mucus could not be removed despite repeated washing with water containing a mucolytic agent (pronase) (Fig. 5.6) [5]. The lesion was diagnosed as an invasive cancer deeply extending into the submucosal layer. An endoscopic biopsy was taken after warfarin was replaced with intravenous administration of heparin, disclosing an adenocarcinoma histologically (Fig. 5.7). We finally recommended surgical resection for treatment, as the above-mentioned endoscopic findings strongly suggested a submucosal deeply invasive cancer. Systemic imaging including computed tomog-

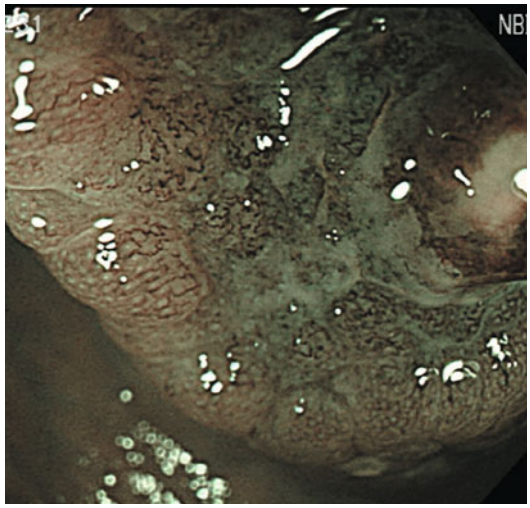

Fig. 5.5 Tiny avascular areas were also identified in the depressed area, which seemed to be a type IIIB CP

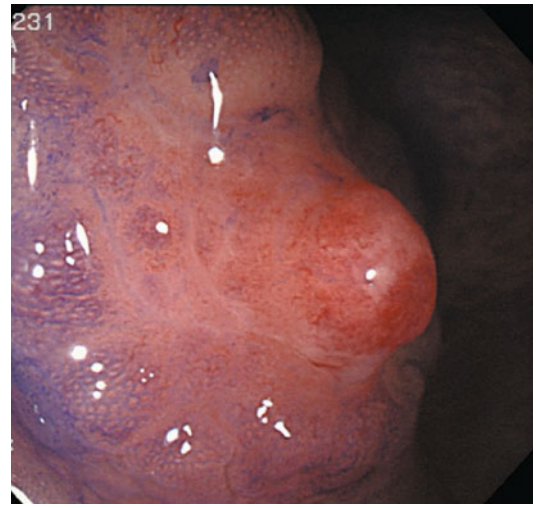

Fig. 5.6 Magnifying chromoendoscopy (0.4% indigocarmine and 0.05% crystal violet staining) could not clearly show the pit pattern for diagnosis, as dense mucus could not be removed despite repeated washing with water containing a mucolytic agent (pronase)

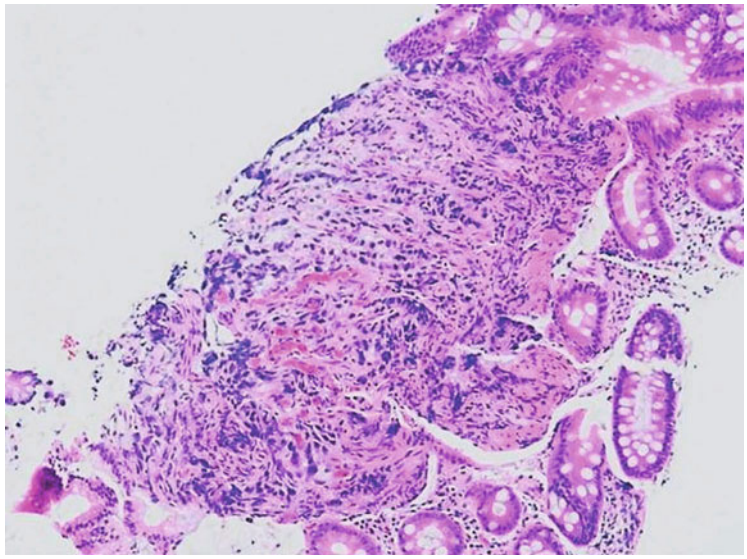

Fig. 5.7 The lesion was diagnosed as an invasive cancer deeply extending into the submucosal layer. An endoscopic biopsy was taken after replacing warfarin with the intravenous administration of heparin, disclosing an adenocarcinoma histologically

raphy showed no lymph node swelling or distant metastasis. A laparoscopy-assisted right-sided hemicolectomy was performed for treatment. Histological examination of the resected specimen revealed a moderately differentiated adenocarcinoma in the mucosal layer, and a poorly differentiated adenocarcinoma extended through the submucosal layer into the proper muscle layer (Fig. 5.8). Marked lymphatic invasion but not venous invasion of cancer cells was seen mainly in the deeper part of the mucosal layer (Fig. 5.9). Unfortunately, the data of k-ras point mutation were unavailable; however, p53 was diffusely positive on immunostaining. Both MLH1 and MSH2 were negative immunohistochemically.

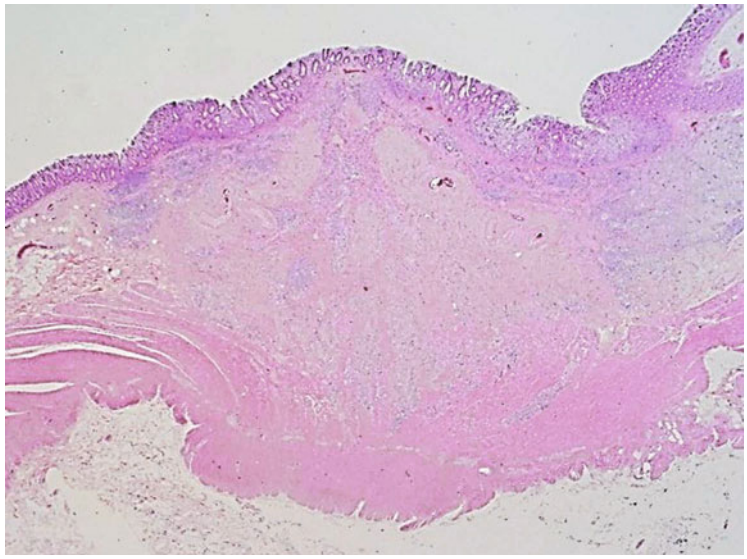

Fig. 5.8 Histological examination of the resected specimen revealed a moderately differentiated adenocarcinoma in the mucosal layer, and a poorly differentiated adenocarcinoma extending through the submucosal layer into the proper muscle layer

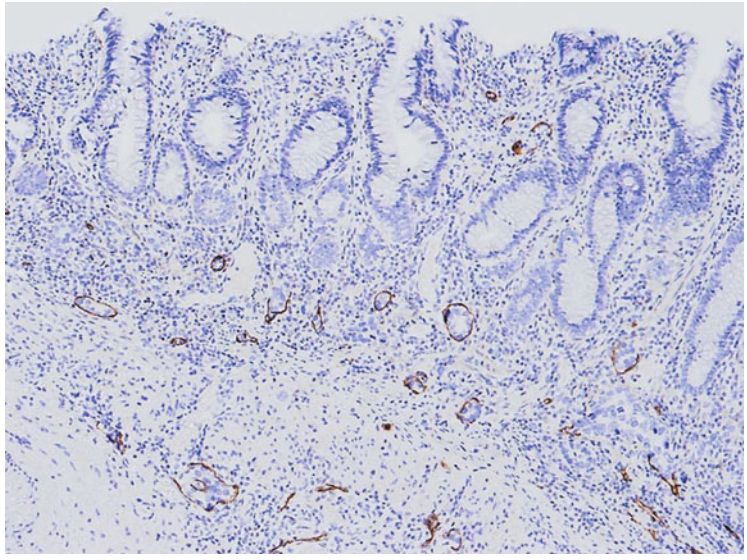

Fig. 5.9 Marked lymphatic invasion but not venous invasion of cancer cells is seen mainly in the deeper part of the mucosal layer

Nodal involvement was detected in one out of 19 dissected lymph nodes (Fig. 5.10). This case was finally classified as stage IIIA (T2, N1, M0) according to the TNM classification. The patient received adjuvant chemotherapy thereafter, and no recurrence was detected at the last follow-up of 2 years after surgery.

Open Issues

It is extremely difficult to detect such a small invasive cancer in a spastic colon. Magnifying chromoendoscopy was ineffective for pit pattern analysis due to the covered dense mucus. Correct endoscopic estimation of cancer depth

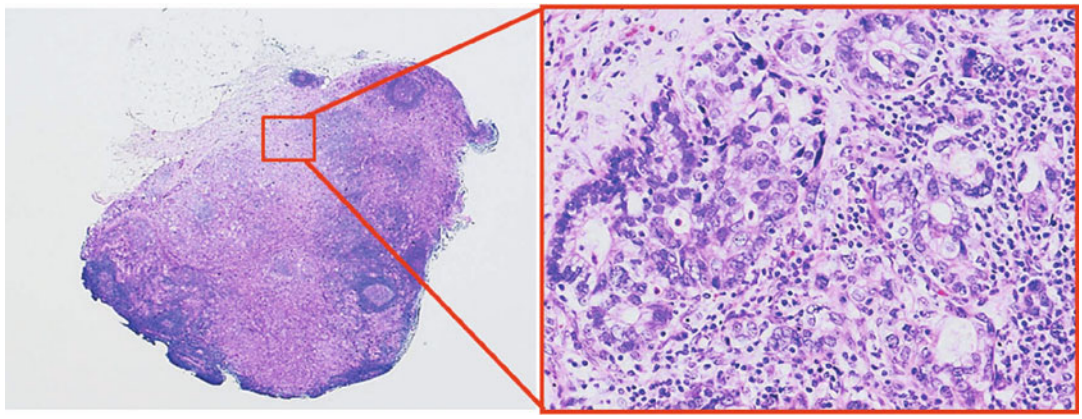

Fig. 5.10 Nodal involvement was detected in one out of 19 dissected lymph nodes

invasion could be achieved after considering the submucosal-tumor-like elevation around the depressed area on conventional view in addition to the capillary patterns observed on magnifying NBI. Moreover, although it looked like a non-polypoid early cancer endoscopically, surprisingly, the tumor had already invaded the muscle layer. Remarkably, there were unfavorable histological findings including poorly differentiated adenocarcinoma, marked lymphatic invasion, and nodal involvement. Although there was no family history of colon cancer and immunostaining of MLH1 and MSH2 was negative, considering a right-sided poorly differentiated adenocarcinoma, this patient may be the initiator of hereditary nonpolyposis colorectal cancer and should be followed up carefully.

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Multiple Choice Questionnaire

1) Which is the incidence of small colorectal cancers less than 10 mm in size?

a. 1%
b. 2%
c. 0.05%
d. 3%

2) What is the incidence of perforation after colorectal EMR?

a. 3%
b. 0%
c. 5%
d. 2%

3) Which is the incidence of perforation after colorectal ESD?

a. 8%
b. 15%
c. 10%
d. 5%

1.c – 2.a – 3.a

Case 5.2
The Hidden Cecal Region: Highlighted in a Clinical Case

Takahiro Fujii

Background

Accurate exploration of the cecal region represents a fundamental step in achieving a proficient colonoscopy and in improving the adenoma detection rate [1].

The morphology of the cecal valve or a redundant tenia coli may sometimes obscure or mimic the cecal region. Thus, the endoscopist should carefully inspect this region by locating the endoscope below the valve's lower lip in order to reduce the rate of missed early colorectal cancers [2]. In addition, intubation of the terminal ileum has been suggested by many authors as definitive proof of a total colonoscopy [3].

An improved endoscopic view is now available with new techniques such as virtual chromoendoscopy. A magnifying view provides added value in the early detection of colorectal cancer; however, the endoscopist must keep in mind the "red flags" regarding the mucosal appearance of these tumors [4].

Laterally spreading tumors account for 5% of all polypoid and non-polypoid lesions, with a higher prevalence in the cecum and rectum; thus, a careful inspection of the cecal region should be mandatory [5].

Clinical Presentation

A 75-year-old female came to our attention for endoscopic treatment of a sessile rectal polyp of 10 mm detected in another hospital. An outpatient total colonoscopy (PCF-Q260AI, Olympus, Tokyo, Japan) performed in our institution showed a faint reddish change and a slight unevenness of the cecal mucosa, just below the lower lip of the cecal valve.

Chromoendoscopy with 0.4% indigo carmine dye disclosed a laterally spreading tumor, granular type, with a small sessile component 20 mm in diameter. Narrow-band imaging (NBI) showed the meshed capillary vessels as dark-brown areas suggesting a non-invasive neoplastic lesion (Fig. 5.11). After the injection of a hyaluronic acid formulation (Mucoup, Seikagaku, Tokyo, Japan) into the submucosal layer, an en bloc resection was performed with a Captivator Micro-Hex snare (Boston Scientific, MA, USA). The cecal defect was closed with three endoclips (Olympus, Tokyo, Japan). The clinical course of the patient was uneventful, without bleeding or perforation (Fig. 5.12).

Histologically, the removed specimen was a well-differentiated adenocarcinoma with submucosal invasion of the lymph follicle (Fig. 5.13).

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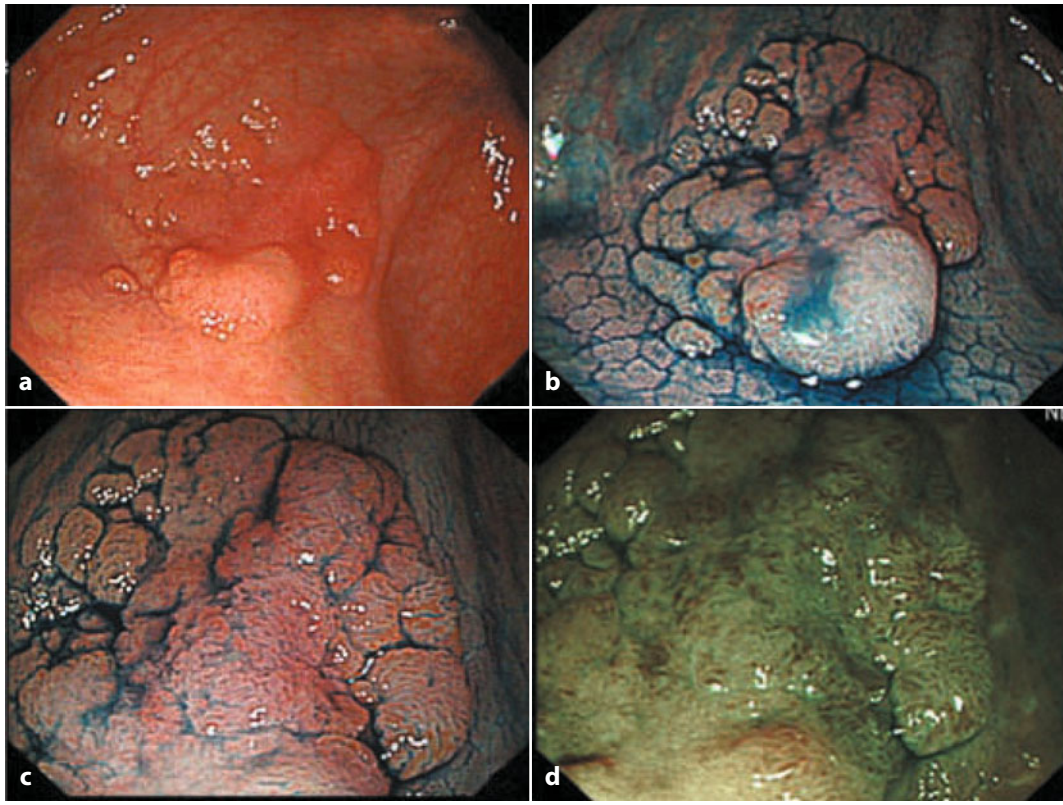

Fig. 5.11 **a** The lesion was detected as faint reddish change and mucosal unevenness, just below the lower lip of the cecal valve. **b** Chromoendoscopy disclosed a laterally spreading tumor, granular type, 20mm in diameter. **c** The image shows the center of the tumor suggesting carcinoma in endoscopic findings. **d** Narrow band imaging showed the meshed capillary vessels suggesting a non-invasive neoplastic lesion

Open Issues

A well-performed total colonoscopy should include accurate inspection of the cecal region in order to reduce the rate of missed colorectal lesions during the endoscopic procedure. Correct positioning of the colonoscope in the ileocecal region is an essential step in obtaining a thorough inspection. Straightening the colonoscope in the left colon and in the transverse colon improves its maneuverability.

The use of an anticholinergic agent or CO2, or a transparent hood attached to the tip of the colonoscope provides better visualization of the cecal region and facilitates intubation of the ileal valve [6]. This endoscopic

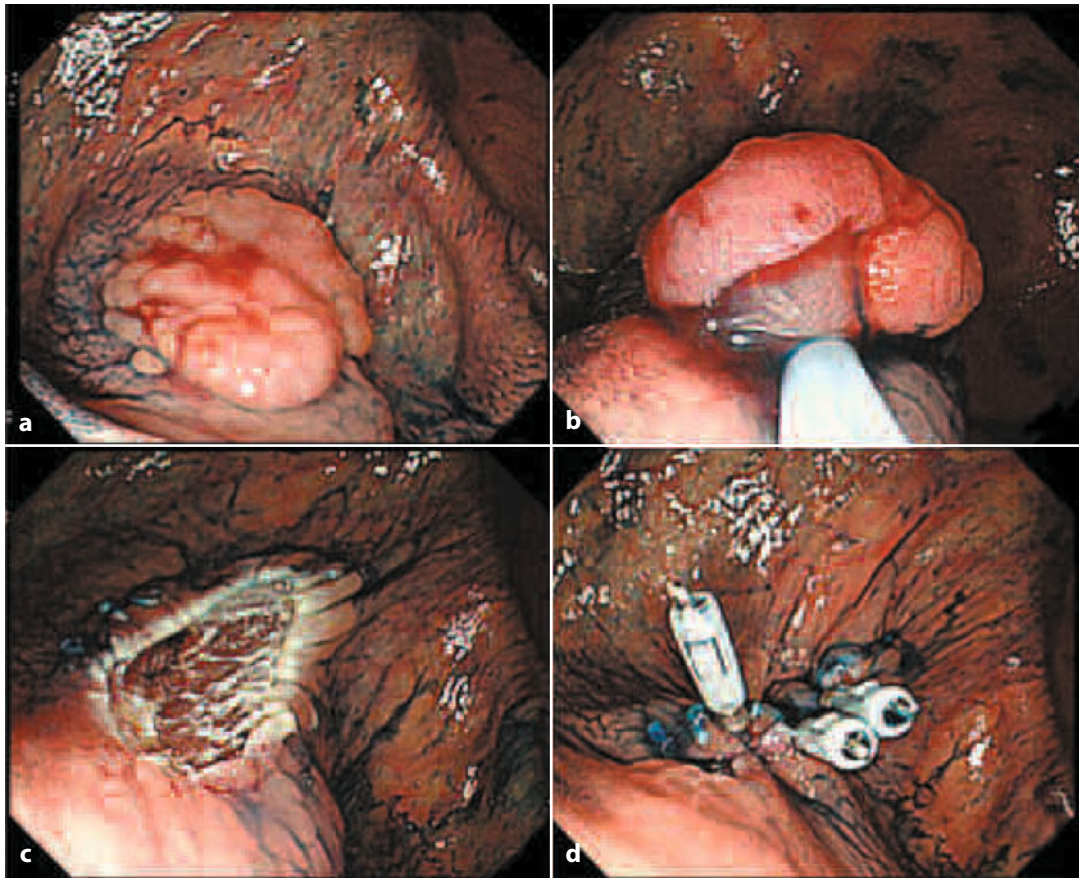

Fig. 5.12 **a** The injection of a hyaluronic acid formulation, reveals negative "no-lifting sign". **b** An en-bloc resection was performed with a snare. **c** Mucosal defect was seen. **d** The cecal defect was closed with three endoclips

"armamentarium" must be part of the endoscopist's know-how, as confirmed in our patient in whom the previous colonoscopy failed to detect a laterally spreading tumor in the cecum.

En-bloc endoscopic mucosal resection is the ideal treatment of these lesions, even if their large diameter can necessitate a piecemeal resection [7]. However, this endoscopic procedure cannot guarantee an accurate histological examination, accounting for the higher incidence of recurrence. Nonetheless, high-definition endoscopy based on a skilled technique can assure a safe and definitive oncologic treatment, as confirmed in this case by the histological examination [8].

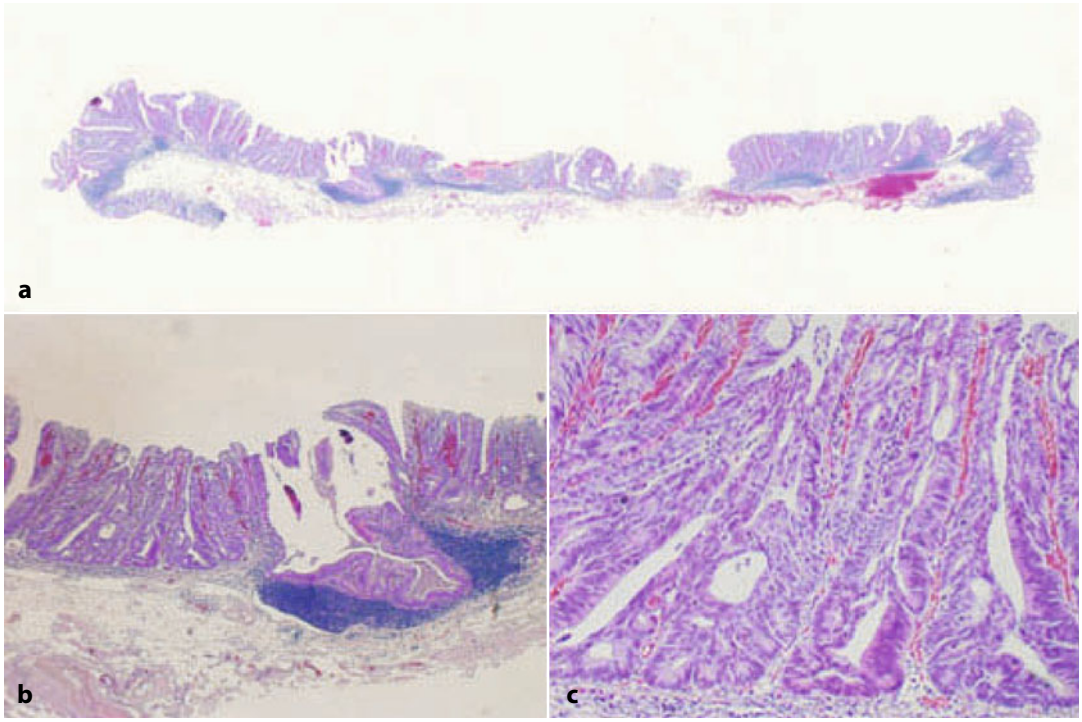

Fig. 5.13 a Histologically, the lesion was shown to have been completely resected by EMR. **b** Well differentiated adenocarcinoma slightly invading the submucosal layer. **c** High power view of the well differentiated adenocarcinoma

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Multiple Choice Questionnaire

1) What is the incidence of laterally spreading tumor, granular type among all colorectal lesions?
a. 10%
b. 20%
c. 30%
d. 5%

2) Which is the more frequent site of a laterally spreading tumor?
a. left colon
b. rectum
c. cecum and rectum
d. sigmoid colon

3) Which is the most frequent location of tumors with a flat-type appearance?
a. transverse colon
b. right colon
c. sigmoid colon
d. rectum

4) Which is the most frequent location of tumors with a polypoid-type appearance?
a. transverse colon
b. right colon
c. left colon
d. rectum

5) What are the current indications for colorectal endoscopic submucosal dissection?
a. polypoid lesions <40 mm in diameter
b. all laterally spreading tumors and non-polypoid lesions
c. laterally spreading tumors located in the rectum
d. lesions with submucosal invasion ≤1000 μm, with negative lymphovascular invasion and a well differentiated component

1.d – 2.c – 3.a – 4.c – 5.d

Case 5.3
Depressed-Type Adenoma of the Ileum

Shin-ei Kudo

Background

The prevalence of neoplastic lesions of the small bowel is quite low. Early detection of these small-bowel neoplasms is difficult as tumors of the small bowel grow slowly and extraluminally and remain asymptomatic for years. In addition, they often develop insidiously such that patients present with non-specific complaints such as abdominal pain, diarrhea, iron deficiency anemia, bleeding, and extra-intestinal symptoms.

Clinical Presentation

An 80-year-old man was admitted to our hospital with an adhesive intestinal obstruction. His intestinal obstruction was resolved after decompression with an ileus tube. Total colonoscopy was subsequently performed to investigate the cause of the obstruction. Insertion of the colonoscope into the terminal ileum incidentally detected a clearly demarcated, slightly reddish, 9-mm lesion with marginal elevation. Chromoendoscopy with indigo-carmine dye demonstrated a star-shaped depression within the lesion (Fig. 5.14), which was of macroscopic type 0-IIc according to the Paris classification [1]. The macroscopic appearance was similar to that of depressed-type early colorectal cancer, as described by Kudo [2]. Narrow-band imaging (NBI) showed a clearly demarcated brownish lesion (Fig. 5.15). On magnifying NBI, a regularly arranged network of vessels was seen that corresponded to the network pattern in the colorectal lesion reported by Wada et al. [3]. Magnifying chromoendoscopy with crystal violet revealed small, round, tubular glandular orifices resembling colon cancer within the depressed area [4] (Fig. 5.16).

Since the patient was on an anticoagulant, endocytoscopy was performed to evaluate the lesion in order to reach a decision regarding further management. The endocytoscopic images showed fusiform and regularly arranged nuclei along the basement membrane and a smooth and slit-like lumen (Fig. 5.17), remarkably similar to our endocytoscopic images of colonic adenoma and intramucosal neoplasia. Furthermore, these findings were not seen in the villous mucosa of the small intestine. Therefore, the diagnosis was either an adenoma or an intramucosal neoplasm of the ileum, and endoscopic mucosal resection was performed. Histological examination indicated a tubular adenoma with low-grade

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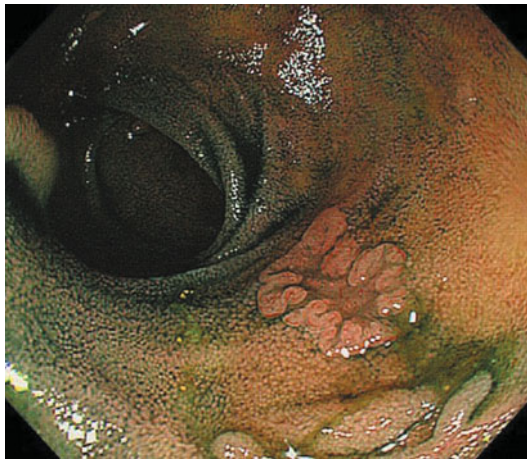

Fig. 5.14 Chromoendoscopy with indigo-carmine dye: star-shaped depression within the lesion

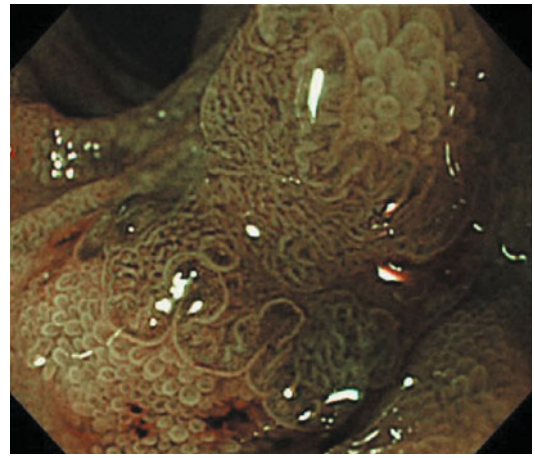

Fig. 5.15 Narrow-band imaging (NBI): clearly demarcated brownish lesion; magnifying NBI: regularly arranged network of vessels

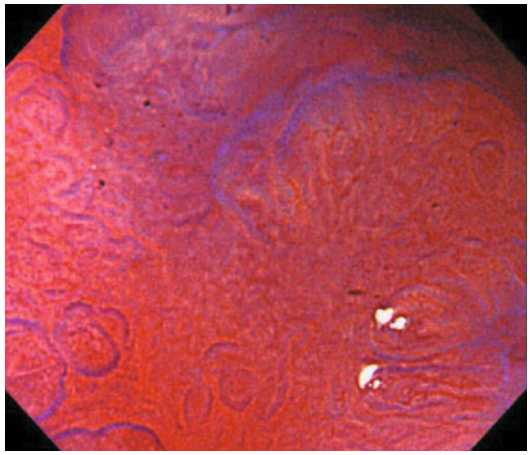

Fig. 5.16 Magnifying chromoendoscopy using crystal violet: within the depressed area, small, round, tubular glandular orifices resembling colon cancer

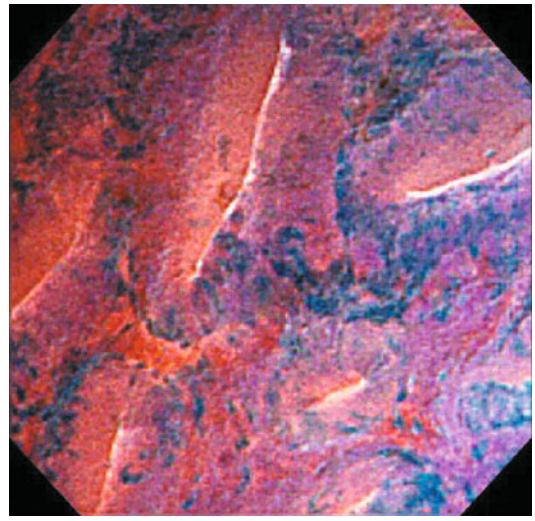

Fig. 5.17 Endocytoscopy: fusiform nuclei regularly arranged along the basement membrane, and a smooth and slit-like lumen

atypia, consistent with our endocytoscopic prediction (Figs. 5.18, 5.19).

Open Issues

Endocytoscopy, which involves a contact light microscopy system integrated into the distal tip of a conventional colonoscope, is a novel emerging endoscopic system. Unlike other modalities, the ultra-magnification capability of endocytoscopy enables on-site observation of both structural and cytological atypia [5, 6]. It may be particularly useful in patients who are on anticoagulant therapy, in whom routine biopsy for histological analysis may be haz-

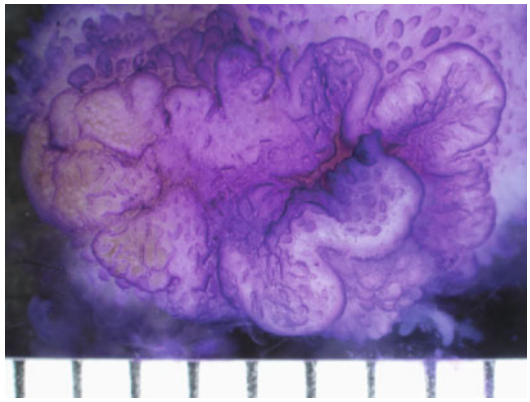

Fig. 5.18 Macroscopic view of the slightly depressed lesion

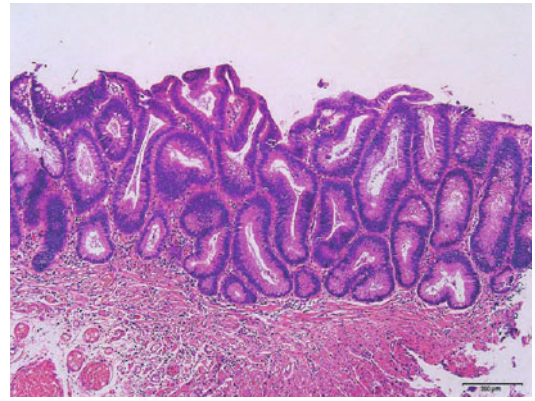

Fig. 5.19 Microscopic view of the resected specimen (hematoxylin-eosin staining). Diagnosis: tubular adenoma with low-grade atypia

ardous and further endoscopic treatment may be more difficult because of scarring.

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Multiple Choice Questionnaire

1) The prevalence rate of adenocarcinoma among small bowel malignant tumors is:
a. about 30-50%
b. about 10-20%
c. less than 5%
d. more than 5%
e. 15%

2) What is the percentage of small bowel tumors located in the ileum?
a. 0.1%
b. about 3%
c. 5%
d. 8%
e. 10%

3) What is the incidence rate of small bowel neoplasms as a percentage of all primary gastrointestinal tumors?
a. 5%
b. more than 10%
c. 10-30%
d. 2%
e. less than 5%

1.a – 2.b – 3.c

Case 5.4
Neoplastic Disease of the Ileocecal Region

Makomo Makazu, Takahisa Matsuda, Taku Sakamoto, Takeshi Nakajima and Yutaka Saito

Background

Relative to the length and surface area of the small intestine, malignancies at this site account for only 3% of all gastrointestinal tract cancers and less than 0.5% of all cancers in the United States [1]. However, the incidence of malignancies in the small intestine appears to be increasing [2]. With recent developments in the technology of digestive endoscopy, such as double-balloon enteroscopy and capsule endoscopy, the detection rate of small intestinal neoplasms is expected to increase further. Therefore, state-of-the-art knowledge of neoplastic diseases in this anatomical area is of utmost importance.

Clinical Presentation

Case 1

A 64-year-old woman underwent a total colonoscopy because of a positive fecal occult blood test. An elevated lesion, 30 mm in diameter, with a nodular surface was detected on the ileocecal valve (Fig. 5.20a). Narrow-band imaging (NBI) with magnifying colonoscopy revealed slightly dilated capillary vessels (Fig. 5.20b). NBI also showed that the tumor extended back into the ileocecal valve. The scope was therefore inserted into the terminal ileum, which made it clear that the tumor we recognized as a cecal tumor was actually a pedunculated tumor originating from the ileum. Magnification with chromoendoscopy using indigo-carmine dye showed a gyrus-like pattern, consistent with the type IV pit pattern according to Kudo's classification of colorectal tumors [3] (Fig. 5.20c). The tumor was expected to be an adenoma or at least carcinoma in situ. Surgery was chosen over endoscopic resection because it was difficult to observe the base of the tumor stalk and the tumor size was relatively large. Ileocecal resection was performed (Fig. 5.20d). Histologically, the lesion was identified as a tubular adenoma with low-grade atypia (Fig. 5.20e).

Case 2

A 64-year-old woman underwent a total colonoscopy as part of a routine medical evaluation. An elevated lesion, 20 mm in diameter, with a lobulated surface was detected on the ileocecal valve (Fig. 5.21a). NBI with magnifying colonoscopy did not evidence any remarkable capillary vessels (Fig. 5.21b).

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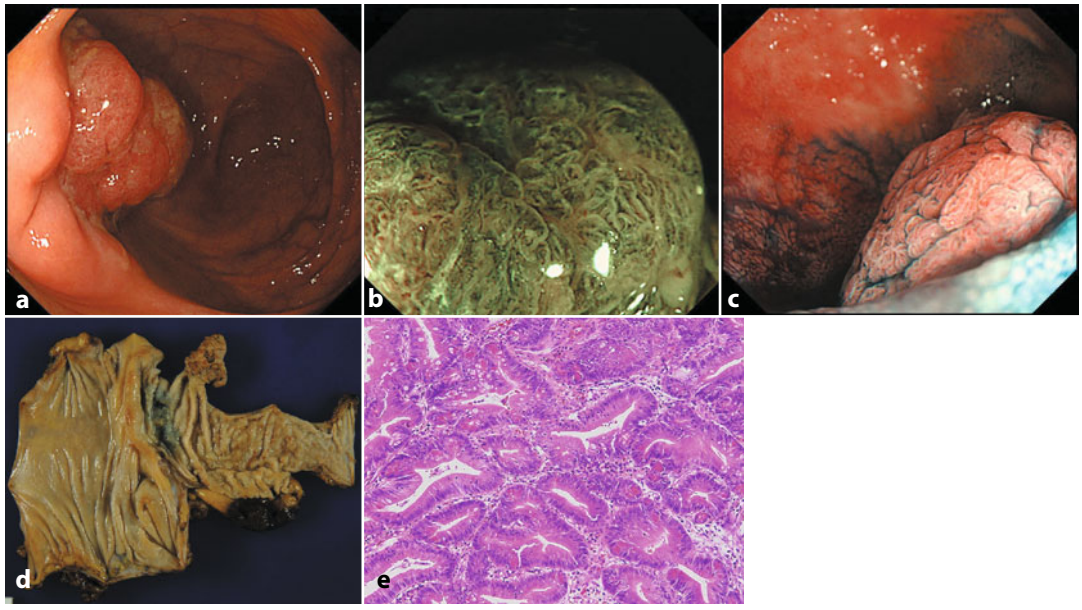

Fig. 5.20 a An elevated lesion, 30 mm in diameter, with a nodular surface was detected on the ileocecal valve in a 64-year-old woman who underwent colonoscopy because of a positive fecal occult blood test. **b** Narrow-band imaging with magnifying colonoscopy revealed slightly dilated capillary vessels. **c** Magnification with chromoendoscopy using indigo-carmine dye showed a gyrus-like pattern. **d** The surgically resected specimen. **e** The lesion was identified as a tubular adenoma with low-grade atypia

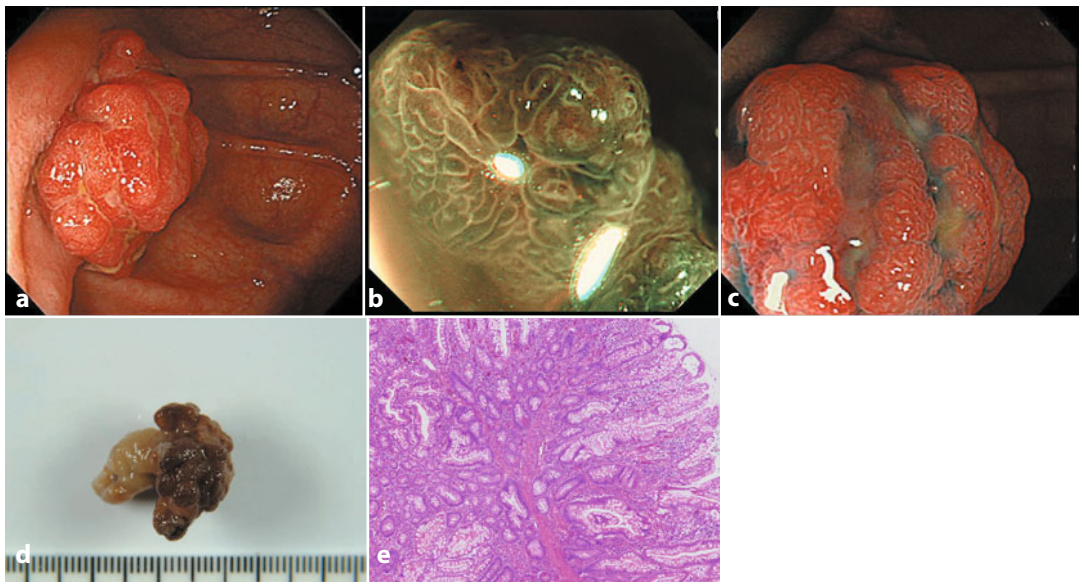

Fig. 5.21 a An elevated lesion, 20 mm in diameter, with a lobulated surface was detected on the ileocecal valve in a 64-year-old woman who underwent routine colonoscopy. **b** Narrow-band imaging with magnifying colonoscopy did not reveal remarkable capillary vessels. **c** Magnification with chromoendoscopy using indigo-carmine dye showed a villous surface pattern. **d** The endoscopically resected specimen. **e** The lesion was identified as a hamartoma

Magnification with chromoendoscopy using indigo-carmine dye spraying did not show a typical pit pattern, rather, only a villous surface pattern (Fig. 5.21c). On chromoendoscopy, the tumor was found to extend back into the ileocecal valve. The scope was therefore inserted into the terminal ileum, where a pedunculated tumor originating from the ileum was seen. The tumor was expected to be a hamartomatous polyp and was completely removed en bloc with endoscopic mucosal resection (conventional lift and cut technique) (Fig. 5.21d). Histologically, the lesion had branching muscularis mucosae with an overlying small-intestinal mucosa and was identified as a hamartoma (Fig. 5.21e).

Open Issues

Ileal pedunculated tumors sometimes prolapse through the ileocecal valve and mimic cecal lesions. Thus, for tumors located on the ileocecal valve or in the cecum their origin should be checked. If an ileal tumor is expected to be an adenoma or at least carcinoma in situ, endoscopic resection may be chosen. The surface and capillary vessel patterns of the tumor are useful in estimating its histological type and its depth. However, because of the villous structures of the small intestinal mucosa, findings relevant to the colorectal area are not directly applicable.

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Multiple Choice Questionnaire

1) What are the determinants of a "complete" total colonoscopy (more than one answer possible)?
a. scope insertion into the end of the ileum
b. scope insertion up to 80 cm
c. scope insertion up to 100 cm
d. observation of the orifice of the appendix
e. observation the Bauhin valve

2) Which is the actual incidence of small bowel malignancies?
a. 3%
b. 5%
c. < 10%
d. 15%

3) What is the current rate of recurrence after colorectal piecemeal resections?
a. >10%
b. <5%
c. <10%
d. 20%

1.a – 2.a – 3.a

Case 5.5
Chronic Diarrhea: The Importance of Terminal Ileoscopy

Antonello Trecca, Pasquale Trecca, Francesca De Laurentiis and Giancarlo D'Ambrosio

Background

Carcinoids are rare tumors that arise from neuroendocrine cells throughout the body [1]. Their incidence seems to have increased over the past few decades (5.25/100,000 in 2004 in the USA), mainly because of more accurate diagnostic devices [2].

In the USA, carcinoids originating in the small intestine have a reported incidence of 0.67/100,000. They comprise the second largest subgroup after those of pancreatic origin and are probably the largest subgroup of carcinoids within the gastroenteropancreatic system [3].

Recently, many techniques for the detection of these tumors have been introduced into clinical practice but diagnosis remains a matter of concern. The following case report deals with a patient whose carcinoid was incidentally detected during an ileoscopy.

Clinical Presentation

A 62-year-old female patient with a 5-kg weight loss and a 6-month history of diarrhea not responsive to loperamide was referred to our institution. She also reported lactose intolerance and intermittent mild crampy abdominal pain. Based on this history, she was started on medical treatment. Blood tests showed unremarkable findings.

A colonoscopy was carried out and the terminal ileum was intubated. A submucosal mass with a deep ulceration was identified at a distance of 4 cm from the Bauhin valve (Fig. 5.22). The obstructive mass determined a substenosis of the terminal ileum, but exploration of the other 10 cm of the ileum was negative.

Multiple biopsies were taken, but the histological report was negative. The patient subsequently underwent a CT scan, which revealed multiple hepatic metastases (Fig. 5.23).

Surgical intervention consisted of a right hemicolectomy including a resection of the last 10 cm of the terminal ileum. The more superficial of the liver metastases was also biopsied.

Analysis of the specimen defined a tumor stage of T2N1M1 according to the AJCC classification, with a proliferation index of 5% (Fig. 5.24).

After an uneventful postoperative course, the patient was discharged. One month later a biological therapy was started using longastatin, 20 mg i.m. once a month. The diarrhea disappeared and the patient was in good general condition without any symptoms. No side effects were reported.

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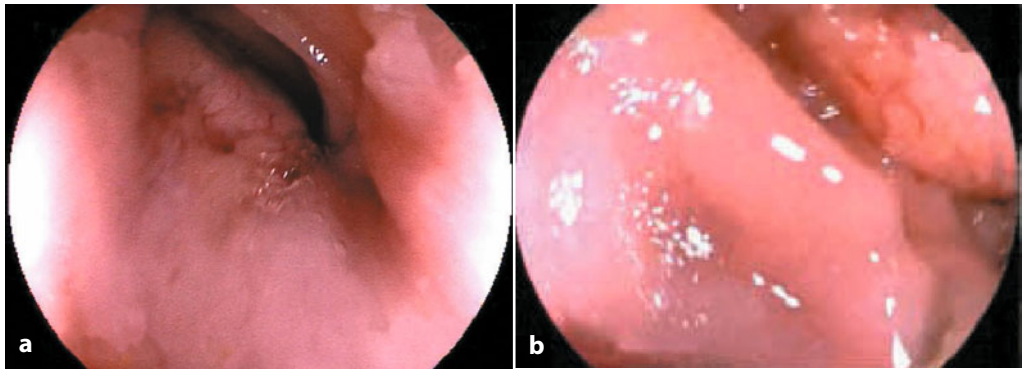

Fig. 5.22 Ileoscopic view of the submucosal mass involving the majority of the intestinal lumen

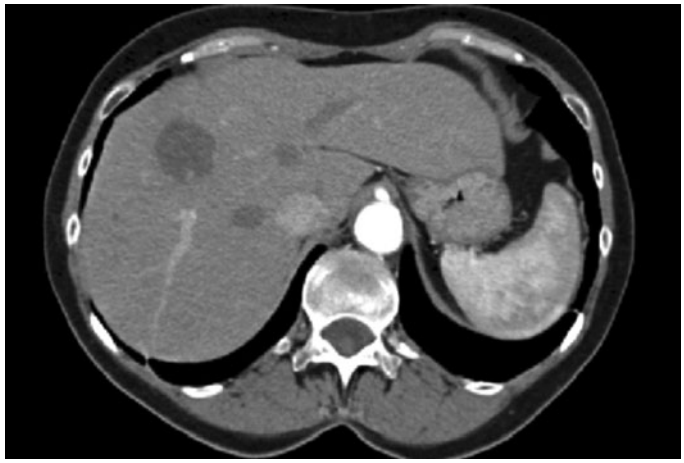

Fig. 5.23 CT scan of the liver showing multifocal metastatic disease

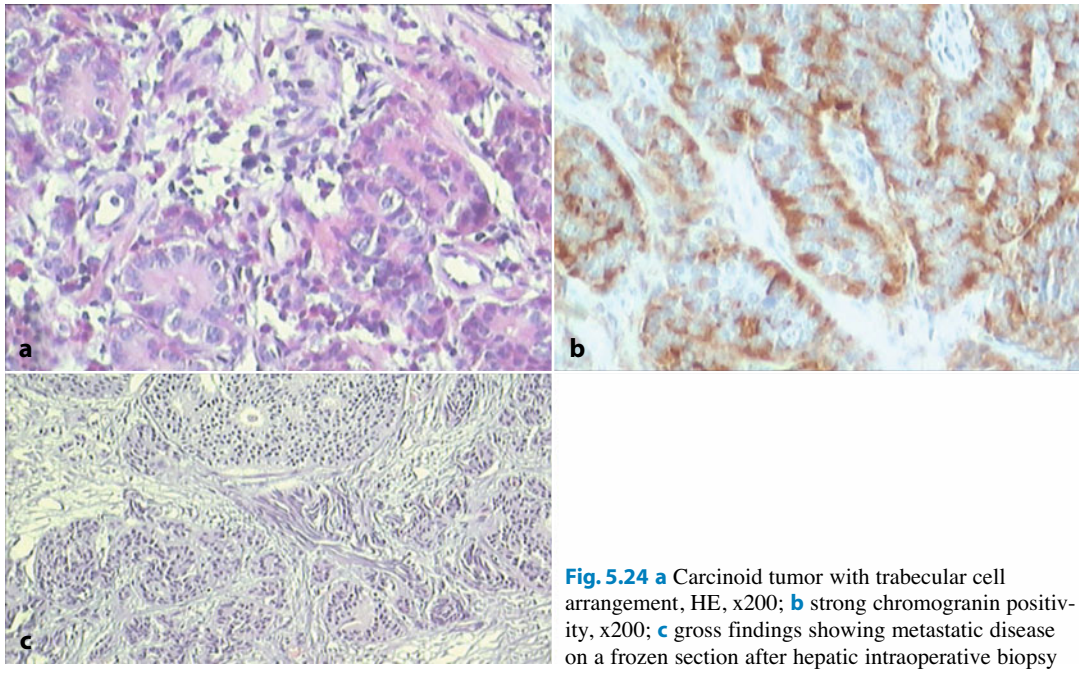

Fig. 5.24 a Carcinoid tumor with trabecular cell arrangement, HE, x200; **b** strong chromogranin positivity, x200; **c** gross findings showing metastatic disease on a frozen section after hepatic intraoperative biopsy

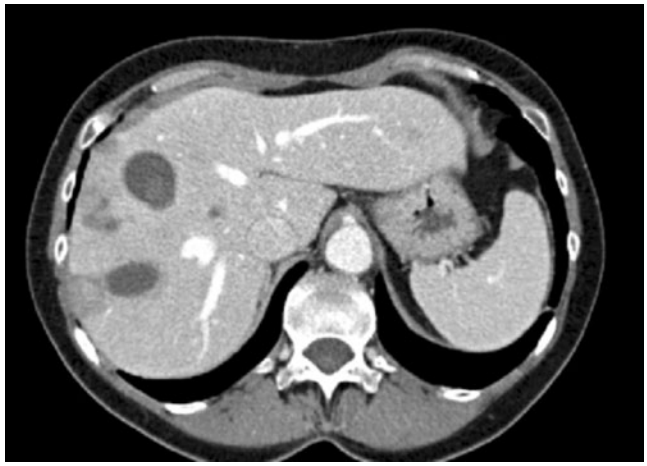

Fig. 5.25 CT scan after chemoembolization and thermoablation

Due to an increased size of three metastases, chemoembolization and thermoablation of the lesions were carried out after 4 months of the above-mentioned medical treatment.

Liver CT scan was repeated and showed no further increase in the size of the metastases (Fig. 5.25).

Fifteen months after the diagnosis, the patient was symptom free, in good general condition and with normal bowel function.

Open Issues

This case confirms once more the importance of exploring the terminal ileum in patients with chronic diarrhea and particularly when colonoscopy does not detect any lesion. Total colonoscopy without ileoscopy would have missed the diagnosis of an advanced disease and an earlier lower gastrointestinal endoscopy would have avoided a late diagnosis. In our patient, the ileoscopy found the tumoral mass involving the terminal ileum, in addition to revealing ileal substenosis and the presence of a deep ulcer in the upper part of the mass, together allowing a presumptive diagnosis [4].

However, ileoscopy has several limitations: i) a wide biopsy does not provide a definitive diagnosis; ii) as more than 20% of these tumors are multifocal, imaging of the entire small bowel is mandatory, even if the presence of liver metastasis suggests the need for immediate surgical treatment to avoid the risk of intestinal obstruction [5]. Medical treatment seems to be effective even in advanced disease, as 15 months after the initial diagnosis the patient was stable and in good general condition [6, 7].

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Multiple Choice Questionnaire

1) What is the clinical incidence of all carcinoid tumors?
a. 2 per 100,000 per year
b. 5 per 100,000 per year
c. 10 per 100,000 per year
d. 20 per 100,000 per year
e. 50 per 100,000 per year

2) Which percentage of small bowel carcinoids are multifocal?
a. 5%
b. 10%
c. 15%
d. 25%
e. 50%

3) Which organ is the most frequently involved in carcinoid tumors?
a. lungs
b. pancreas
c. liver
d. large bowel
e. small bowel

1.a – 2.d – 3.e

Case 5.6
Ileal Neuroendocrine Tumor: Clinical Case and Hot Topics

Prashant Kant, Nigel Scott and Bjorn J. Rembacken

Background

Colonoscopy is frequently requested as a first-line investigation for patients with long-standing lower gastrointestinal symptoms. Whilst great emphasis is currently placed on achieving successful caecal intubation, relatively neglected is the importance of intubation and visualisation of the terminal ileum (TI), particularly if the colonic mucosa and ileo-caecal valve appear normal. In one large series of 21,638 colonoscopies, the authors reported an overall TI intubation rate of only 18% [1]. Yet, the yield of pathology detected in the TI following normal colonoscopy is reportedly as high as 22% [2]. Whilst the majority of ileal pathology often involves inflammatory changes that lead to the diagnosis of Crohn's ileitis, infrequent findings also include asymptomatic neoplastic lesions. Here we present the unexpected finding of an ileal lesion and point out the important clinical implications.

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Clinical Presentation

A 45-year-old woman presented with a long history of erratic bowel habit, worsening over the preceding 18 months but with no blood or mucous in her stools. She also reported left iliac fossa pain and had a mild microcytic anaemia but no weight loss.

At an initial colonoscopy the colon was unremarkable. However, a 6-mm, subpedunculated (Paris classification 1sp), submucosal polyp was found approximately 10 cm from the ileo-caecal valve. Biopsies from the polyp showed a normal ileal mucosa.

In view of the persisting symptoms a magnetic resonance (MR) enterography was arranged, which could not definitively identify the ileal polyp (Fig. 5.26). Consequently, the polyp was referred for specialist endoscopic characterisation and resection.

On the follow-up assessment, the lesion was determined to be a firm nodule rather than a lipoma, and endoscopic mucosal resection was undertaken (Fig. 5.27). The polyp was removed as an 8-mm single fragment. Subsequent histology confirmed a well-differentiated neuroendocrine tumour (NET) without vascular channel invasion. Immunohistochemical markers demonstrated a chromogranin-positive lesion with a low mitotic index (Ki-67 index = 2%), confirming a grade 1 NET (Fig. 5.28). As the lesion extended to the deep resection margin, it was reported as

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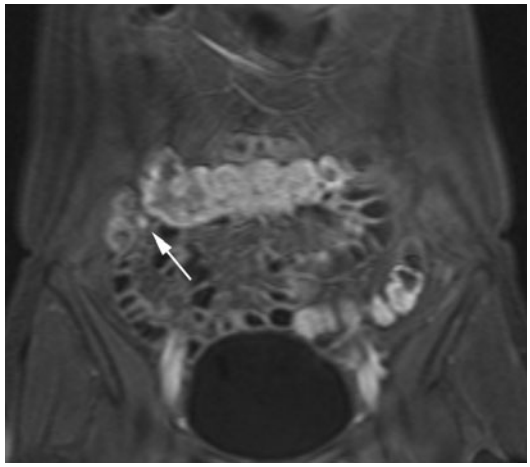

Fig. 5.26 Enhanced T1 magnetic resonance image. The coronal image shows the terminal ileum and proximal colon, with the neuroendocrine tumor indicated by the *white arrow*

disciplinary team, a limited right hemi-colectomy with resection of the terminal ileum was decided upon. A laparoscopic hemicolectomy was performed, removing 95 mm of the TI with draining lymph nodes. No residual NET was found in the specimen. Metastatic neuroendocrine carcinoma was confirmed in one of the 14 lymph nodes resected. The tumour was ultimately staged as T1 N1 V0 R0.

Open Issues

Small-bowel tumors are relatively rare in the gastrointestinal (GI) tract. NETs were originally described over a hundred years ago and

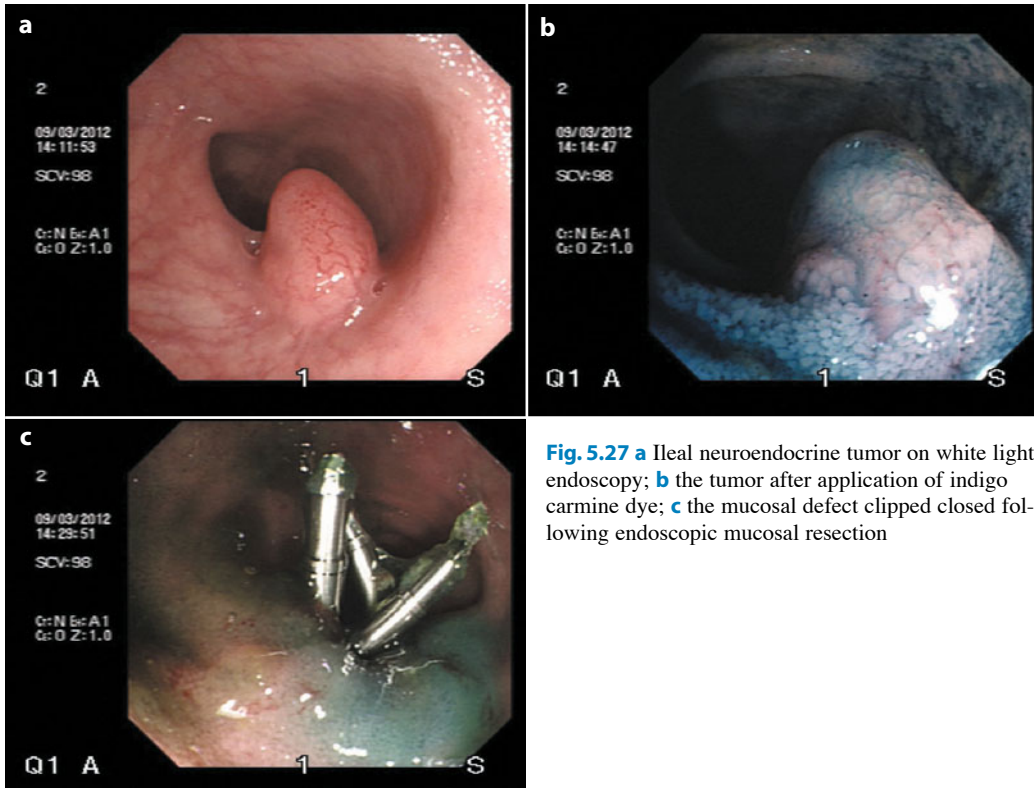

Fig. 5.27 a Ileal neuroendocrine tumor on white light endoscopy; **b** the tumor after application of indigo carmine dye; **c** the mucosal defect clipped closed following endoscopic mucosal resection

incompletely excised. A subsequent CT scan was reassuring, showing no abnormalities at the TI or surrounding lymph nodes. The serum chromogranin A level was normal.

On further review by the colorectal multi-

were previously considered to be a rare occurrence. They are frequently referred to as "carcinoid", implying a carcinoma-like appearance yet a benign course. NETs arise from diffuse enterochromaffin cells and may

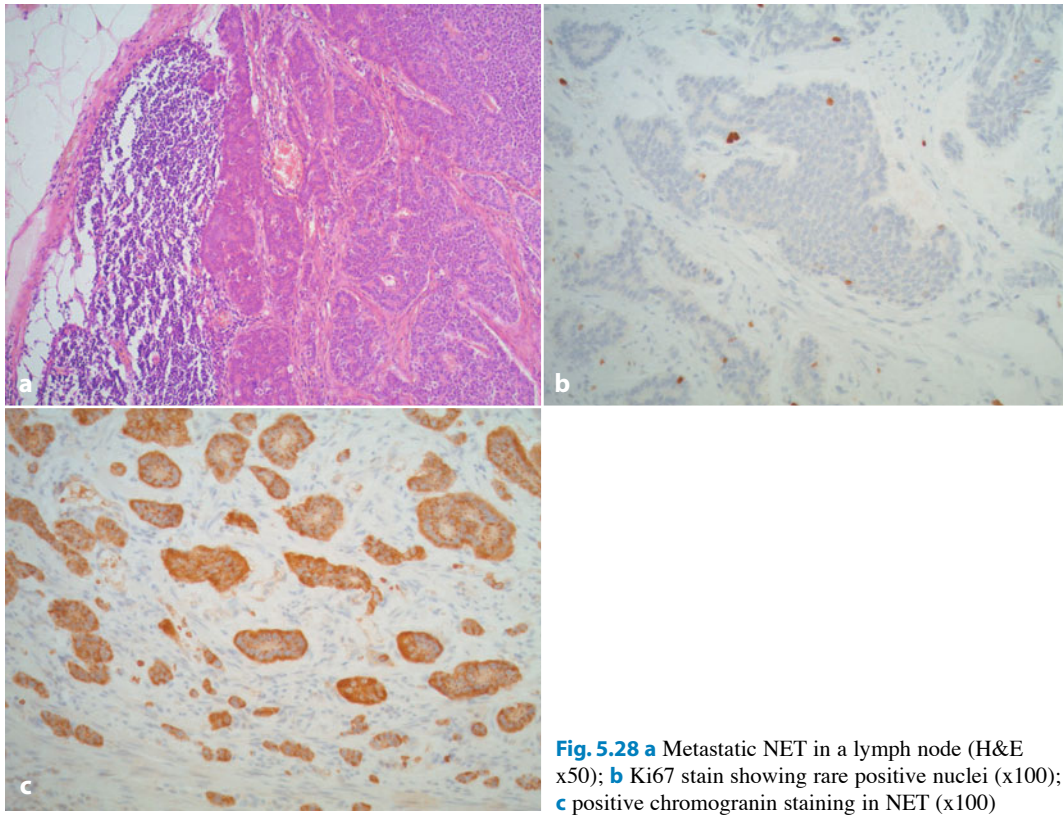

Fig. 5.28 a Metastatic NET in a lymph node (H&E x50); **b** Ki67 stain showing rare positive nuclei (x100); **c** positive chromogranin staining in NET (x100)

affect various organs in the GI tact, bronchus, thymus, kidney, ovary and testes, but 75– 90% occur in the GI tract.

The "mid-gut" is the most common primary site and about 38% of all GI NETs develop in the small intestine, usually in the distal ileum [3]. Mid-gut NETs are typically small, 47% are less than 20 mm in diameter, and they are usually covered by a normal smooth mucosa. As the accessibility of the small bowel has improved in recent decades, so too has the awareness of this group of lesions and the recognition of their great malignant potential. In fact, the majority of mid-gut NETs are malignant, with a reported mean 5-year survival rate of 63% [3]. Between 25% and 30% of these tumors are multiple and there is a significant association with other GI tract malignancies [4].

The age-adjusted incidence of ileal NETs has risen over three-fold over the last 30 years [3]. This apparent increase can most likely be attributed to the increased awareness of the condition and to the increased utility of enteroscopy, ileo-colonoscopy, capsule endoscopy and higher resolution cross-sectional imaging. The true prevalence of these lesions remains unknown. In one study of 4,000 routine colonoscopies with a TI intubation rate of 80%, five NETs were detected [5].

Morphologically, the tumors arise submucosally, and are characteristically slow growing. Early cases are usually asymptomatic, as was the case with this lesion and diagnosis is often late. Patients with malignant disease usually present in the fifth and sixth decade of life and the average time from onset of symptoms to diagnosis exceeds 9 years [6]. Approximately 10% of patients develop hepatic metastases that result in the carcinoid syndrome, a constellation of flushing, secretory diarrhea, telangiectasia and bronchial constriction caused by

the release of numerous vasoactive peptides which have bypassed hepatic metabolism.

Clinico-pathological properties vary with the site of the lesion. As opposed to NETs of the rectum, appendix, pancreas or bronchus, small bowel carcinoids have a high rate of transmural invasion and are more likely to metastasise. Multiplicity rates are also greater for small bowel NETs, ranging from as low as 2–4 % for rectal tumors but as high as 40 % for small bowel tumors [7]. Patients with multiple small bowel NETs are younger, have a greater risk of developing carcinoid syndrome and have a poorer prognosis than patients with solitary tumors [8].

Within the small intestine itself, jejuno-ileal NETs differ from those occurring in other sites of the gut. Whilst duodenal tumors are often detected early, owing to the relative ease of discovery at routine upper GI tract endoscopy, jejuno-ileal lesions are often discovered either after resection for small bowel obstruction or after exploratory surgery for an unknown primary tumor after distant metastases have occurred. Due to the rarity of NETs, very little data are available regarding the incidence of small intestinal NETs, although two large series from Japan suggest that between 0.2% and 8% of all small intestinal malignant tumors are NETs [9]. Another series of 167 cases of jejuno-ileal carcinoid highlighted the clinicopathological differences between jejunal and ileal lesions. Small intestinal NETs occur 6.5 times more frequently in the ileum than in the jejunum [10]. In addition, small bowel NETs have a strong association with synchronous tumors of the gut, with the frequency of second malignancies ranging from 10% to 29% and most commonly being colorectal adenocarcinomas [10].

In the current case the tumor was well differentiated (grade 1), small and limited to the submucosa. Nevertheless, a lymph node metastasis was found at subsequent surgery. Although endoscopic resection is usually curative for type 1 and 2 NETs of the stomach, appendix tumors < 10 mm in size and small well differentiated NETs of the rectum, there is a substantial risk of nodal metastasis in early-stage small bowel carcinoids such that intestinal resection with wedge resection of the adjacent mesentery is usually required [11]. The guidelines of the British Society of Gastroenterology for the management of gastroentero-pancreatic neuroendocrine tumors specifically recommends surgical resection of small bowel NETs [12]. In a series of 167 carcinoid tumors of the jejunum and ileum collected by the AFIP, regional lymph node metastases were found in 21% of the tumours < 10 mm in diameter and 17% of the 24 tumours confined to the submucosa [10]. Sutton and colleagues reported an even higher rate of 40% lymph node involvement in NETs < 10 mm in their review of midgut carcinoids [13].

Given their relatively low incidence and the likelihood of occurrence in endoscopically remote reaches within the small intestine, there are no published series on endoscopic mucosal resection (EMR) of NETs of the small intestine. EMR of rectal NETs, the majority of which are 10 mm or less in size, restricted to the submucosal layer and without evidence of metastases, is relatively safe. The rectum is relatively fixed to the retroperitoneum, allowing safer endoscopic resection, with less risk of perforation assuming the use of techniques incorporating conventional polypectomy, band-ligation and endoscopic submucosal dissection. This is in contrast to the experience reported with the much thinner lumen of the small intestine [14]. The small intestinal lumen is less than half the thickness of the rectum [15] and EMR carries greater challenges and higher risks of complications. While the current data on complications stems from published series of EMR of sporadic duodenal adenoma and thus may not be directly applicable to NETs, they offer the closest approximation of the procedural risks. In large series, perforations occur in approximately 0.7% of cases, whilst intraprocedural bleeding managed endoscopically occurs in approximately 9%. Delayed bleeding (up to 2 weeks post-EMR) is estimated to occur in 4.4% of cases, corresponding to an overall complication rate of 14.3% [16].

Ileal tumours are rare but carry significant malignant burden, often presenting late. Around 22% of these patients present with distant metastases and in half no primary tumour can be found [17]. This case illustrates the value of routine ileoscopy, which resulted in the detection of a small NET that was too small for radiologic detection.

Ileoscopy is a relatively neglected quality indicator of a complete colonoscopy. Reasons for poor intubation rates include perceived technical challenges, thus adding to the procedure time, and patient discomfort. In fact, however, with practice, TI intubation can be achieved with an 80% success rate while adding only a median 1.5 minutes to the procedure [18]. A normal colonoscopy in patients with lower GI symptoms diminishes the index of suspicion for genuine pathology. It is recommended that ileoscopy is routinely performed in all patients to improve quality assurance and the diagnostic yield.

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Multiple Choice Questionnaire

1) Which one of the following symptoms is not recognised to be part of the carcinoid syndrome?

a. diarrhea
b. flushing
c. bronchospasm
d. left-sided valvular lesions
e. pellagra

2) Ileal carcinoids are mostly asymptomatic, although abdominal pain is a common presentation. What is the suspected mechanism?

a. intussusception
b. ischaemia
c. obstruction
d. hormone release
e. adhesions

3) Which of the following risk factors for mortality is incorrect?

a. male gender
b. distant metastases
c. carcinoid syndrome
d. ki67 index greater than 1%
e. solid growth pattern

1.e – 2.b – 3.a

Case 5.7
A Pedunculated Adenoma in the Terminal Ileum

Kuangi Fu and Takayoshi Shimizu

Background

Tumors of the small intestine are rare. Among all such tumors, about 25% are benign, with smooth muscle tumors as the most frequently detected type. Generally, adenomas are commonly seen in the large intestine whereas those in the small intestine, accounting for about 10% of the benign tumors, are rarely encountered endoscopically. While the duodenum is the most frequently involved site, an ileal adenoma is extremely rare. During colonoscopy, intubation of the terminal ileum remains controversial, as the diagnostic value is limited [1]. Moreover, it is often technically difficult to reach the terminal ileum, especially when patients are in a lateral position.

Clinical Presentation

An 82-year-old man presented to our department with a chief complaint of hematochezia and anemia. He had hypertension and had previously suffered a cerebral infarction and was now under medication. He had no family history of cancer. Esophagogastroduodenoscopy revealed a healing duodenal ulcer. Ileoscopy was easily achieved with a transparent hood (D-201-12704, Olympus, Tokyo, Japan) attached to a pediatric colonoscope (PCF-260AI, Olympus, Tokyo, Japan). A pedunculated polyp, about 8 mm in size, was detected in the terminal ileum, about 3 cm proximal to the ileocecal valve (Fig. 5.29). There was no other lesion responsible for the bleeding. Chromoendoscopy with 0.4% indigo carmine dye disclosed long and branching crypt orifices, with a type IV pit pattern according to Kudo's classification. Accordingly, a tubulovillous or villous adenoma was estimated endoscopically (Fig. 5.30). Although the patient was taking aspirin for anticoagulation therapy, a complete polypectomy, in which an endoclip was placed at the base of the stalk, was uneventful (Fig. 5.31). Another endoclip was added for prophylactic reasons (Fig. 5.32). No complication, such as post-polypectomy bleeding or perforation, was identified. Histologically, the removed specimen was a tubulovillous adenoma with high-grade atypia.

Open Issues

Due to its perceived technical difficulty, intubation of the ileum during colonoscopy is not routinely performed; rather, it is done on a

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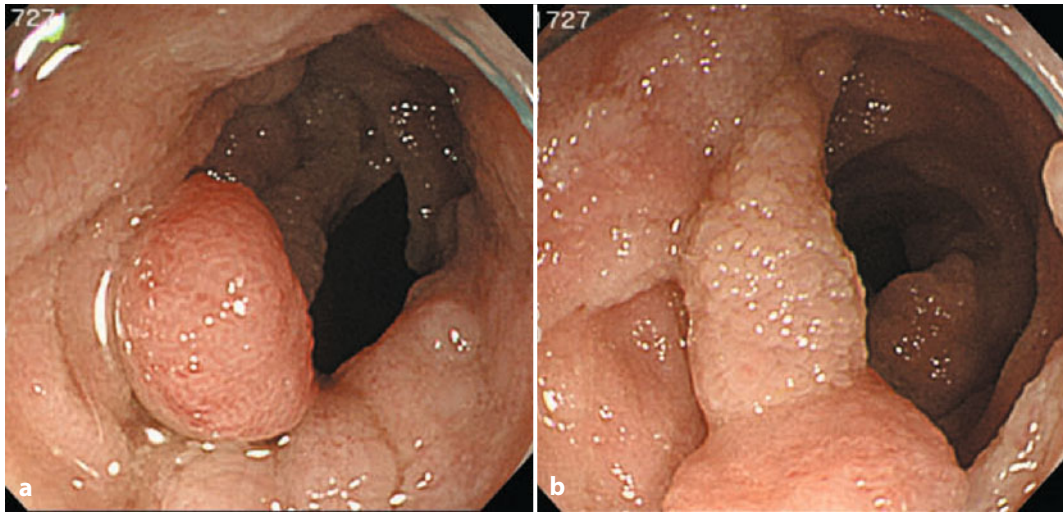

Fig. 5.29 A pedunculated polyp, about 8mm size, was detected in the terminal ileum about 3cm proximal to the ileocecal valve

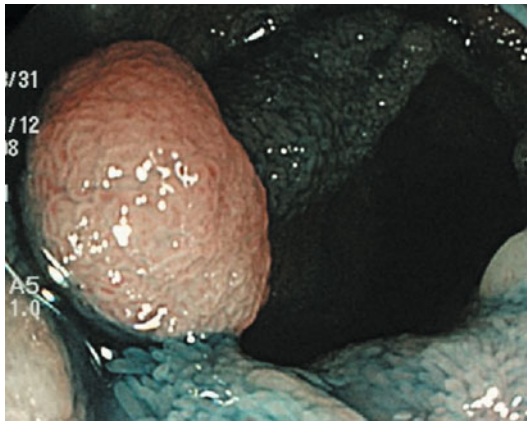

Fig. 5.30 Chromoendoscopy with 0.4% indigo carmine dye disclosed long and branching crypt orifices which were classified as type IV pit pattern according to Kudo's classification, and thus a tubulovillous or villous adenoma was estimated endoscopically

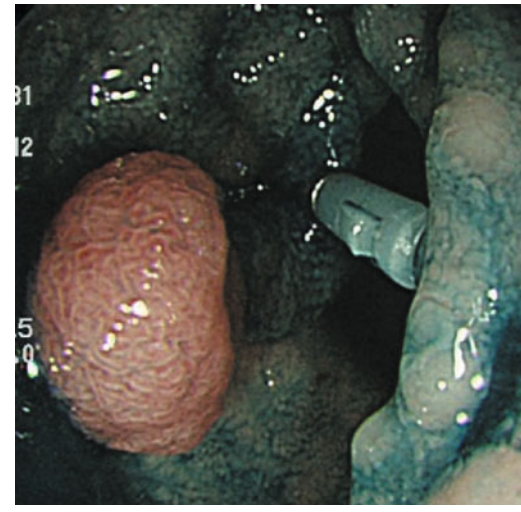

Fig. 5.31 An endoclip was placed at the base of the stalk before polypectomy

case-by-case basis [1]. In our patient, we used a transparent hood attached to the tip of a pediatric colonoscope, which facilitated ileocecal intubation. Although rarely encountered during ileoscopy, various intestinal disorders (Crohn's disease, tuberculosis, lymphoma, and cancer) can involve the ileocecal region. Moreover, intramucosal cancer or adenoma is exceptionally detected in this location [2, 3-6]. To the best of our knowledge, only five cases of adenoma or early cancer are reported in the English literature so far [3-6]. The details of these reported cases as well as ours are summarized in Table 5.1. All but one large lesions were successfully treated endoscopically. Although our patient was under antiplatelet therapy and the lesion was located in the terminal ileum, with its thin wall, the polyp could be safely removed with the assistance of prophylactically placed endoclips. Saline-assisted polypectomy (or EMR) is another choice for the treatment of this kind of lesion.

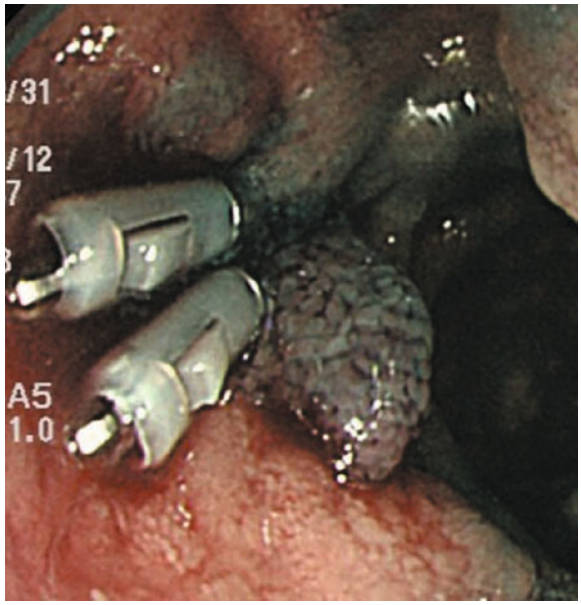

Fig. 5.32 Another endoclip was added with prophylactic intent after polypectomy

Table 5.1 Summary of reported cases of ileal adenoma

Case	Author	Age	Sex	Symptom	Macroscopic type	Size (mm)	Histology	Treatment
1	Tsuchida	73	M	Hematochezia	IIa	35	Adenoma	EMR
2	Ohwan	69	F	Abdominal pain	IIa+IIc	7	Tubular adenoma	EMR
3	Tsuchida	65	M	Hematochezia	Isp	7	Tubular adenoma	EMR
4	Tsuchida	65	M	None	Ip	6	Tubulovillous adenoma	EMR
5	OD Osflo	13	F	Abdominal pain	Not informative	550	Tubulovillous adenoma	Surgery
6	Our case	82	M	Anemia	Ip	8	Tubulovillous adenoma	Polypectomy

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Multiple Choice Questionnaire

1) Which is the incidence of benign small bowel tumors?

a. 20%
b. 30%
c. 10%
d. 5%

2) Which is the incidence of small bowel malignancies?

a. 5%
b. 30%
c. 10%
d. 5%

3) How many centimeters of terminal ileum can be visualized by ileoscopy?

a. no more than 20
b. up to 5
c. up to 10
d. more than 20

1.c – 2.b – 3.a

Case 6.1
Yersinia Enterocolitis

Kuangi Fu, Hironori Konuma and Ichiro Konuma

Background

Yersinia species are gram-negative coccobacilli that are facultative anaerobes. *Y. enterocolitica* and Y. *pseudotuberculosis* are the species that most commonly cause enterocolitis. Clinically, *Yersinia* enterocolitis leads to ileitis and mesenteric adenitis and thus is often misdiagnosed as acute appendicitis. Endoscopic findings include small ulcers mainly involving the ileocecal region such that the disease should be differentiated from inflammatory bowel disease [1, 2]. The final diagnosis is based on the isolation of *Y. enterocolitica* in the feces or biopsied tissues, e.g. obtained during endoscopic biopsy, and/or elevated serum antibody titers to the bacteria.

Clinical Presentation

A 14-year-old girl presented with a 1-week history of mild fever, abdominal pain, and watery diarrhea. On physical examination, tenderness without muscular defense or rebound tenderness was seen in the right lower abdomen. Laboratory data showed leukocytosis and increased C-reactive protein. Colonoscopy revealed a multiple, relatively uniform distribution of discrete, small round ulcers in the ileum end (Fig. 6.1), cecum (Fig. 6.2), and ascending colon (Fig. 6.3). Each small ulcer was covered with a yellowish exudate at the base with a raised erythematous margin. The small ulcers were distributed longitudinally and transversely. The swollen ileocecal valve (Fig. 6.4) and terminal ileum were also covered with the same diffuse small ulcers. The ulcers were not seen through the transverse colon to the sigmoid colon. The mucosa was edematous and mucosal vascularity was faint superficially (Fig. 6.5). No abnormal endoscopic findings were detected in the rectum (Fig. 6.6). Endoscopic biopsies taken from the terminal ileum and right-sided colon showed non-specific inflammation but granulomas were not identified histologically. Stool cultures were negative for various bacteria; ova, parasites, and *Clostridium difficile* toxin were also negative. Her medical history was unremarkable. She had no record of overseas travel before her symptoms developed. At first, a clinical diagnosis of inflammatory bowel disease (IBD) was made, and mesalazine (1200 mg/day) was prescribed. While her symptoms improved with the medication, she still complained of intermittent abdominal pain. Re-examination of the endoscopic biopsies disclosed an inflammatory change mainly consisting of neutrophils,

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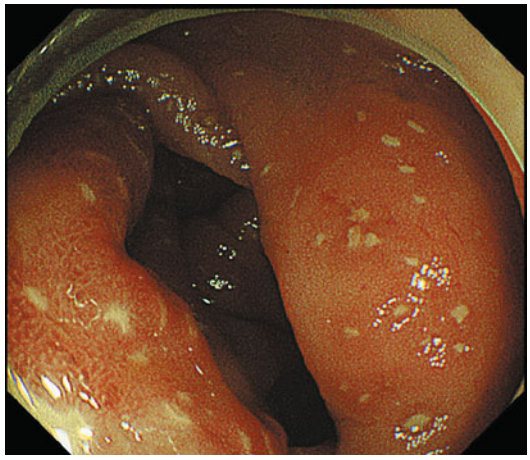

Fig. 6.1 Colonoscopy revealed multiple relatively uniform distribution of discrete, small round ulcers in the ileum end (*Fig. 6.1*), cecum (*Fig. 6.2*), and ascending colon (*Fig. 6.3*). Each small ulcer was covered yellowish exudate in the base with raised erythematous margin. The small ulcers distributed longitudinally and transversely

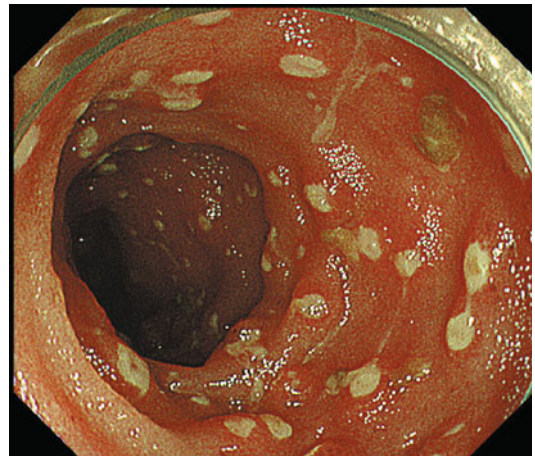

Fig. 6.2

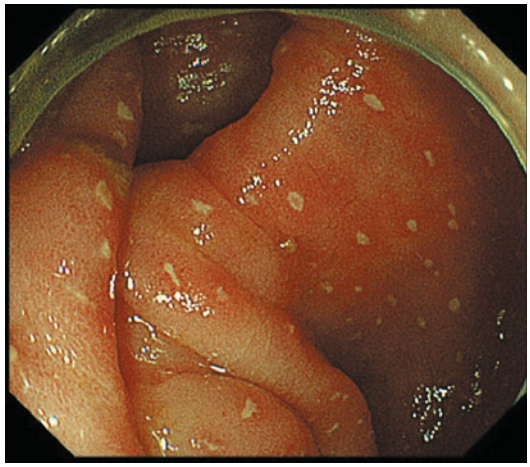

Fig. 6.3

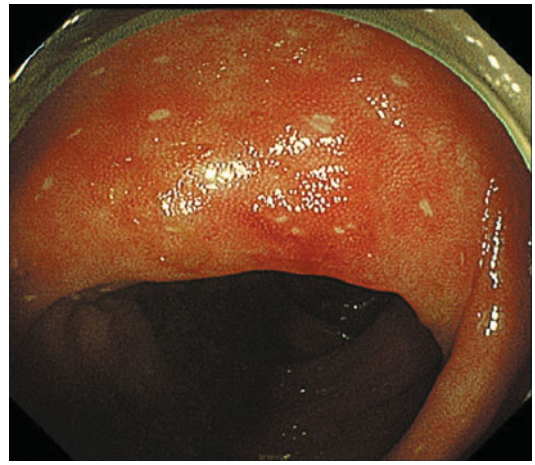

Fig. 6.4 The swollen ileocecal valve was also covered with same diffuse small ulcers

which suggested infectious enterocolitis rather than IBD (Fig. 6.7). Considering the involved sites and endoscopic findings in addition to the negative stool culture, a diagnosis of *Yersinia* enterocolitis was suspected. A serologic test revealed increased serum IgM antibody to *Y. enterocolitica* (1:320 in the bacterial agglutination test). A fluoroquinolone (levofloxacin hydrate, 500 mg/day) was prescribed for one week. Her symptoms subsided thereafter. A focused history-taking disclosed that she had not eaten uncooked meat food or unpasteurized milk, but she did have two dogs in her house, which suggested cross-contamination, although no one in her family exhibited similar symptoms.

Open Issues

There are no distinct clinical, radiologic, hematologic, or chemical findings that confirm yersiniosis. Generally, the diagnosis is established by culture isolation of the organism. Although stool culture is the most preferred method in the clinical setting, the cold-enrichment technique is necessary for culturing *Yersinia* species, which sometimes

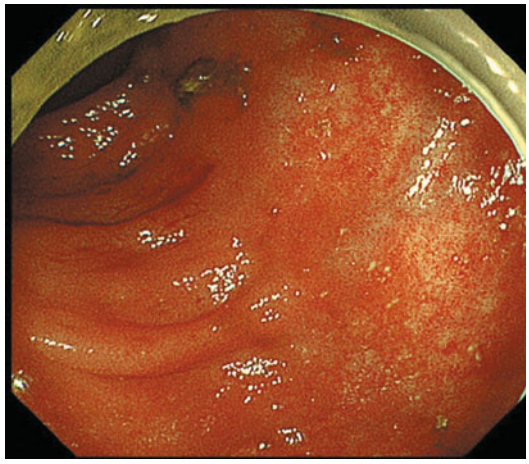

Fig. 6.5 Although no ulcer was seen through the transverse colon to the sigmoid colon, the mucosa was edematous and mucosal vascularity was faint superficially

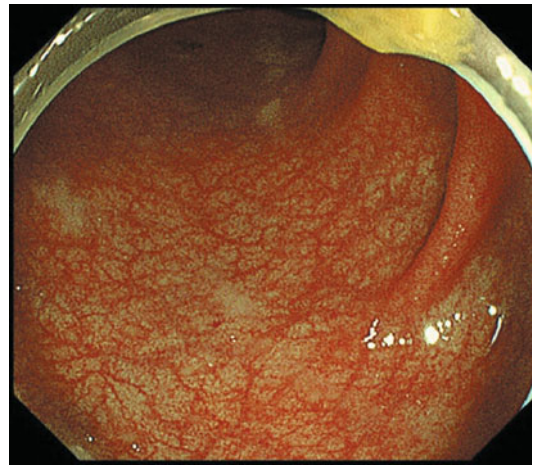

Fig. 6.6 No abnormal endoscopic finding was seen in the rectum

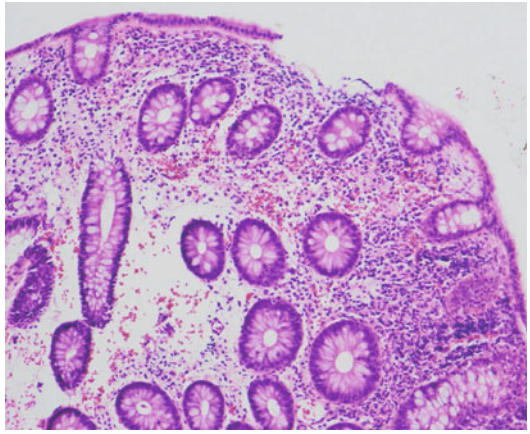

Fig. 6.7 The endoscopic biopsies disclosed that the inflammatory change mainly involved neutrophils, which suggested infectious enterocolitis rather than IBD

results the in failure of isolation [3]. In this case, elevated antibody titers are of high diagnostic value if the disease has been included in the differential diagnosis.

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Multiple Choice Questionnaire

1) How do you define *Yersinia Enterocolitica*?

a. gram-negative coccobacilli
b. gram-positive coccobacilli
c. parasite
d. gram-negative bacterium
e. virus

2) Which are the most common endoscopic findings of *Y. Enterocolitica*?

a. small ulcers of the ileocecal region
b. deep ulcerations of the terminal ileum
c. aftoid lesions of the terminal ileum
d. deep colonic ulcerations

3) How can you detect *Yersinia* infection?

a. isolation in the feces
b. endoscopic biopsy
c. elevated serum antibody titers
d. all of them

1.a – 2.a – 3.d

Case 6.2
Intestinal Occlusion-Like Syndrome Caused by Anisakiasis of the Small Bowel: Case Report

Paola Cesaro, Lucio Petruzziello, Cristiano Spada and Guido Costamagna

Background

Anisakis is a nematode parasite that may infect humans following the consumption of uncooked fish or seafood containing third-stage larvae of this nematode. Humans are only accidental hosts of the parasite. After ingestion, the larvae can penetrate the host stomach or intestinal wall. The resulting disease, known as anisakiasis or anisakidosis, is due mainly to two mechanisms: allergic reaction and direct tissue damage [1, 2]. The first case of a child vomiting an "ascarid" was described in 1876, but it was not until 1960 that the nematode was identified by Van Thiel in a patient from the Netherlands who had consumed raw herring [3-6].

Clinical Presentation

A 57-year-old otherwise healthy man was admitted to our hospital for epigastric pain, lower abdominal pain, and vomiting without diarrhea that had abruptly occurred in the afternoon of the day before. His symptoms had gradually worsened. He had no recent travel history, however he had eaten uncooked fish 2 days prior to admission. On admission he was conscious, his blood pressure was 130/90 mmHg, pulse 96 beats/min, and body temperature 37.8°C. He had no history of a laparotomy and was not taking medications. On physical examination, the patient's abdomen was tympanic and painful but there was no palpable mass. Peristalsis was heard on auscultation and there was flatus. Laboratory examinations showed a white blood cell count of 11 920/mm^3 with a normal eosinophil count. Plain radiography of the abdomen showed bowel air-fluid levels. An abdominal CT scan demonstrated a concentric wall thickening of the distal ileum, cecum, and appendix and a hyperdensity of the perivisceral adipose cells, with enlarged lymph nodes (up to 1 cm) in the mesenteric artery (Figs. 6.8, 6.9). On colonoscopy, the mucosae of the cecum and ileocecal valve were edematous, in the absence of visible lesions (Fig. 6.10). The distal ileum had an area of inflammation and mucosal edema that extended for 2–3 cm, with a slight reduction in the caliber of the lumen (Fig. 6.11). At this level, a cylindrical whitish threadlike worm, vital, and 2.5- to 3-mm long, was removed with biopsy forceps (Figs. 6.12, 6.13).

Histopathologic evaluation of endoscopically obtained biopsy specimens showed marked edema and inflammation and an

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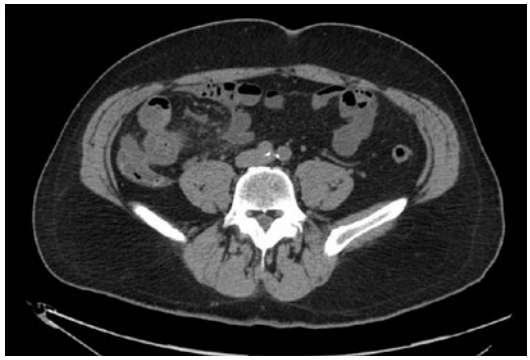

Fig. 6.8 Abdominal CT without contrast

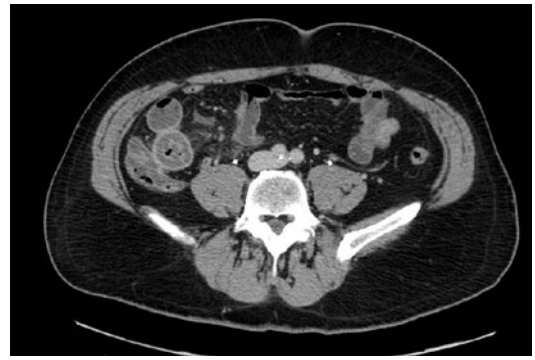

Fig. 6.9 Abdominal CT with contrast

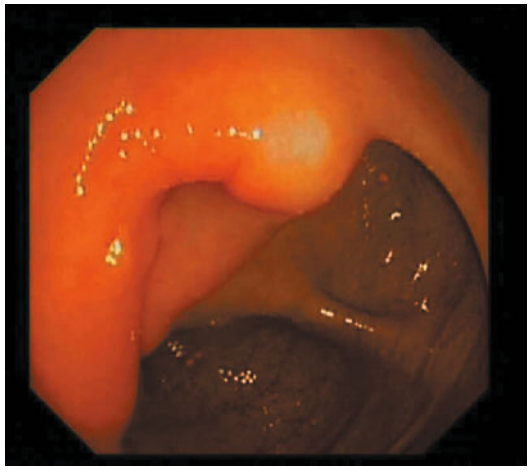

Fig. 6.10 Ileocecal valve

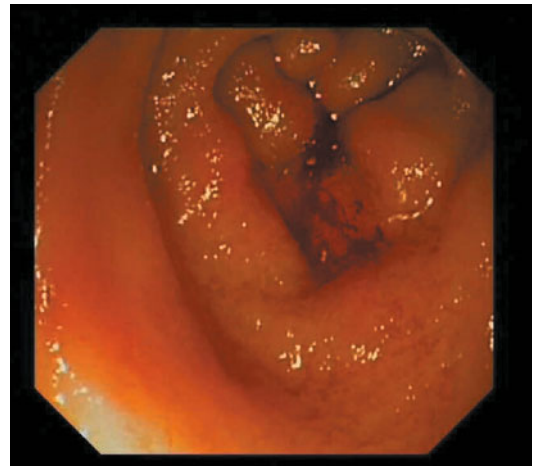

Fig. 6.11 The distal ileum

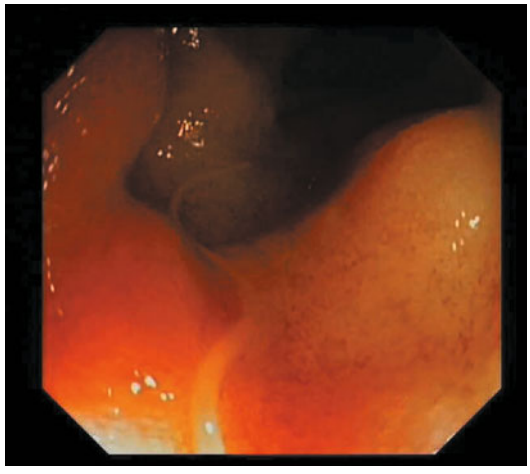

Fig. 6.12 A cylindrical whitish threadlike worm (Anisakis)

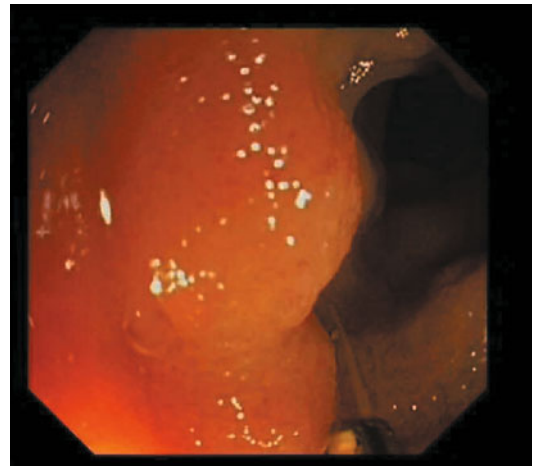

Fig. 6.13 Removal of Anisakis with biopsy forceps

eosinophilic cellular infiltration. The microbiological examination confirmed the pathogen as a larva of Anisakis *simplex*.

Abdominal pain and fever rapidly disappeared after removal of the worm.

Open Issues

The consumption of raw fish, such as sushi and sashimi, has been increasing worldwide, reflecting current food trends, but it has been associated with a higher risk of contracting a parasitic disease such as anisakiasis.

In most cases, anisakiasis occurs in the stomach. Enteric anisakiasis is very rare and its diagnosis is typically made only after laparotomy performed for an acute abdomen or intestinal obstruction [7, 8]. The disease should be suspected when abdominal symptoms occur following raw fish ingestion. An accurate endoscopy should be performed to remove the parasite [9] since an effective medical treatment of anisakiasis is not yet available.

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Multiple Choice Questionnaire

1) What is anisakiasis?

a. an infestation of third-stage larvae of *Anisakis*
b. a viral infection of the stomach and small bowel
c. an infestation with second-stage larvae of *Anisakis*
d. a gastrointestinal infection
e. a fungal infection of the stomach and small bowel

2) What is the most frequent pathogen responsible for anisakiasis?

a. *contracaecum* species
b. *anisakis pegreffi*
c. *anisakis simplex*
d. *thynnascaris* species
e. *anisakis physeteris*

3) What are the principal clinical manifestations of anisakiasis?

a. abdominal pain
b. vomiting
c. fever
d. depends on the localization
e. none of the above

4) What is the pathogenic mechanism of anisakiasis?

a. allergic reaction
b. direct tissue damage
c. both
d. neither
e. hemolysis

5) What is the most effective treatment for patients with anisakiasis?

a. laparotomy
b. endoscopic removal of the parasite
c. oral therapy with antibiotics
d. parenteral corticosteroids
e. albendazole

1.a – 2.c – 3.d – 4.c – 5.b

Case 6.3
An Unusual Guest in the Terminal Ileum

Gianfranco Tappero

Background

Tapeworm infection can pose a significant problem especially in developing countries [1]. Human contamination is rare but infected individuals may be asymptomatic [2]. *Taenia* species are recognized following the expulsion of eggs or proglottids in the stool of the host [3]. The following describes a case of *Taenia saginata* that was both identified and definitively treated by endoscopy.

Clinical Presentation

A 28-year-old woman underwent an outpatient colonoscopy. For the last 3 months, she had complained of sudden vomiting, alternating bowel habits, and weight loss of about 7 kg. Blood examinations revealed only slight normocytic anemia (Hb 11.8 gr%, MCV 86) with minimal vitamin B12 levels (188). The fecal test showed unremarkable findings, while fecal occult blood testing was positive. During colonoscopy, a long, yellow, flat worm-like structure was observed in the right colon, arising from the ileo-cecal valve (Fig. 6.12). The worm was grasped by a biopsy forceps and, by following it, the colonoscope was pushed into the terminal ileum, where the worm was grossly encircled. No mucosal damage was seen either in the ileum, or in the colon (Fig. 6.13). The grasped worm was almost completely retrived from the channel of the colonoscope (Fig. 6.14). Subsequent examination showed that it contained several tighly-connected segments (proglottids). Histological examination of the specimen confirmed it to be *Taenia saginata*. The patient was treated with four tablets of Yomesan in a single dose, which allowed her complete recovery. She had no further problems during follow-up of more than 3 years.

Open Issues

Taenia saginata, the beef tapeworm, is most frequently detected in the small bowel, with the head usually residing in the jejunum or ileum. Each segment, known as a proglottid, has a complete set of reproductive organs [4]. High prevalence areas of *Taenia* are sub-Saharan African, southeast Asia, and the Middle East and it can affect both humans and cattle, especially due to the consumption of raw beef or where sanitation conditions are poor. Tapeworm infection can be asymptomatic in humans, except in the case of *Taenia*

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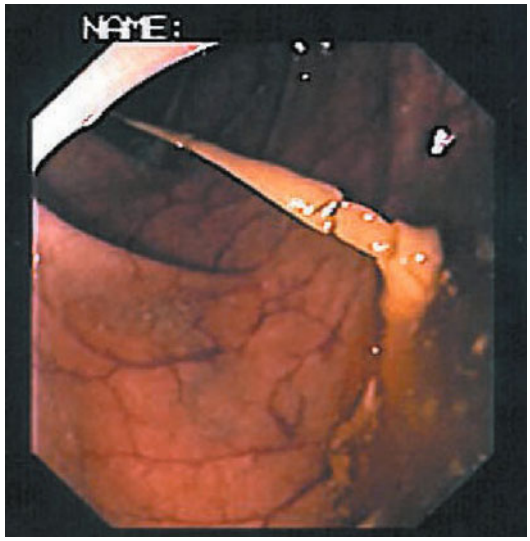

Fig. 6.12 Endoscopic view of colonic Tenia, with a close-up view showing its morphology

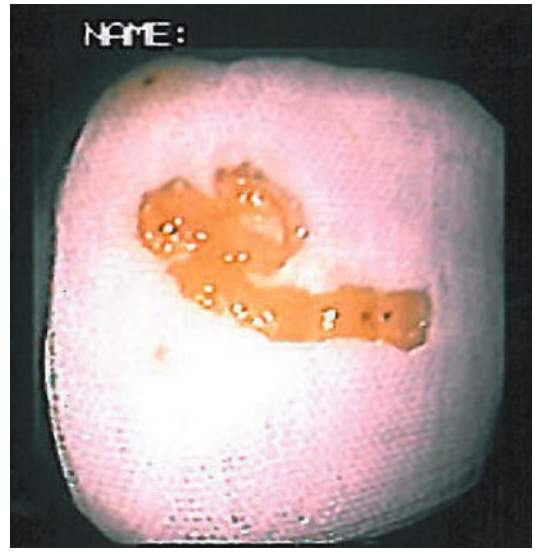

Fig. 6.14 A fragment of the retrieved Tenia, with a close-up view showing its morphology

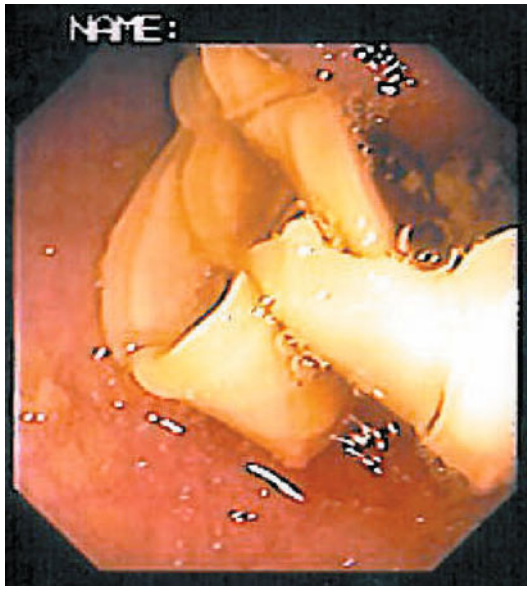

Fig. 6.13 Endoscopic view of the encircled Tenia in the distal ileum

in which the eggs or proglottids are sometimes visible in the stool.

Our young patient was not aware of the infection. She came to our attention complaining vomiting and had suffered significant weight loss. Obstructive diseases due to tapeworm infection have been rarely described. Typical symptoms are weight loss, B12 deficiency, and megaloblastic anemia reflecting competition of the worm with the human host for vitamin B12 absorption. This is particularly severe with *Diphyllobothrium* species. The juvenile form of *Taenia solium*, the pork worm, can provoke cysticercosis in the brain, associated with significant morbidity [5]. Medical treatment includes Praziquantel, a synthetic heterocyclic isoquinolone-pyrazine derivative that induces ultrastructural changes with increased permeability to calcium ions, inducing a paralysis of the worm, which is finally eliminated via peristalsis. Medical treatment is always recommended even in case of endoscopic removal of the parasite. This was the approach to our patient, in whom no regrowth of the worm was evident after 3 years of follow-up.

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Multiple Choice Questionnaire

1) Which are the high prevalence areas of *Taenia* infection?

a. Europe
b. North America
c. Sub-Saharan African
d. China

2) In the human body, where is *Taenia saginata* most commonly present?

a. right colon
b. sigmoid colon
c. rectum
d. small bowel

3) Which tapeworm can induce cysticercosis in the brain?

a. *taenia saginata*
b. *taenia solium*
c. *ascaridia*
d. *diphyllobothrium*

1.c – 2.d – 3.b

Case 7
Jejunal Adenocarcinoma in a 16-Year-Old Patient: An Unusual Case

Filippo Torroni, Erminia Romeo, Paola De Angelis, Francesca Foschia, Paola Francalanci, Tamara Caldaro, Francesca Rea, Giovanni Federici di Abriola, Alessandro Inserra and Luigi Dall'Oglio

Background

Small bowel tumors (SBT) are rare. Among primary small bowel malignancies, adenocarcinoma is the most common histological subtype. Patients at risk for SBT include those with Crohn's disease, familial adenomatous polyposis (FAP), celiac sprue, and peptic ulcer disease [1, 2]. In the following we present an unusual case of jejunal adenocarcinoma in a 16-year-old boy.

Clinical Presentation

A 16-year-old boy was brought to the emergency room for severe anemia, vomiting, and weight loss for 2 months. Inflammatory bowel disease (IBD) was clinically suspected despite the negative inflammatory blood values. A family history of IBD or SBT was negative; laboratory parameters showed anemia (hemoglobin 7,5 g/dl). No condition necessitating surgery was detected by the physical examination. A scintigraphy excluded Meckel's diverticulum; an upper and a lower endoscopy with histology were normal; however, wireless capsule endoscopy (WCE) showed jejunal lesions suggestive of Crohn's disease (Fig. 7.1). The capsule was never evacuated. Therefore, 11 days later, a single-balloon enteroscopy with biopsies was performed, which identified a jejunal mass with capsule retention (Figs. 7.2, 7.3). Adenocarcinoma was diagnosed based on the biopsies. The total body CT scan was negative for metastasis. The patient underwent surgical resection of 30 cm of small bowel (jejunum) (Fig. 7.4). The diagnosis was confirmed by the histology of the surgical specimen (Fig. 7.5) and the patient received chemotherapy. An endoscopic follow-up was performed 8 months after surgery. A normal mucosa was seen at the level of the anastomosis (Fig. 7.6).

Open Issues

The recently development of deep enteroscopy has changed the diagnostic and therapeutic approach to small bowel diseases. Double-balloon enteroscopy, to study the small bowel, was introduced at the beginning of 2001 by Yamamoto. This technique is available for routine clinical use in both the adult and the pediatric population, just as with single-balloon enteroscopy.

The indications for double-balloon enteroscopy include obscure gastrointestinal bleed-

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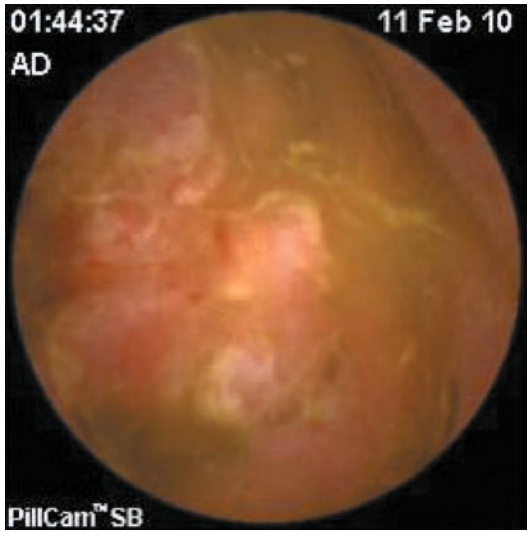

Fig. 7.1 Wireless capsule endoscopy: jejunal findings

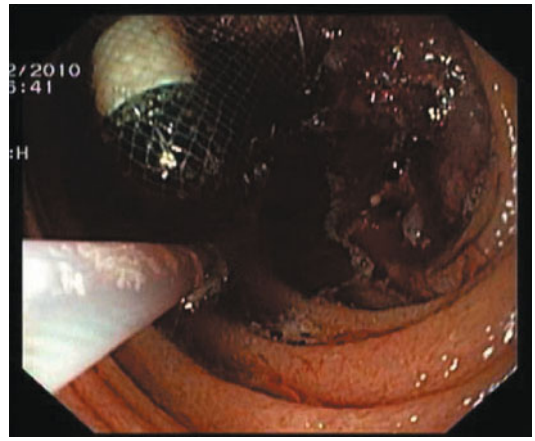

Fig. 7.2 Single-balloon enteroscopy: capsule retention

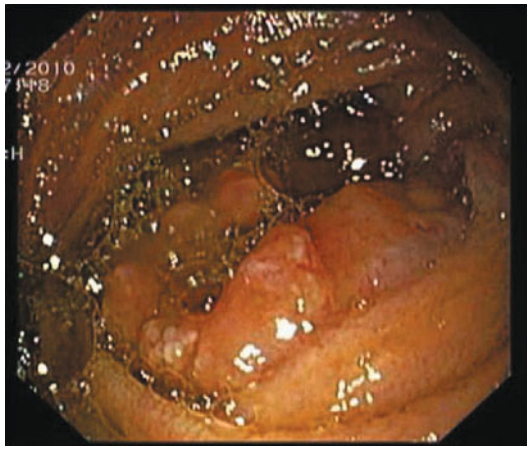

Fig. 7.3 Single-balloon enteroscopy: jejunal mass

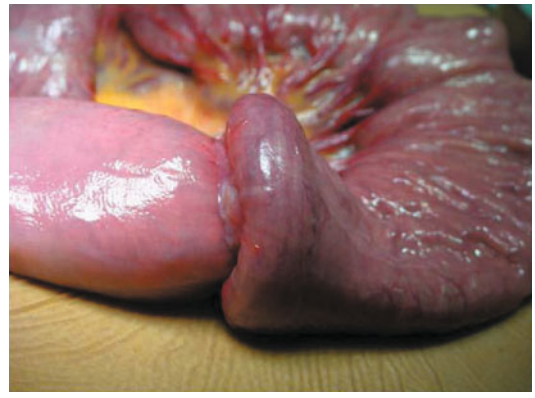

Fig. 7.4 Surgical findings of a small bowel tumor

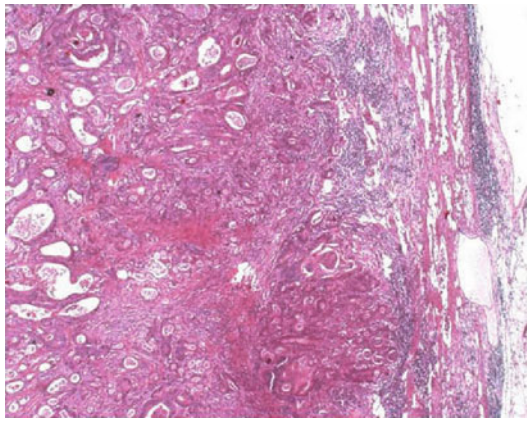

Fig. 7.5 Histology the of surgical specimen: jejunal adenocarcinoma

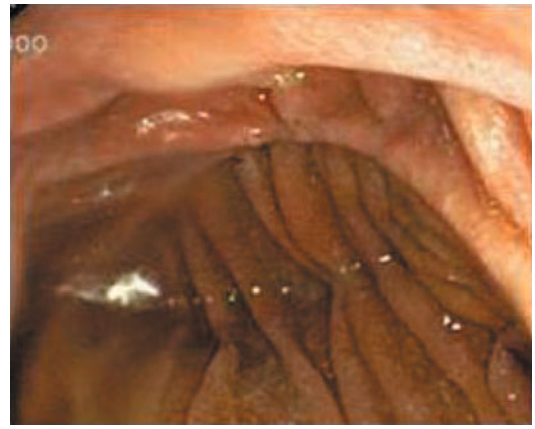

Fig. 7.6 Enteroscopy: post-operative follow-up

ing, Crohn's disease, intestinal polyposis, and suspected intestinal tumors [3, 4].

In our case, the combination of WCE and single-balloon enteroscopy was essential to diagnose SBT, especially considering that the risk factors and family history provided no indication of the possibility of tumor in this young patient.

We want to point out also that capsule imaging must be evaluated with attention paid to avoiding misleading interpretations of the frame.

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Multiple Choice Questionnaire

1) What is the minimum age for performing capsule endocopy in children?

a. 3 years
b. < 10 years
c. 7 years
d. < 3 years
e. 1 year

2) Which is the main WCE indication?

a. recurrent abdominal pain
b. inflammatory bowel disease
c. obscure gastrointestinal bleeding (OGIB)
d. polyposis
e. celiac disease

3) Which is the timing of WCE in OGIB?

a. after 72 hours
b. at least 1 week
c. in 24-48 hours
d. no specific timing
e. during bleeding

1.d – 2.c – 3.c

Radiology

Case 8.1
A Collection of Clinical Cases

Laura Maria Minordi, Maria Gabriella Brizi, Amorino Vecchioli, Alessandra Farchione, Luigi Larosa, Rosa Marra and Lorenzo Bonomo

Case 1

A 24-year-old man with a recent onset of intermittent diarrhea, mild diffuse abdominal pain, as well as weight loss.

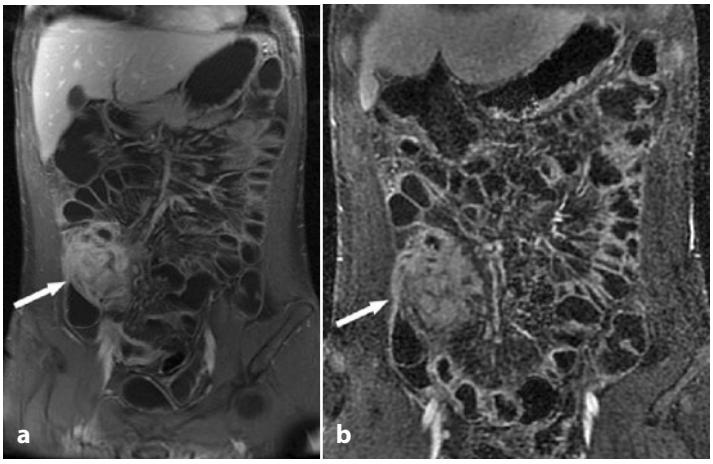

Fig. 8.1 Enterography MR, performed after oral ingestion of polyethylene glycol solution (2L), shows in coronal fat suppressed T1 weighted fast spin echo (FS-FSE) sequence (**a**) marked parietal thickening of the last ileal loop with luminal stenosis and altered signal intensity of the perivisceral mesentery (*arrow*). Coronal contrast enhanced fat-suppressed T1-weighted FSE sequence (**b**) shows intense parietal enhancement of the last ileal loop (*arrow*), referred to an active inflammatory bowel disease (Crohn's disease)

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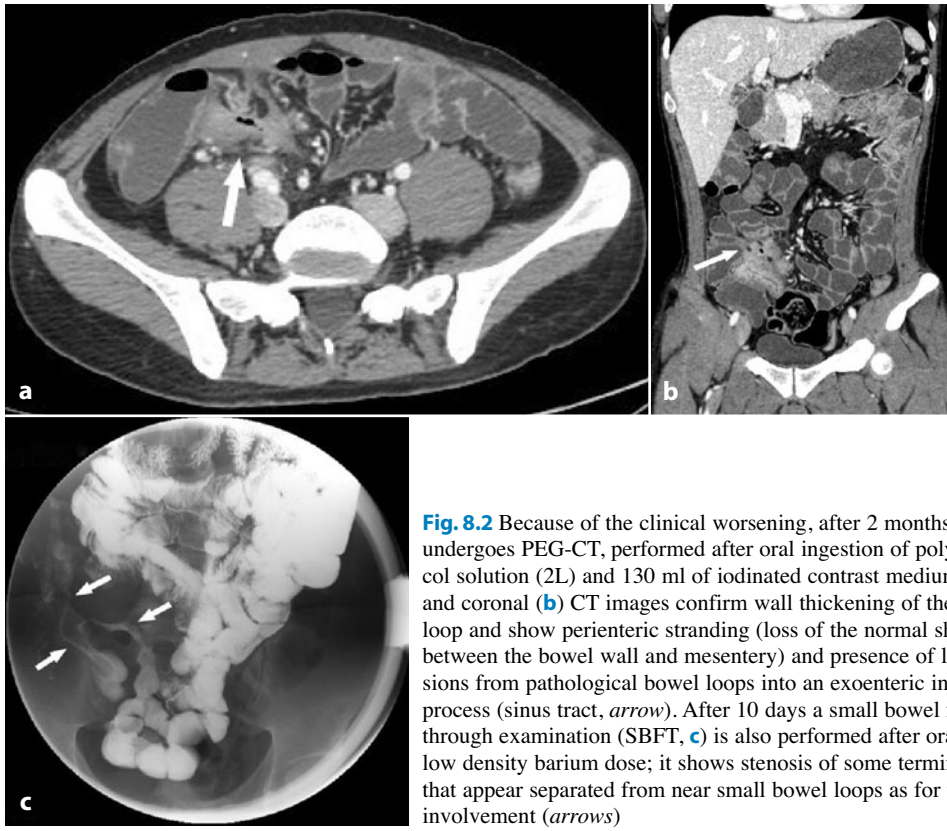

Fig. 8.2 Because of the clinical worsening, after 2 months the patient undergoes PEG-CT, performed after oral ingestion of polyethylene glycol solution (2L) and 130 ml of iodinated contrast medium; axial (**a**) and coronal (**b**) CT images confirm wall thickening of the last ileal loop and show perienteric stranding (loss of the normal sharp interface between the bowel wall and mesentery) and presence of linear extensions from pathological bowel loops into an exoenteric inflammatory process (sinus tract, *arrow*). After 10 days a small bowel follow-through examination (SBFT, **c**) is also performed after oral ingestion of low density barium dose; it shows stenosis of some terminal ileal loops that appear separated from near small bowel loops as for the mesentery involvement (*arrows*)

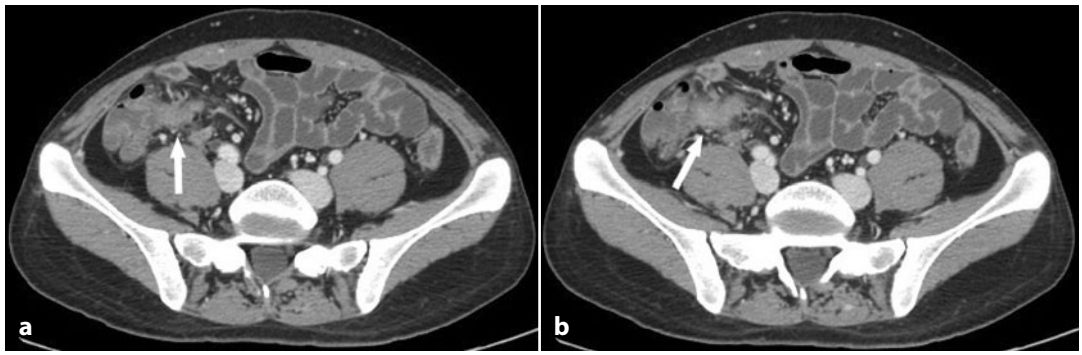

Fig. 8.3 After antibiotic therapy (one month) PEG-CT is performed again and shows reduction of the parietal thickening of the last ileal loop (**a**, *arrow*) and the involvement of the perivisceral mesentery (**b**, *arrow*).

Case 2

A 62-year-old man with a history (7 years) of ileal Crohn's disease.

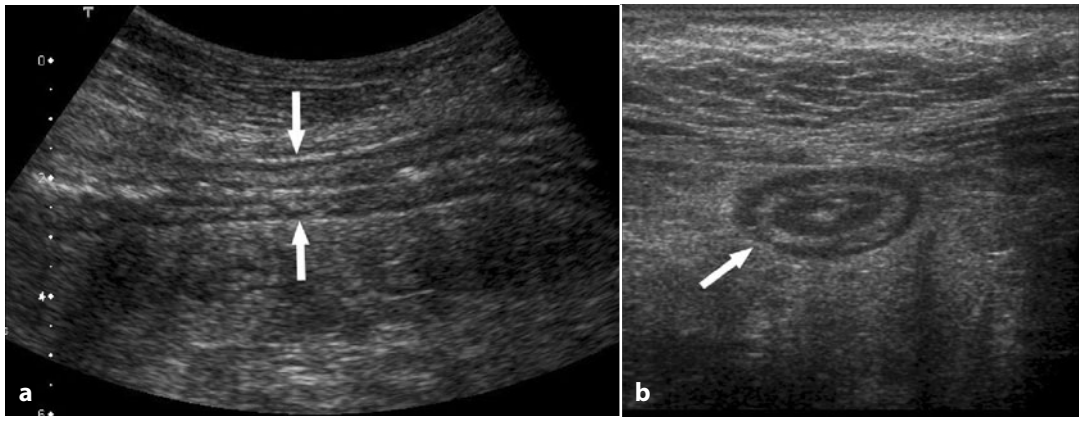

Fig. 8.4 Because of the clinical worsening (abdominal pain in right iliac fossa, weight loss and asthenia), the patient undergoes an abdominal ultrasonography (US), at low (**a**) and high (**b**) frequencies; bowel wall thickening of the terminal ileum, a prominent submucosal layer, a narrowed lumen, and mesenteric fat hypertrophy are evident (*arrows*)

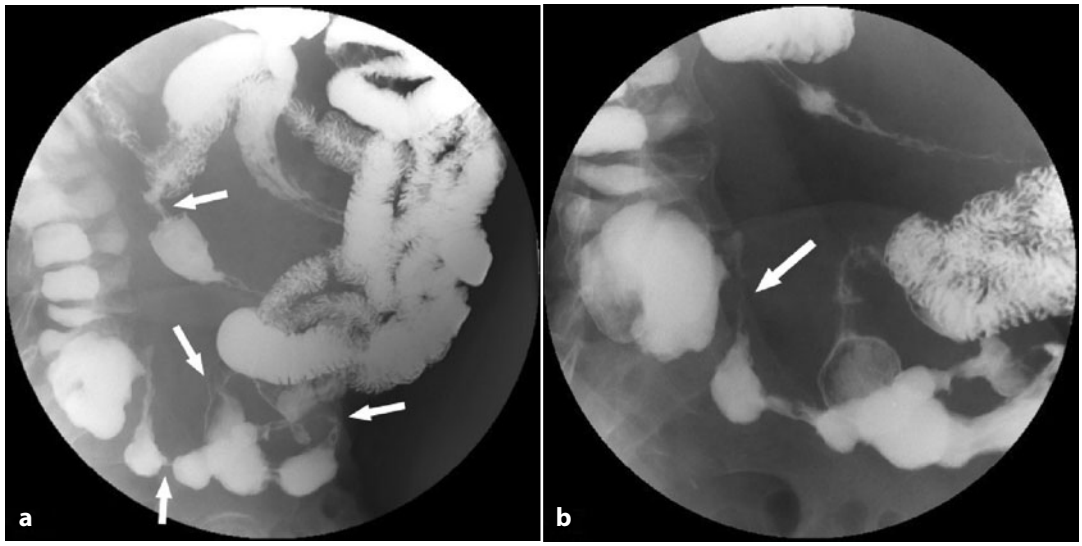

Fig. 8.5 Small bowel follow-through examination (SBFT) shows linear deformities and loss of the normal fold patterns of the ileal loops due to the presence of alternating stenotic (*arrows*) and normal tracts (**a**). The last ileal loop is also involved (**b**, *arrow*)

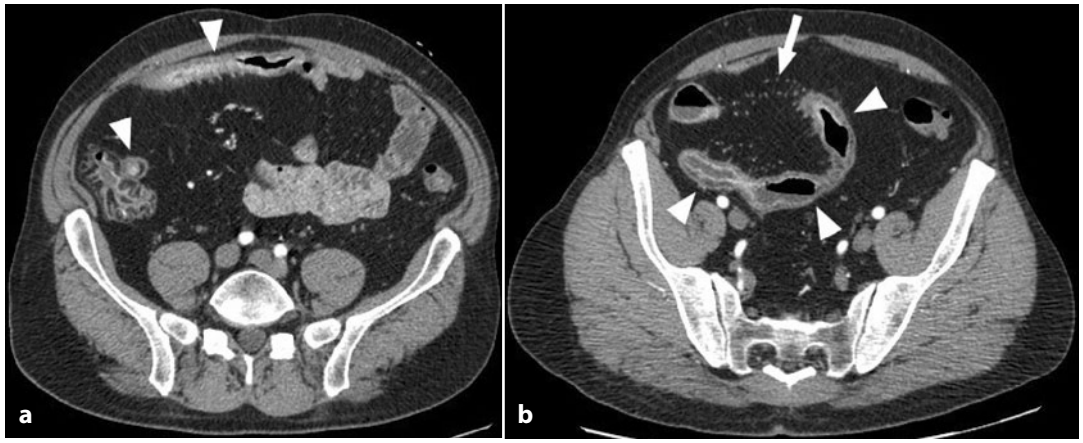

Fig. 8.6 Contrast-enhanced CT demonstrates mural hyperenhancement, mural stratification, and wall thickening of multiple ileal loops (**a** and **b**, *arrowheads*), in association with hypervascularity of the involved mesentery (*comb sign*, *arrow* in **b**).

Case 3

An 83-year-old man with melena, diffuse abdominal pain, and weight loss of abrupt onset.

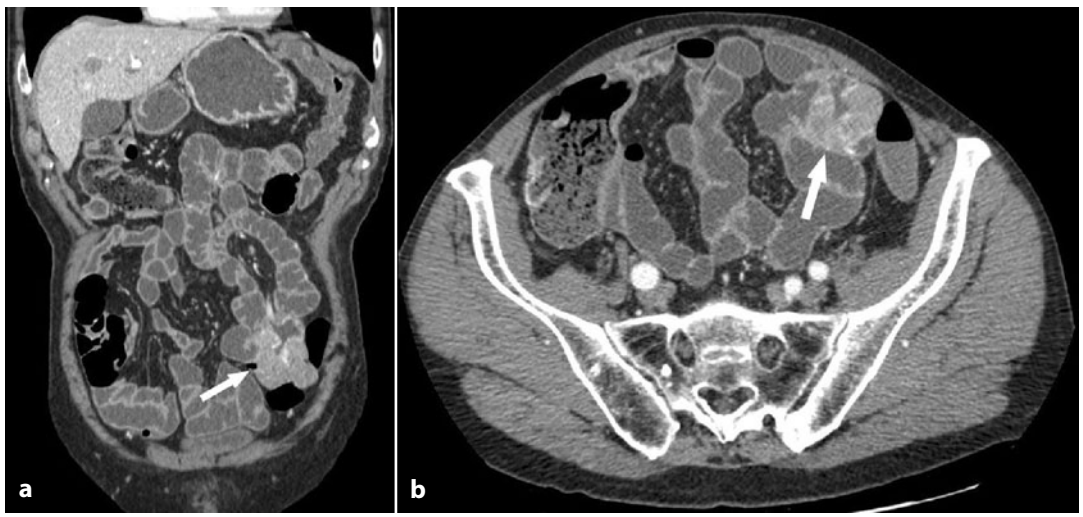

Fig. 8.7 Abdominal enterography-CT, coronal (**a**) and axial (**b**) images, show marked asymmetric wall thickening of an iliac loop (*arrow*), localized in the left iliac fossa, associated with a reduction of lumen caliber. This radiological aspect suggested a small bowel tumor, confirmed by surgery (surgical report: gastrointestinal stromal tumor)

Case 4

A 33-year-old man with flushing and rectal bleeding

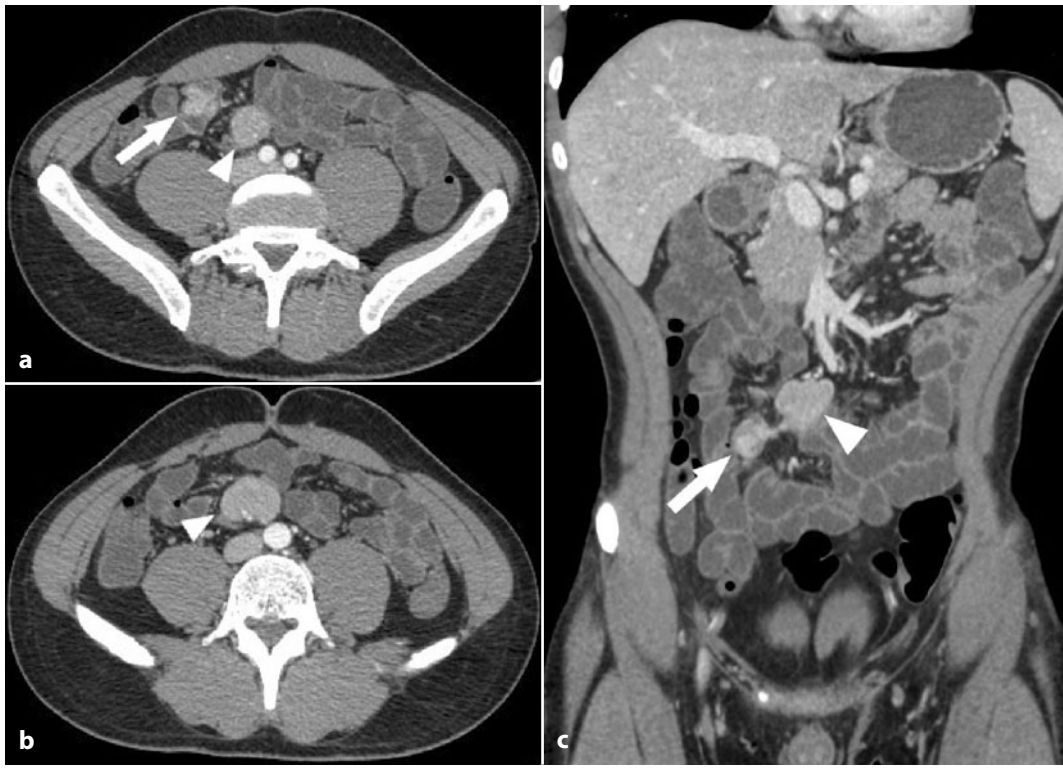

Fig. 8.8 PEG-CT, performed after oral ingestion of polyethylene glycol solution (2L) and 130 ml of iodinated contrast medium. Axial (**a, b**) and coronal (**c**) images, in the arterial phase, show an ileal polypoid anular lesion (*arrows*) with early contrast enhancement; regional metastatic lymphadenopathy is also seen (*arrowheads*). This radiological aspect suggested a small bowel tumor, confirmed by surgery (neuroendocrine gastrointestinal tumor)

Case 5

A 49-year-old man with recent onset of right abdominal pain and a positive history of raw fish consumption.

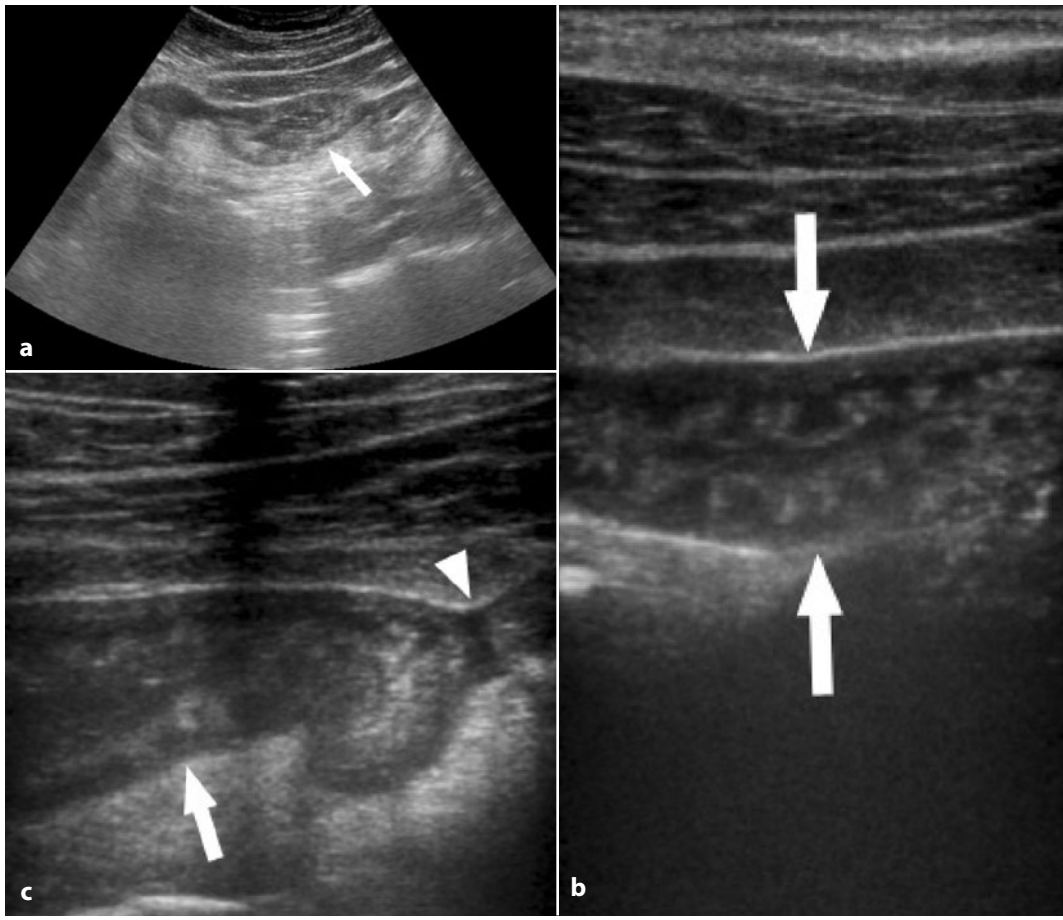

Fig. 8.9 Abdominal ultrasonography, at low (**a**) and high frequencies (**b, c**) demonstrates stratified wall thickening of the terminal ileum (*arrows*) and a small amount of ascitic fluid beside the affected bowel (*arrowhead*)

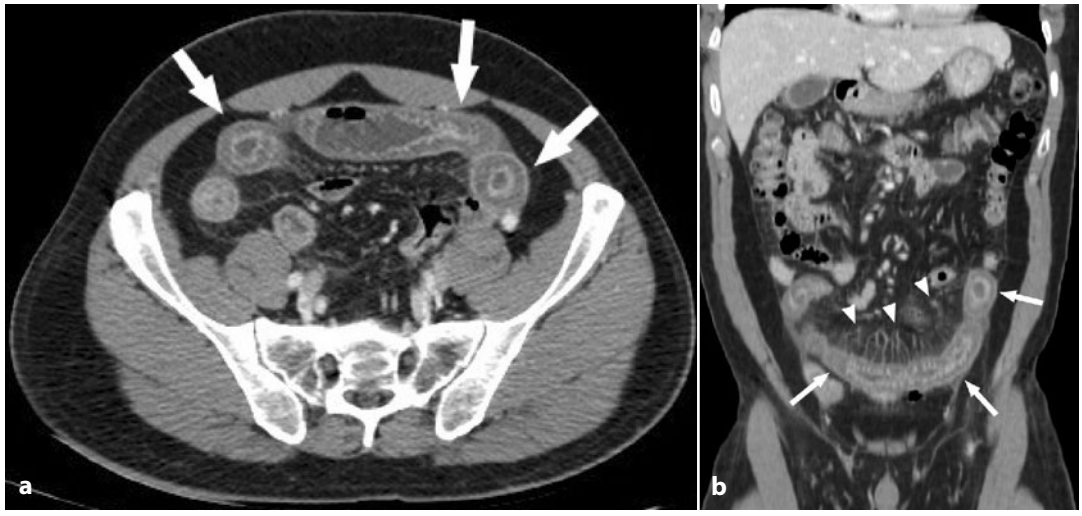

Fig. 8.10 Axial (**a**) and coronal (**b**) contrast-enhanced CT images show wall thickening of a long section of the ileum with alternating layers of high and low attenuation ("target sign", *arrows*), in association with hyperdensity and hypervascularity of the involved mesentery (*arrowheads*)

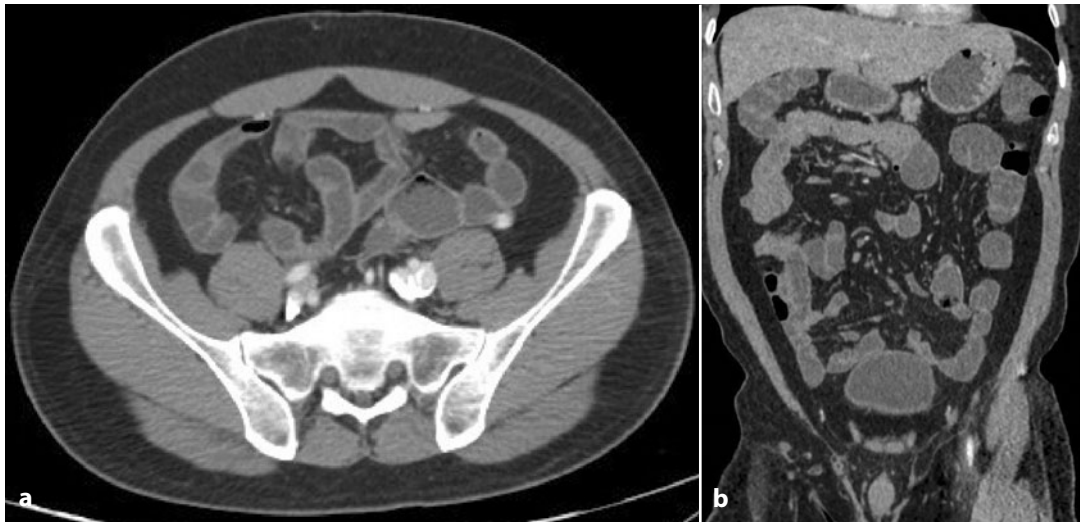

Fig. 8.11 a, b Because of suspected inflammatory bowel disease, after seven days the patient undergoes PEG-CT, performed after oral ingestion of polyethylene glycol solution (2L) and 130 ml of iodinated contrast medium; at the same levels the exam demonstrates complete resolution of the previous alterations. Parasitological examination of stool shows Anisakis parasitic infestation of the ileum

Multiple Choice Questionnaire

1) In CT enterography we use

a. only a IV iodinated contrast medium; no oral contrast medium for small loop distension
b. only a contrast medium for small loop distension; no IV iodinated contrast medium
c. no IV iodinated contrast medium; no oral contrast medium for small loop distension
d. sometimes IV iodinated contrast medium and oral contrast medium for small loop distension
e. always IV iodinated contrast medium and oral contrast medium for small loop distension

2) If we use a naso-jejunal tube for the distension of the small bowel loop in CT or in MRI (enteroclysis) instead of the oral administration of contrast medium (enterography)

a. in CT/MRI enteroclysis the distension is better both in the jejunum and in the ileum than achieved with the CT/MRI enterography
b. the distension is better only for jejunum in CT/MRI enteroclysis than achieved with the CT/MRI enterography
c. the distension is better only for the ileum in CT/MRI enteroclysis than achieved with the CT/MRI enterography
d. no difference is present between the two techniques
e. the distension is inferior in CT/MRI enteroclysis compared to the CT/MRI enterography

3) In Crohn's disease the differentiation between edematous stenosis and fibrotic stenosis

a. is not possible with both CT or MRI
b. is possible only in CT; fibrotic stenosis shows reduced contrast enhancement and inferior parietal thickening compared to edematous stenosis
c. is possible only in MRI; fibrotic stenosis shows reduced contrast enhancement and inferior parietal thickening compared to edematous stenosis
d. is possible with both CT and MRI; fibrotic stenosis shows increased contrast enhancement and superior parietal thickening compared to edematous stenosis
e. is possible with both CT and MRI; stenotic stenosis shows reduced contrast enhancement and inferior parietal thickening compared to edematous stenosis

4) The hallmark of small bowel disease is

a. always homogenous mural thickening in benign disease and inhomogeneous mural thickening in malignant disease
b. always homogenous mural thickening in malignant disease and inhomogeneous mural thickening in benign disease
c. usually homogenous mural thickening in benign disease and inhomogeneous mural thickening in malignant disease
d. usually homogenous mural thickening in malign disease and inhomogeneous mural thickening in benign disease
e. no difference between mural thickening between benign or malignant disease

1.e – 2.a – 3.e – 4.c

Case 8.2
Adenocarcinoma of the Small Bowel in a 48-Year-Old Male: Radiological-Surgical Correlation

Franco Iafrate, Marcella Iannitti, Paolo Baldassari, Davide Diacinti and Andrea Laghi

Background

According to the literature, small bowel tumors (SBTs) account for 2–3% of all gastrointestinal malignancies. Adenocarcinoma is the most frequent histological type, followed by carcinoid tumor, lymphoma, and gastrointestinal stromal tumor (GIST) [1]. The development of SBTs shows an increasing incidence in patients with inflammatory bowel diseases such as Crohn's disease and celiac disease [2, 3]. Clinical symptoms are usually nonspecific and include abdominal pain, anorexia, weight loss, bowel obstruction, jaundice, perforation, or bleeding. Over the last few decades, radiological diagnostic tools for the early detection of SBTs has significantly improved. Surgery represents the treatment of choice, but diagnosis is often reached late. Currently, 5-year survival is only 15–30%. [4]

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Clinical Presentation

A 48-year-old man with a recent diagnosis of celiac disease was placed on a gluten-free diet, which he followed for 6 months. He was admitted to the hospital with nausea, vomiting, and increasing autoantibodies for celiac disease. An ultrasound study of the small bowel revealed non-specific thickening of the small bowel wall. The patient underwent MR enterography in order to assess the severity of the celiac disease. He was administered an oral positive contrast agent (PEG 4000) in 1.5 L of water 45 min prior to the study, in order to distend the small bowel. The MRI protocol consisted of TSE single shot T2-weighted sequences, true-FISP imaging, diffusion weighted imaging, and GR echo T1-weighted sequences before and after intravenous administration of gadolinium. The exam revealed an endoluminal stenosing lesion within a distal ileal loop, with consequent marked dilatation of the proximal bowel loops (Fig. 8.12). Local lymphadenopathies were observed within peritoneal fat tissue. Ileal "jejunalization" was detected as well, a typical aspect of celiac disease. In order to stage the ileal neoplasm, the patient underwent multidetector CT study with intravenous administration of non-ionic iodinated contrast agent. Both arterial and venous phase images were acquired. Oral administration of hypodense contrast medium

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was avoided because of the marked dilatation of the small bowel loops due to a sub-occlusion condition. The CT study clearly demonstrated an eccentric thickening of the ileal wall, with a diameter of 3 cm. Along with the resulting marked fluid distention of the proximal small bowel, gas-fluid levels were present. After the patient was injected with an

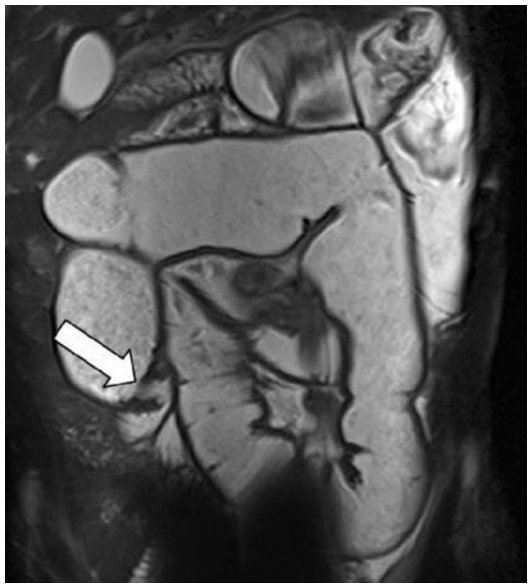

Fig. 8.12 MR enterography. Coronal TSE single shot T2-weighted sequence shows the endoluminal lesion (*arrow*) within a distal ileal loop, determining a stenosis with consequent marked dilatation of the proximal bowel loops

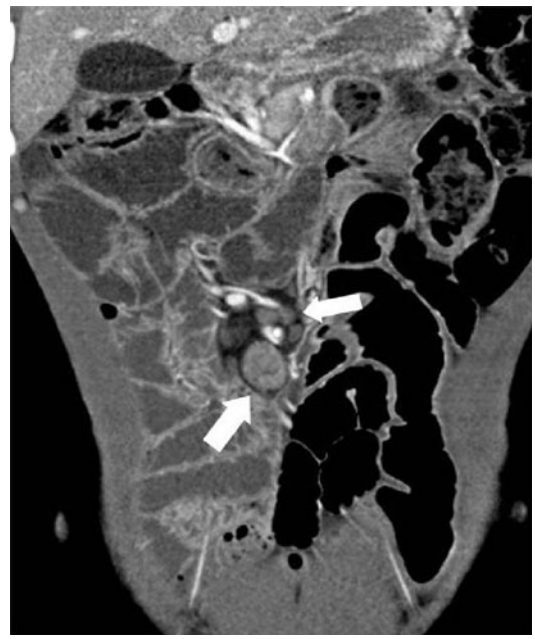

Fig. 8.14 MDCT coronal reformatted image shows pathological lymphadenopathies along the course of the ileal vessels (*arrows*)

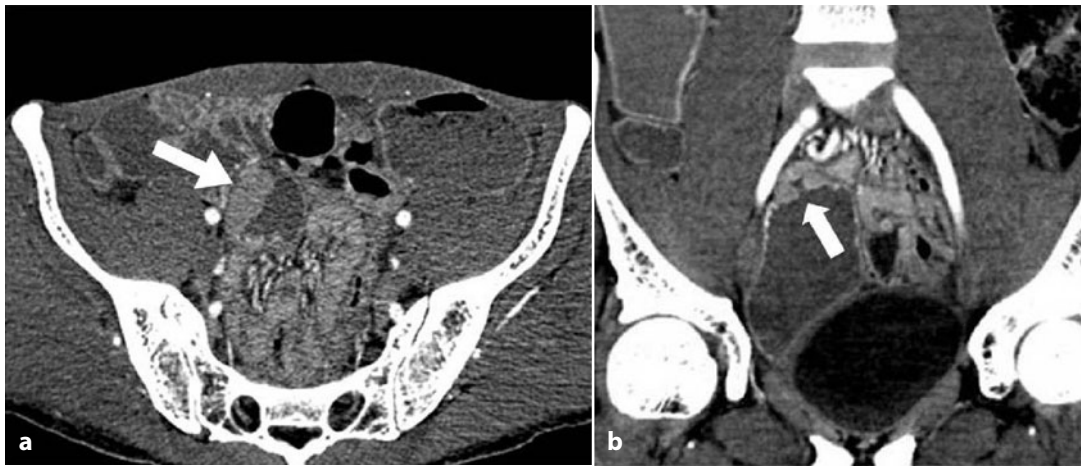

Fig. 8.13 Axial (**a**) and coronal (**b**) MDCT enterography confirms an eccentric wall thickening of the small bowel at the level of the distal ileum. There is marked enhancement after intravenous contrast administration (*arrow*)

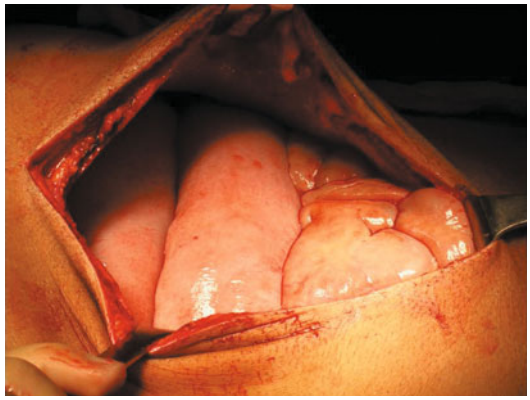

Fig. 8.15 Surgical opening of the abdomen confirms the marked distention of the small bowel loop proximal to the lesion

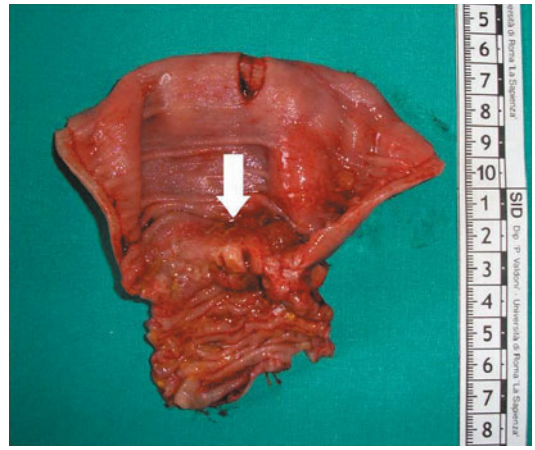

Fig. 8.16 The surgical specimen highlights the eccentric lesion, of about 3 cm in diameter and involving the wall of the distal ileum (*arrow*)

intravenous contrast agent, the lesion showed moderate and non-homogenous enhancement. Local lymphadenopathies with a frankly pathologic aspect were confirmed (Figs. 8.13–8.15) but other, metastatic lesions were not detected. Treatment consisted of surgical ileal resection with associated lymphadenectomy (Fig. 8.16). Histological analysis revealed that the neoplasm was a rare small bowel adenocarcinoma of the ileum [5].

Open Issues

Both MRI and MDCT play a key role in the detection of SBTs. In the literature, the sensitivity and specificity of CT and MR enterography in the detection of tumors of the small bowel are quite variable, with some authors reporting rates of close to 90% [6]. In our experience, SBTs can be misdiagnosed at MR enterography, thus recommending MDCT as the preferred imaging modality in order to obtain an accurate pre-surgical evaluation. The advantages of MDCT are its higher spatial resolution, faster examination time, and the absence of artifacts related to peristaltic motion and breathing.

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Multiple Choice Questionnaire

1) Which signs are typical for adenocarcinoma of the small bowel?

a. homogeneously enhancing bowel wall
b. enlargement of the mesenteric lymph nodes
c. "comb sign"
d. "cobblestone appearance"
e. short thickened segment causing an ileal stenosis that enhances with intravenously administered contrast agent

2) Which is the most common histological type of malignant SBT?

a. lymphoma
b. carcinoid tumor
c. gastrointestinal stromal tumor (GIST)
d. adenocarcinoma
e. lipoma

3) Which are the the advantages of MDCT?

a. high spatial resolution
b. fast examination time
c. the absence of artifacts related to peristaltic motion and breathing
d. accurate preoperative TNM staging
e. all of the above

1.e – 2.d – 3.e

Case 8.3
A 34-Year-Old Woman with Chronic Inflammation Due to Crohn's Disease: Typical MRI Signs

Franco Iafrate, Maria Ciolina, Alessandro Pichi and Andrea Laghi

Background

Crohn's disease is a chronic granulomatous inflammatory disease of the gastrointestinal tract. According to epidemiological studies, the pathology is not uncommon, with 400,000–600,000 people affected in North America alone [1]. The disease occurs in males and females with the same prevalence; the peak age range of disease development is 15–25 years. The etiology is multifactorial but a familial tendency has been described, with an increased risk in first-degree relatives [1-3]. While many new clinical and surgical therapeutic strategies have been recently introduced, their success depends on an accurate assessment of the extension and severity of the disease.

Clinical Presentation

A 34-year-old woman with Crohn's disease treated with immunosuppressive agents for about 5 years presented with symptom exacerbation, characterized by severe abdominal pain, rectal bleeding, and diarrhea alternating with bowel subileus.

The patient was referred to our center for MR enterography in order to assess the severity of her disease. Patient preparation for the

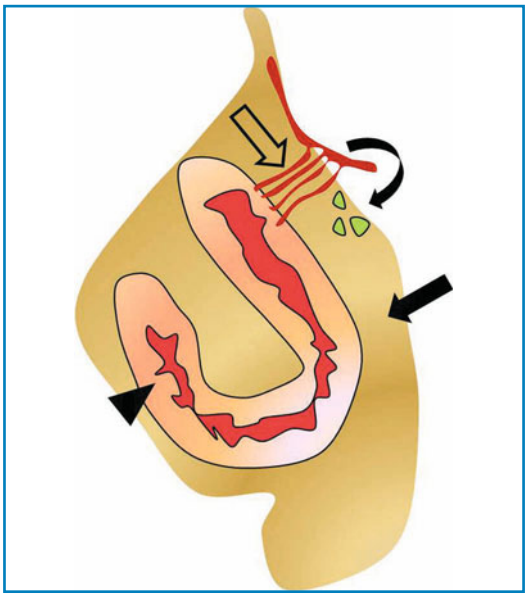

Fig. 8.17 Drawing of a pathologic small bowel, showing a pathologically thickened segment of the distal ileum (*arrowhead*) associated with fibrofatty proliferation (*arrow*), the "comb sign" due to vascular engorgement (*empty arrow*), and local reactive lymphadenopathies (*curved arrow*)

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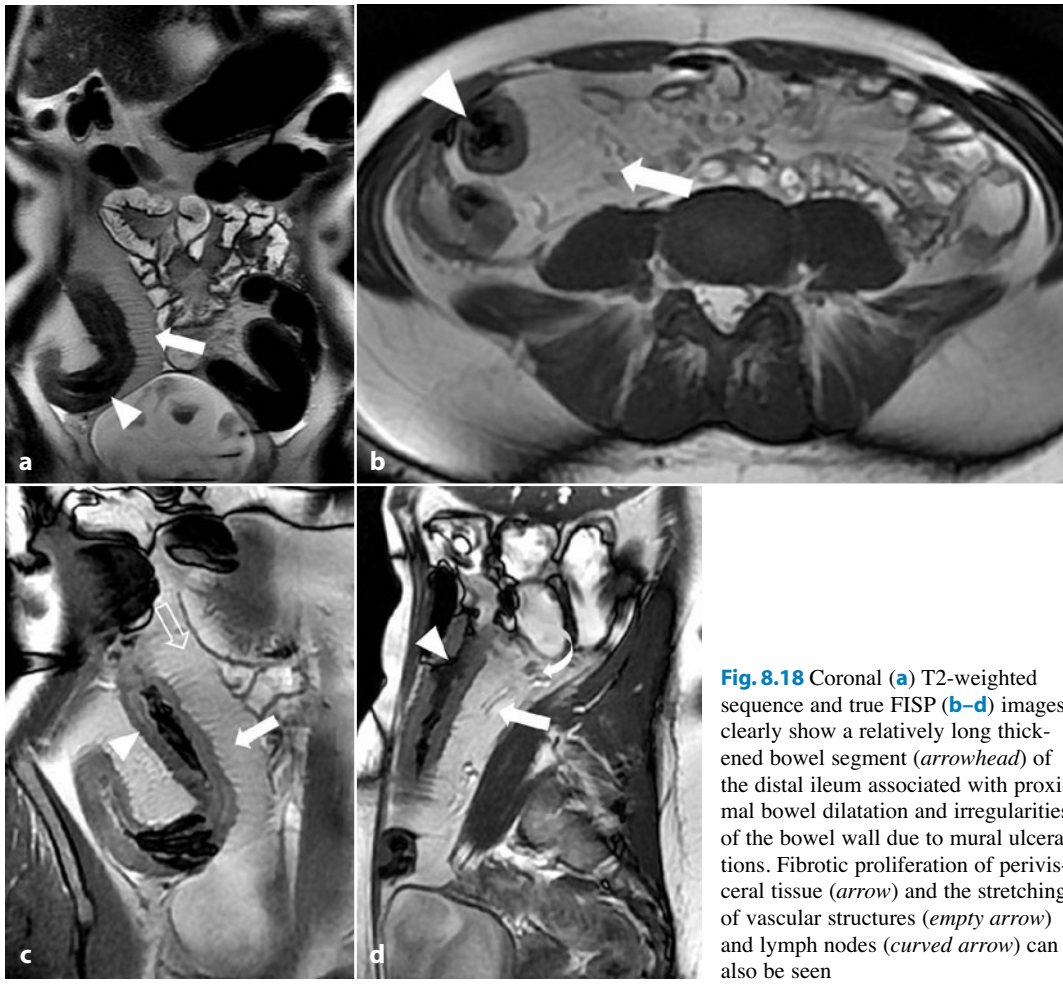

Fig. 8.18 Coronal (**a**) T2-weighted sequence and true FISP (**b–d**) images clearly show a relatively long thickened bowel segment (*arrowhead*) of the distal ileum associated with proximal bowel dilatation and irregularities of the bowel wall due to mural ulcerations. Fibrotic proliferation of perivisceral tissue (*arrow*) and the stretching of vascular structures (*empty arrow*) and lymph nodes (*curved arrow*) can also be seen

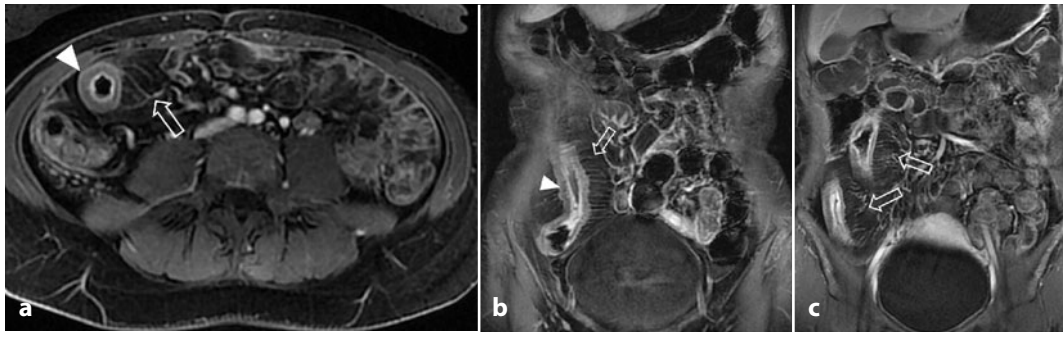

Fig. 8.19 Axial (**a**) and coronal (**b, c**) T1-weighted sequences show the stratified pattern of the hyper-enhancing mucosa and hypo-enhancing submucosa (*arrowhead*). Congestion of the mesenteric vessels, producing the "comb sign" (*empty arrow*), is evident around the involved segment. These findings are strongly suggestive of the active lesions of Crohn's disease

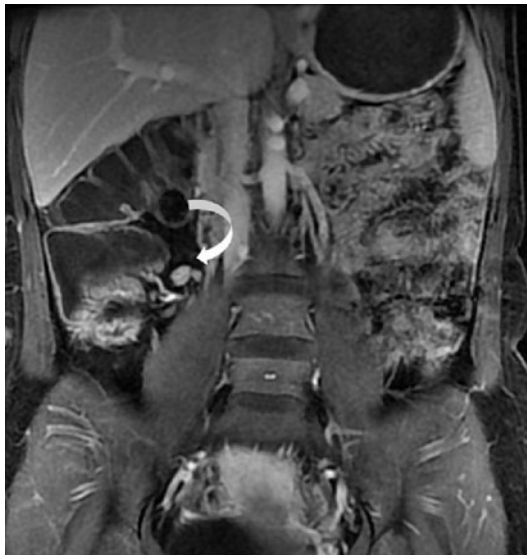

Fig. 8.20 Coronal T1-weighted image shows the enlargement of mesenteric reactive lymph nodes (*curved arrow*)

exam consisted of fasting for 6 h, oral administration of positive contrast agent (PEG 4000) in 1.5 L of water 45 min prior to the study, and the intravenous administration of a spasmolytic agent (hyoscine butylbromide). MRI was carried out using TSE T2 weighted single shot sequences, true FISP imaging, and GR fat-suppressed T1 weighted sequences, with the intravenous administration of a paramagnetic contrast agent. Axial, coronal, and multiplanar images according to the orientation of the affected bowel segment were acquired. T2-weighted sequences demonstrated mural thickening of more than 5 mm involving the distal ileum, with an extension in length of 20 cm. Multiple mucosal irregularities due to the presence of mural ulcerations and fissures alternated with superficial pseudo-nodules, resulting in a "cobblestone appearance" [4, 5].

On GR T1-weighted contrast-enhanced sequences, the pathologic segment showed a stratified pattern of enhancement with a "target" or "double halo" appearance, related to the hyperintensity of the inflamed mucosal layer and the hypointensity of the outer layer due to fibrotic involution of the submucosa and muscularis propria. Contrast-enhanced sequences revealed the vascular engorgement of the vasa recta, in which the alternating tubular and straight pattern referred to as the "comb sign," indicative of active inflammation, was seen. Extraintestinal changes detectable in every sequence were the presence of mesenteric reactive lymph adenopathy and fibrotic proliferation of the perivisceral mesenteric fat, i.e., "fibro-fatty proliferation," due to the chronic inflammation, with a loss of the normal interface between the bowel wall and mesentery. True FISP imaging demonstrated the absence of residual peristaltic activity, again due to the fibrotic changes in the muscle layer as a direct consequence of chronic inflammation [4, 5].

Open Issues

The reported overall sensitivity of MR enterography in the evaluation of Crohn's disease is 96–100% according to a per patient analysis, and over 85% in a per segment analysis [6-8]. To achieve this high sensitivity, bowel distention is crucial regardless of the type of oral contrast medium (positive, negative, or biphasic). We prefer to administer positive oral contrast diluted in 1.5 L of water. Generally, we use PEG 4000 or mannitol with a concentration not higher than 2–2.5%, in order to avoid side effects such as diarrhea, excess intestinal gas, and abdominal cramps [9, 10].

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Multiple Choice Questionnaire

1) Which signs are typical consequences of fibrotic involution due to the chronic inflammation of Crohn's disease?

a. homogeneously enhancing bowel wall
b. enlargement of mesenteric lymph nodes
c. stratified enhancement of the bowel wall with a "target" appearance and fibrofatty proliferation
d. "cobblestone appearance"
e. mural ulcerations

2) Which is the best sequence to rule out the presence of the "comb sign"?

a. TSE T2 weighted single shot sequences
b. TSE T2 weighted sequences with fat-suppression
c. true FISP sequences
d. GR fat-suppressed T1 weighted sequences with the intravenous administration of a paramagnetic contrast agent
e. GR T1 weighted sequences with the intravenous administration of contrast agent and without fat suppression

3) Which are the main advantages of true-FISP sequences?

a. very fast imaging
b. GRE unspoiled sequences benefit from the residual transverse magnetization
c. assessment of the residual peristaltic activity of the pathological bowel segment
d. reduced motion artifacts
e. all of the above

1.c – 2.d – 3.e

Capsule Endoscopy

Case 9.1
Bulging or Mass? This Is the Question

Emanuele Rondonotti, Silvia Paggi, Andrea Anderloni, Nadia Di Lorenzo, Alessandra Baccarin, Luciana Ambrosiani, Giancarlo Spinzi and Roberto de Franchis

Background

Obscure gastrointestinal bleeding (OGIB) is the leading indication for capsule endoscopy (CE), accounting for 70–80% of all procedures performed [1]. The CE diagnostic yield in this setting (50–60%) is superior to that of other diagnostic techniques in the study of the small bowel [2]. This mostly depends on the capability of CE to identify small/flat lesions, often missed by other techniques. On the other hand, protruding lesions (polyps or masses), due to a lack of insufflation, can be difficult to identify; therefore, distinction between masses and bulges still represents a challenging task for CE.

Clinical Presentation

A 60-year-old woman with an unremarkable past medical history was admitted because of an acute episode of melena. She had no abdominal pain, vomiting, or weight loss. However, she had been taking ibuprofen (400 mg/day) for a toothache for the last 5 days. The physical examination was normal. Blood tests revealed iron deficiency anemia (Hb: 8.5 mg/dL; ferritin: 4 ng/mL; MCV: 60 fl). The patient underwent urgent gastroscopy, which showed antral erosions (biopsies failed to detect *Helicobacter pylori*), and subsequently ileocolonoscopy, which was negative. She was therefore referred for CE, performed 22 days later. The CE study showed, in the mid small bowel (at 66% of the small bowel transit time) only in a few frames, a bulge protruding into the lumen, with an ill-defined margin and covered by normal mucosa (Fig. 9.1). Based on

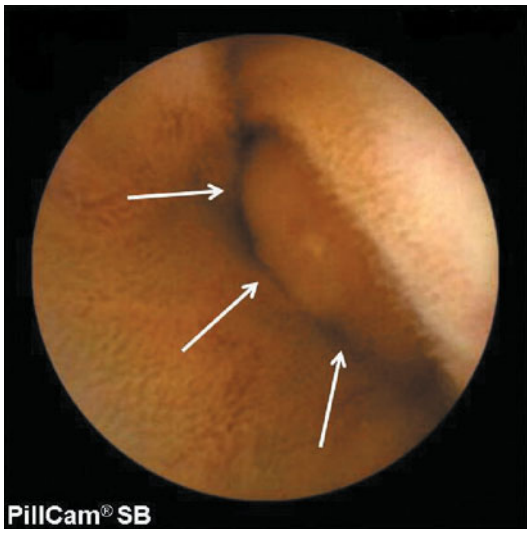

Fig. 9.1 Capsule endoscopy shows a bulge (*white arrows*) with a large base protruding in the lumen and having an ill-defined margin, covered by normal mucosa

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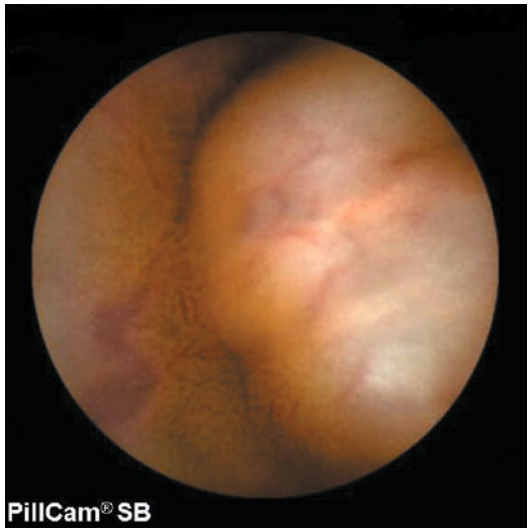

Fig. 9.2 Capsule endoscopy shows a protruding lesion with a well-defined edge, covered by a pale and tense mucosa

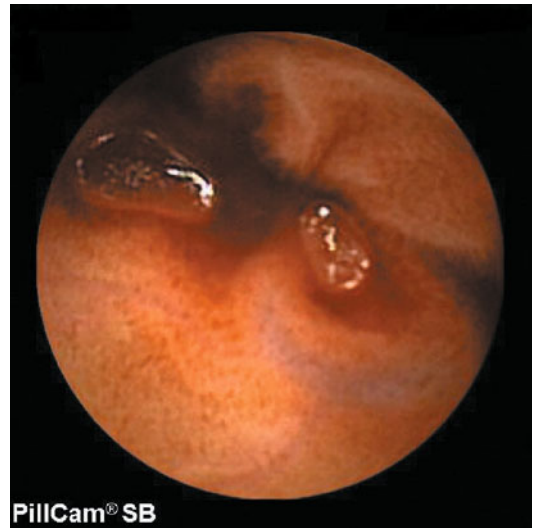

Fig. 9.3 Capsule endoscopy shows red blood in the lumen preventing the evaluation of the small bowel mucosa

the CE appearance alone, the presence of a submucosal mass could not be excluded with certainty; therefore a CT-enterography was planned, which showed no abnormality. A combination of the CE and CT-enterography findings led to classification of the possible lesion identified at CE as an "innocent" bulge. The patient was discharged with iron supplementation and proton pump inhibitors therapy. The blood tests performed 3 and 6 months later showed an increased Hb level (Hb 12.2 g/dl) and the iron supplementation was discontinued. About 6 months later, the patient experienced a new episode of melena; the blood test showed severe anemia (Hb 6.2 g/dl) and she was newly admitted. Immediately after RBC transfusion and urgent gastroscopy, which was negative, the patient swallowed the capsule (48 h after admission). The second CE examination showed in the mid small bowel, roughly in the same area as the previously identified bulge (at about 60% of the small bowel transit time), a protruding lesion with a well-defined edge, covered by pale and tense mucosa (Fig. 9.2). A precise estimate of the size and the shape of the lesion was not possible because it was not fully visible in any single frame. Distally to this lesion, up to the passage through the ileocecal valve, a large amount of red blood was observed in the lumen, preventing the evaluation of the small bowel mucosa (Fig. 9.3). The capsule was excreted naturally within 48 h. An antegrade double-balloon enteroscopy, performed 2 days after capsule excretion, showed a 2.5-cm submucosal mass with a small, non-bleeding ulcer, 2.2 m beyond the ligament of Treitz (Figs. 9.4, 9.5). A biopsy was performed at the ulcer edge and a tattoo was placed a few centimeters proximally to the lesion. The patient underwent laparotomy and during surgery the tattoo was easily identified. A well-circumscribed lesion, measuring 3.5 × 1.5 × 2.5 cm, was discovered in the mid jejunum (Fig. 9.6). A small bowel resection and a side-to-side anastomosis of the small bowel were performed. The patient had an uneventful postoperative course and was discharged on the sixth postoperative day. The pathological examination of the resected lesion revealed a mesenchymal tumor. The resection margins

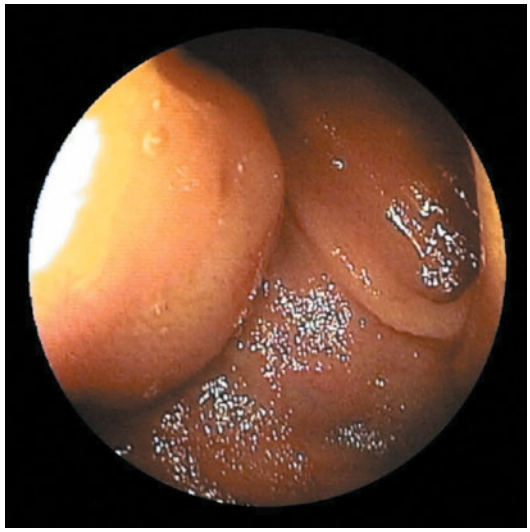

Fig. 9.4 Double balloon enteroscopy findings: submucosal mass protruding into the lumen

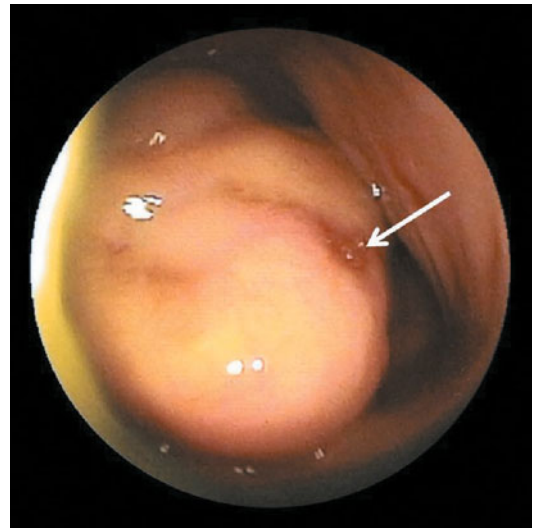

Fig. 9.5 Double balloon enteroscopy findings: submucosal mass with a small ulcer (*arrow*)

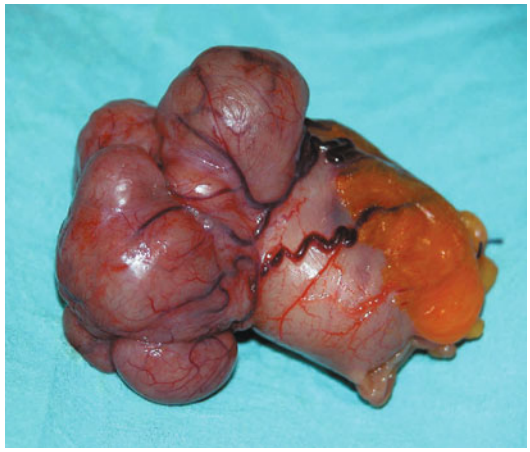

Fig. 9.6 Resected small bowel

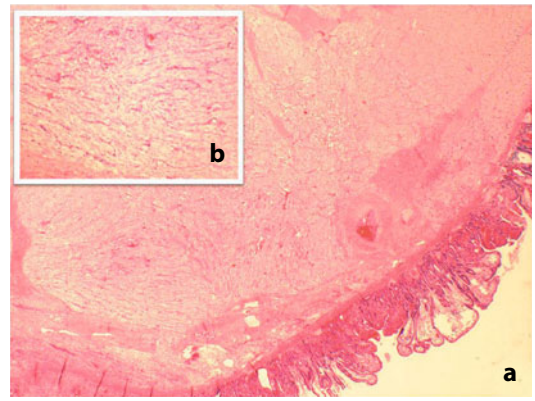

Fig. 9.7 Microscopy image of a mesenchymal neoplasm composed of spindle cells with elongated nuclei. H&E stain; magnification 2× (**a**) and 10× (**b**).

were negative for neoplastic tissue and no metastases were noted in any of the regional resected lymph nodes. The mesenchymal tumor was classified as a gastrointestinal stromal tumor (GIST): hematoxylin and eosin staining demonstrated spindle cells with moderate pleomorphism (Fig. 9.7), low mitotic activity (<5 mitoses/50 HPF; MIB1 proliferative index=1%) and extracellular collagen globules ("skenoid fibers"). No necrosis was observed. Immunohistochemical stainings for CD34, DOG1, and cKIT protein were positive. The patient was followed-up for 2 years after the small bowel resection; blood tests were persistently normal and she did not experience any new episode of gastrointestinal bleeding.

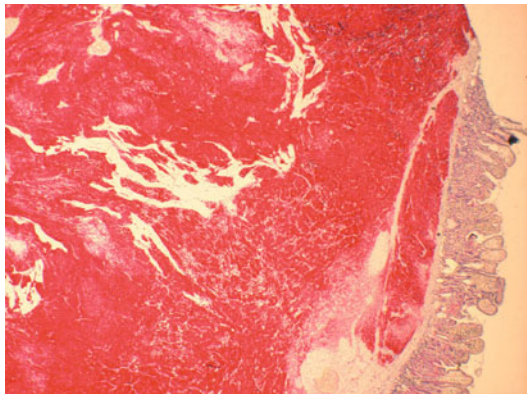

Fig. 9.8 Microscopy image shows uniform and diffuse cKIT immunolabeling (magnification 2×)

Open Issues

Some authors have estimated that the CE miss-rate for neoplastic diseases can reach 18-20% [3-5]. Several reasons contribute to this relatively high percentage, but the crucial one is probably the difficulty in discriminating masses from bulges based on CE findings. However, several visual clues may help, i.e. changes in mucosal characteristics, presence of bridging folds, transit abnormalities, repetitive images, and synchronous lesions. Therefore, scoring systems [6-8] based on these clues have been proposed and were recently validated. Another important issue that affects the capability of CE to identify small bowel lesions seems to be procedure timing. Several authors reported that, in patients with obscure-overt bleeding, CE performed shortly after the bleeding episode has the greatest potential for lesion detection [9, 10].

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Multiple Choice Questionnaire

1) The estimated miss rate of capsule endoscopy (CE) for neoplastic diseases is

a. 1%
b. 5%
c. 10%
d. 20%
e. 30%

2) Among the following visual clues, which one does not suggest the presence of a small bowel mass?

a. changes in mucosal characteristics
b. presence of bridging folds
c. presence of arteriovenous malformations
d. transit abnormalities
e. synchronous lesions

3) Which is the most important parameter that affect the sensitivity of CE to identify small bowel lesions?

a. procedure timing
b. experience of the physician
c. technology of the capsule
d. preparation of the patient
e. all of them

1.d – 2.c – 3.a

Case 9.2
Videocapsule Endoscopy and Therapeutic Enteroscopy for the Management of Small Bowel Polyps in a Patient with Peutz-Jeghers Syndrome

Giovanni Battista Rossi, Giovanni Di Nardo, Mario de Bellis, Salvatore Oliva and Elena Di Girolamo

Background

Peutz-Jeghers syndrome (PJS) is characterized by multiple hamartomatous polyps of the gastrointestinal tract and mucocutaneous pigmentation occurring around the body orifices [1-2]. This autosomal dominant disease is caused by a germline mutation in the STK11/LKB1 gene and carries an increased risk of several types of cancer, such as gastrointestinal tumors, pancreatic cancer, genital tract malignancies and, especially, breast cancer [1-2]. Approximately 50% of patients develop intussusception by the age of 15, because of jejunal and/or ileal polyps [3]. Previously, this complication was usually treated by surgery. Recently, video-capsule-endoscopy (VCE) and enteroscopy have changed both the diagnostic and the therapeutic management of small bowel hamartomas [4-8].

Clinical Presentation

This is the case of a 21-year-old girl diagnosed with PJS when she was 14 years old. At that time she had been suffering recurrent episodes of abdominal pain and occult gastrointestinal (GI) bleeding, with subsequent anemia. After a complete work-up a diagnosis of PJS was made. The latter was complicated by the presence of large small bowel polyps, which had been detected radiologically. The intestinal polyps were causing recurrent episodes of intussusceptions, with subsequent abdominal pain and GI bleeding. Therefore, the patient underwent exploratory laparotomy and an intraoperative therapeutic enteroscopy was performed at the time of surgery. Endoscopy via an enterotomy in the small bowel identified several large polyps, which were simultaneously removed, avoiding multiple enterotomies and the risk of the short bowel syndrome associated with resection. Pathological examination of the specimens confirmed that the removed polyps were hamartomas. After two years of relatively good health, the patient was diagnosed with dysplastic adenomatous foci, which had arisen in a hamartoma of the major papilla, observed during surveillance by means of an upper endoscopy. Since this pathological result carries a high risk of cancer and the size of the ampullar hamartoma had been increasing over time, the patient underwent prophylactic surgical ampullectomy. Surgery was uneventful and the patient resumed a normal life. When she was 18 years old, a surveillance VCE (Pillcam SB, Given Imaging) revealed the presence of multiple small bowel polyps less than 5 mm in diameter. No interven-

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tion was deemed necessary at that time and a repeat VCE was scheduled for two years later. That examination showed that the small bowel polyps had increased in size. Of these, the largest measured approximately 10 mm and they were located in the proximal jejunum (Figs. 9.9, 9.10). Polyps of this size should be removed in order to reduce the risk of cancer, according to the "expert opinion" from recent guidelines [1, 7]. Therefore, the patient underwent single-balloon enteroscopy under general anesthesia. Despite the fact that both anterograde and retrograde routes were used, the enteroscopy was not complete. However, the 10-mm polyps detected by VCE in the proximal jejunum were reached and could be removed by means of snare polypectomy (Figs. 9.11–9.13). Unfortunately, no polyp was retrieved for pathology. The procedure was uneventful and the previous surgeries did not influence the outcome of the enteric polypectomies.

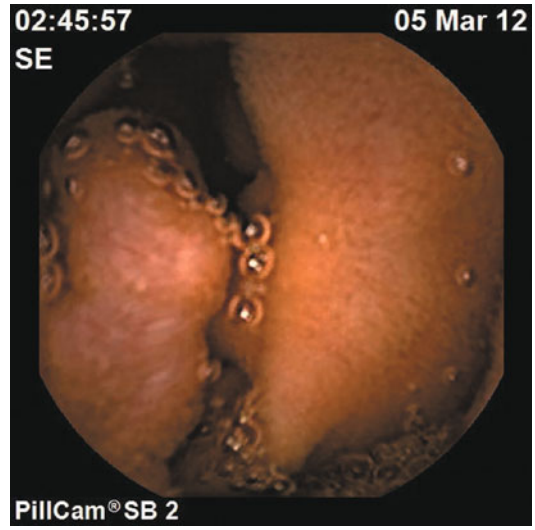

Fig. 9.9 VCE: bilobed polyp in the proximal jejunum

Fig. 9.10 VCE: bilobed polyp in the proximal jejunum

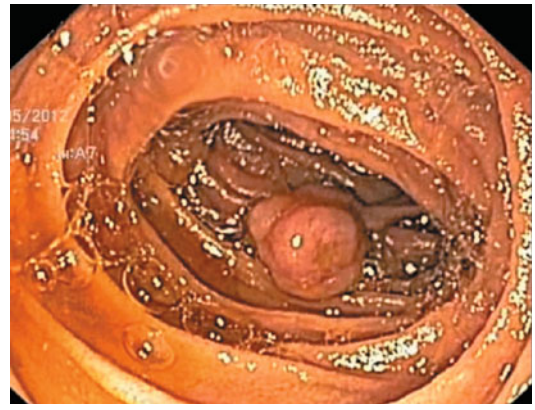

Fig 9.11 Single-balloon enteroscopy: pedunculated polyp in the proximal jejunum

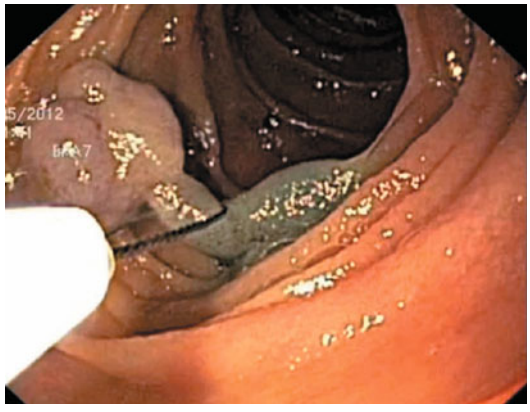

Fig 9.12 Single-balloon enteroscopy: snare polypectomy of a pedunculated polyp in the proximal jejunum

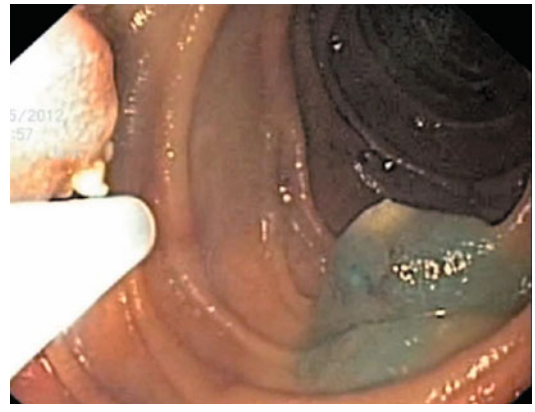

Fig. 9.13 Single-balloon enteroscopy: snare polypectomy of a pedunculated polyp in the proximal jejunum

Open Issues

Compared to traditional radiology, VCE is more accurate in the surveillance of patients with PJS [4]. This diagnostic method is not only able to detect polyps less than 10 mm in diameter, but it also reduces radiation exposure and is well accepted by patients [2]. Therefore, VCE allows complete and regular surveillance of the small bowel in PJS patients. Balloon enteroscopy (single or double balloon) allows the non-invasive resection of small bowel polyps by means of standard snare polypectomy [5-8]. Both procedures have changed the current management of patients with PJS, reducing the need of high-risk surgery.

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Multiple Choice Questionnaire

1) Peutz-Jeghers syndrome (PJS) is

a. characterized by multiple hamartomatous polyps of the gastrointestinal tract and mucocutaneous pigmentation occurring around the body orifices
b. associated with an increased risk of cancer
c. an autosomal dominant disease
d. all of the above
e. none of the above

2) The most common cancer(s) associated with PJS is/are

a. gastrointestinal tumors
b. pancreatic cancer
c. genital tract malignancies
d. lung cancer
e. breast cancer

3) Gastrointestinal polyps may cause

a. gastrointestinal bleeding and anemia
b. abdominal pain due to intussusception
c. gastrointestinal obstruction or infarction
d. all of the above
e. none of the above

4) Which of these statements is false?

a. VCE detects polyps less than 10 mm in diameter
b. VCE reduces radiation exposure and is well accepted by patients
c. balloon enteroscopy allows standard snare polypectomy of small bowel polyps
d. therapeutic balloon enteroscopy has reduced the need for surgical resection of small bowel polyps
e. VCE and balloon enteroscopy has not changed the current management of patients with PJS

1.d – 2.e – 3.d – 4.e

Case 9.3
An Unusual Finding at Videocapsule Enteroscopy

Enrico Ricci and Angelo De Padova

Background

Iron-deficiency anemia represents a major public health problem [1] and is a frequent cause for a referral to a gastroenterologist. Published guidelines for the evaluation of iron-deficiency anemia recommend that both esophagogastroduodenoscopy, with duodenal biopsies to rule out celiac disease, and ileocolonoscopy should be performed to exclude bleeding lesions [2]. However, even when these investigations are carefully executed, up to 30% of patients lacks a definitive diagnosis, and are therefore candidates for **capsule enteroscopy** [3]. Videocapsule findings in most cases include angectasias, arteriovenous malformations, small bowel Crohn's disease, NSAID-associated enteropathy, polyps, neoplasms, and celiac disease, but also rarer etiologies, as described in the following.

Clinical Presentation

A 25-year-old Nigerian man living in Italy for 3 years was referred to the Medical Department of our institution with complaints of asthenia, frequent headache and rapid worsening of his condition over the past 2 months. His vital signs were normal, and on physical examination only a mucosal paleness and leg edema were remarkable. Biochemical tests showed **severe anemia** (hemoglobin 3 g%), with a significant reduction of mean corpuscular volume (52 fL), and low serum iron (9 μg%) and ferritin (3 ng/mL). Total protein was also low (5.1 g%). His bowel habit was regular, and no signs of hemorrhage were found. Chest radiography results were normal, as were the splanchnic organs as seen on abdominal ultrasonography, but a diffuse, non-corpuscular fluid was noted, accompanied by a pleural effusion. After hospital admission, the patient was rapidly transfused, with a good recovery of his hemoglobin level, up to 7.3 g%. An esophagogastroduodenoscopy was performed, which allowed a diagnosis of antral gastritis and bulbar erosions. Gastric biopsies revealed a severe erosive gastritis, with no evidence of *Helicobacter pylori*, while duodenal biopsies identified erosive duodenitis but without specific features. The results of a colonoscopy, with exploration of the ileal mucosafor 30 cm, were normal.

Our diagnostic work up for patients with severe sideropenic anemia with no sign of hemorrhage currently includes a videocapsule enteroscopy. Accordingly, our patient swallowed a Given SB2 capsule for the evaluation

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of the small bowel. The images revealed numerous 6- to 8-mm whitish **parasites**, adhered to the bowel wall via an appendage and scattered throughout the gut, but mainly along the jejunum (Figs. 9.14–9.16). The jejunal and ileal mucosae were patchy and erythematous, in addition to areas of petechiae (Fig. 9.17).

A stool parasitology exam revealed infection with **Ancylostoma duodenale**. This soil-transmitted nematode of the genus *Ancylostoma*, once it penetrates the human body, reaches the gut, and, when mature, lays thousands of eggs per day. **Hookworms** attach to the intestinal mucosa and secrete enzymes that allow them to ingest villous tissue and

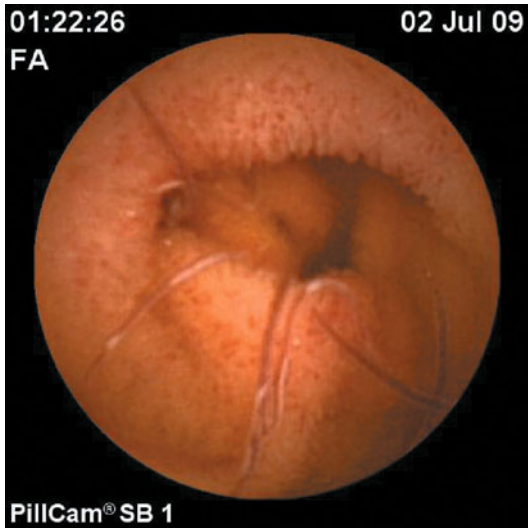

Fig. 9.14 Numerous 6- to 8-mm whitish parasites adhere to the bowel wall via an appendage

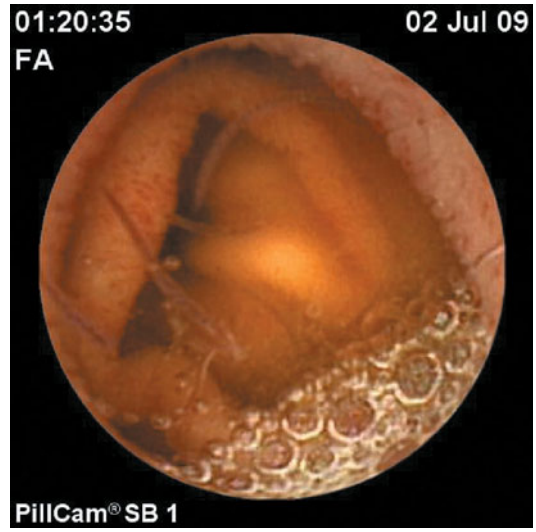

Fig. 9.15 Numerous 6- to 8-mm whitish parasites adhere to the bowel wall via an appendage

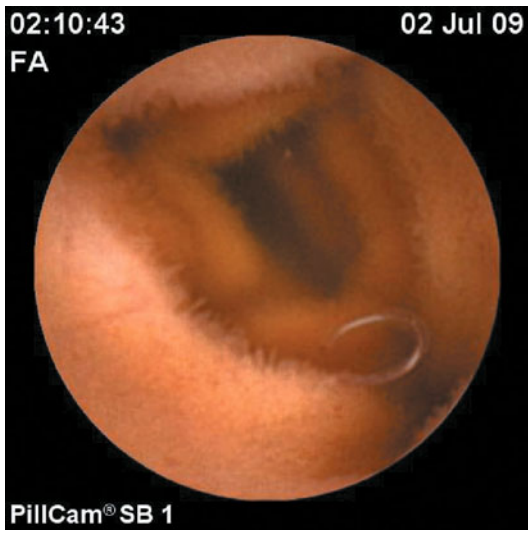

Fig. 9.16 Numerous 6- to 8-mm whitish parasites adhere to the bowel wall via an appendage

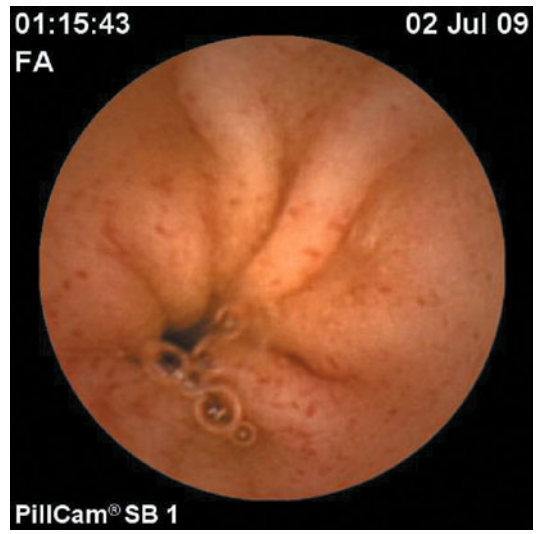

Fig. 9.17 Jejunal and ileal mucosae: patchy and erythematous

blood. Hemoglobinases within the hookworm digestive canal enable the degradation of human hemoglobin for use as an essential nutrient source. The infection can cause abdominal pain, loss of appetite, severe protein deficiency. and iron deficiency anemia. Ancylostomiasis is one of the most important **parasitic infections** worldwide [4], with up to 740 million people currently infected, mostly throughout Sub-Saharan Africa, tropical regions of the Americas, South China, and Southeast Asia.

Our patient was treated with a mono-somministration of 400 mg albendazole. One week later, the stool parasitology exam was negative for any infection. After 2 months, the patient had normal hemoglobin (15 g%), mean corpuscular volume (88 fL), ferritin (52 ng/mL), iron (92 μg%) and total protein (6.9 g%). His general condition likewise improved rapidly.

Open Issues

Iron-deficiency anemia is a conventional reason for VCE investigation when esophagogastroduodenoscopy and ileocolonoscopy are not able to achieve a definite diagnosis. The usual findings include angiectasias, arteriovenous malformations, small bowel Crohn's disease, NSAID-associated enteropathy, polyps, neoplasms, and celiac disease. Our clinical case points out that anemia can also be due other, rarer etiologies, which have to be considered when other diagnoses are ruled out. **Helminthic infections** are widespread throughout theTropics and Subtropics, with hundreds of millions of people vulnerable to infection. A simple stool parasitology exam, when VCE findings are indicative of a helminthic infection, can lead to a clear diagnosis and a focused therapy.

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Multiple Choice Questionnaire

1) Ancylostoma duodenale
- a. is a very common cause of iron-deficiency anemia
- b. cannot be diagnosed by detecting the worms in a stool parasitology exam
- c. rarely triggers severe anemia
- d. can be detected with videocapsule enteroscopy
- e. does not occur worldwide

2) How is Ancylostomiasis considered?
- a. parasitic infection
- b. bacteric infection
- c. viral infection
- d. infection by a coccobacillus

3) In which countries is Ancylostomiasis more frequent?
- a. Saharan Africa
- b. tropical regions of the Americas
- c. South China
- d. Southeast Asia
- e. all of them

1.d – 2.a – 3.e

Case 9.4
Small Bowel Metastases from Squamous Cell Carcinoma of the Lung

Elena Di Girolamo, Mario de Bellis, Pietro Marone, Valentina D'Angelo, Andrea Belli and Giovanni Battista Rossi

Background

Gastrointestinal (GI) metastases of lung cancer are quite rare, ranging from 0.2% to 1.7% in clinical series, even if they occur in 7.3-12.2% of autopsy cases [1-3]. The small bowel is the most frequent site of metastatic lung cancer in the GI tract [4, 5]. Mostly asymptomatic, small bowel metastases from lung cancer are usually diagnosed only in the presence of life threatening symptoms, such as small bowel obstruction, perforation or bleeding.

This is the report of a patient with squamous cell carcinoma of the lung who was eventually diagnosed with small bowel metastases after recurrent episodes of GI bleeding.

Clinical Presentation

A 78-year-old man was initially diagnosed with locally advanced squamous cell carcinoma of the lung. The tumor involved the left superior lobe and was associated with mediastinal metastatic lymphadenopathy (N2) at the diagnosis. Six months after radiotherapy, the patient had several minor episodes of GI bleeding which determined the onset of severe anemia, with a hemoglobin of 6.3 g/dl (normal values = 14-18 g/dl). There were no other major abdominal symptoms or complaints in the personal history of the patient. At upper endoscopy a diagnosis of erosive gastritis was made and this was thought to be the possible cause of GI bleeding. Therefore, high doses of proton pump inhibitors were administered to the patient. Despite therapy, a new episode of GI bleeding occurred a week later. A repeat upper endoscopy revealed that the gastritis was almost completely healed. A complete colonoscopy with exploration of the terminal ileum showed that both the colon and the terminal ileum were normal. Eventually, the patient underwent Video-Capsule-Endoscopy (VCE) (VCE, Pillcam SB, Given Imaging) which showed several polyp-like lesions covered by congested mucosa, with no signs of recent bleeding (Figs. 9.18-9.20). All the lesions were localized in the distal jejunum and proximal ileum; the intestinal lumen was partially obstructed by the larger lesions. A subsequent (18F) Fluorodeoxyglucose Positron Emission Tomography (FDG-PET)/CT scan failed to detect the bowel lesions, while it confirmed the presence of both lung cancer and mediastinal lymphoadenomegaly (Fig. 9.21).

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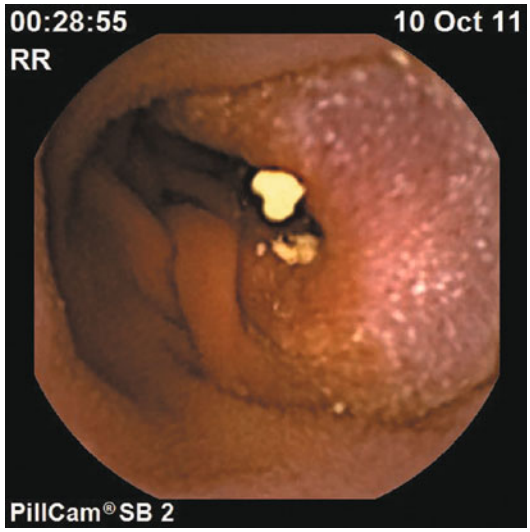

Fig. 9.18 VCE: polyp-like lesion in the distal jejunum. The lesion is covered by congested mucosa

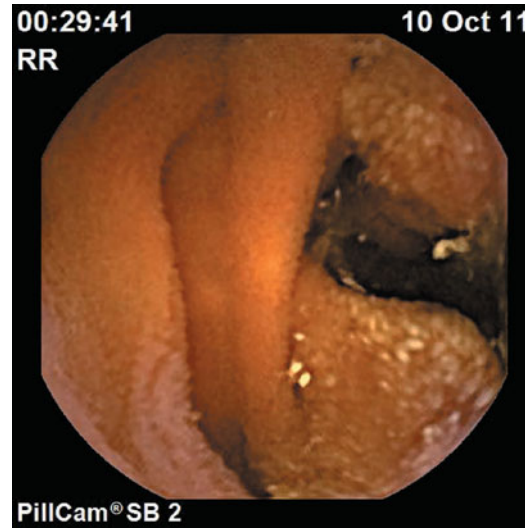

Fig. 9.19 VCE: polyp-like lesion in the distal jejunum/proximal ileum. Congested mucosa with no signs of recent bleeding

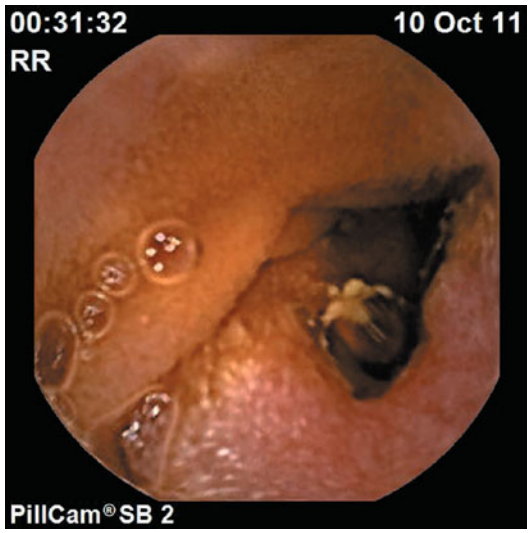

Fig. 9.20 VCE: polyp-like lesions in the distal jejunum/proximal ileum. There is partial obstruction of the intestinal lumen

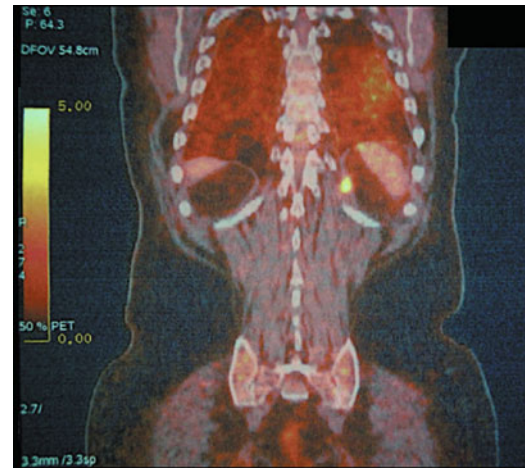

Fig. 9.21 PET/CT: no lesions detected in the small bowel

Recurrent episodes of GI bleeding and a limited neoplastic involvement of the small bowel were felt to be a clear indication for palliative surgery [2, 6]. At the time of surgery, two consecutive strictures were found in the small bowel, respectively, in the distal jejunum and the proximal ileum, which contained the polyp-like lesions detected at VCE. These findings were then confirmed at pathology. Macroscopically, the surgical specimen contained several whitish nodules which infiltrated and ulcerated the enteric mucosa. Microscopically, there were findings consistent with sub-mucosal infiltration of

the small bowel from squamous cell carcinoma of the lung.

After surgery, the patient recovered and had a complete resolution of his abdominal symptoms with no complications. He was able to resume and keep a normal oral intake until he died of his lung cancer 3 months later.

Open Issues

Clinically significant symptoms due to small bowel metastastes from lung cancer are rare. Both CT scan and PET are useful for the diagnosis of small bowel metastases from lung cancer. CT scan shows localized thickening of the small bowel wall and/or polyp-like lesions protruding in its lumen [7]. PET seems more accurate than CT scan, but often is not diagnostic.

In our case, VCE was crucial to make the correct diagnosis. We accordingly recommend VCE after standard endoscopy in order to rule out small bowel metastases from lung cancer, when recurrent episodes of GI bleeding occur.

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Multiple Choice Questionnaire

1) At autopsy, the incidence of small bowel metastases from lung cancer is
a. similar to the clinical incidence
b. more frequent than the clinical incidence, with a frequency of 0.2-1.7%
c. more frequent than the clinical incidence, with a frequency of 7.3–12.2%
d. as high as 35-50% of cases
e. lower than the clinical incidence

2) Clinical presentation of significant small bowel metastases from lung cancer occurs
a. only with gastrointestinal bleeding
b. with life threatening symptoms, such as small bowel obstruction, perforation or bleeding in the advanced stages of the disease
c. with specific symptoms in the early stage of the disease
d. with severe gastrointestinal symptoms in the early stages of the disease
e. with mild gastrointestinal symptoms, such as diarrhea and abdominal pain in the advanced stages of disease

3) The most useful diagnostic methods for diagnosing small bowel metastases from lung cancer is/are
a. upper endosopy
b. complete colonoscopy with exploration of the terminal ileum
c. CT-PET scan
d. capsule endoscopy and CT-PET scan
e. none of the above

4) Surgery with the resection of the affected small bowel is
a. curative
b. palliative, especially for small bowel obstruction and GI bleeding
c. high risk with a high mortality rate
d. none of the above
e. all the above

1.c – 2.b – 3.d – 4.b

Enteroscopy

Case 10.1
Capsule, Enteroscopy, or Radiology? The Gastroenterological Dilemma

Gabriele Marinozzi, Amilcare Parisi, Anselmo Della Spoletina, Antonio Astolfi, Stefano Ascani and Antonello Trecca

Background

The diagnosis of small bowel disease is still a mattern of concern among experts. Each imaging technique has its advantages but also its limits. This is particularly true when dealing with the management of patients with obscure gastrointestinal bleeding. In this group, the positive detection rates are in the range of 43–60% with double-balloon enteroscopy (DBE) [1, 2], 42–77% with capsule endoscopy (CE) [3], 21–24% with computed tomography (CT), and 5–25% with small bowel series [4], with the highest accuracies thus obtained with DBE and CE. Here we present a difficult case of gastrointestinal bleeding.

Clinical Presentation

A 51-year-old patient was admitted for melena (hemoglobin 10.9 g/dl) in January 2007. Upper and lower gastrointestinal endoscopy showed unremarkable findings. Abdominal CT scan was negative and the patient was discharged with iron supplementation. As an outpatient procedure, a CE of the small intestine was performed, which evidenced two nonbleeding telangiectasias in the terminal ileum. The patient was discharged and periodic serological examinations were stable until February 2009, when a second episode of melena occurred. The physical examination was normal, but the Hb had decreased to 10.2 g/dl. A scintigraphic study was negative for focal accumulations due to active gastrointestinal bleeding. Repeat CE confirmed the diagnosis of ileal telangiectasia. DBE showed an ulcerative broad lesion with irregular margins of 4 cm in diameter located within the first jejunal loop (Fig. 10.1). Histological and immunohistochemical examinations resulted in a diagnosis of gastrointestinal stromal tumor (GIST). A CT-enterography was negative for other locations of the disease and confirmed the presence of a mass located in the abdominal cavity (Fig. 10.2). The patient then underwent a small bowel resection with a side-to-side anastomosis (Fig.10.3). Histology performed on the resected specimen confirmed a high-risk GIST, mainly comprising fused cells with a mitotic index of 12 mitoses per 50 HPF, CD 117/C-KIT positive, S-100 negative, desmin negative, and without metastatic disease (Fig. 10.4). The patient had an uneventful postoperative course and was discharged on the seventh postoperative day, with medical therapy of imatinib (Gleevec)

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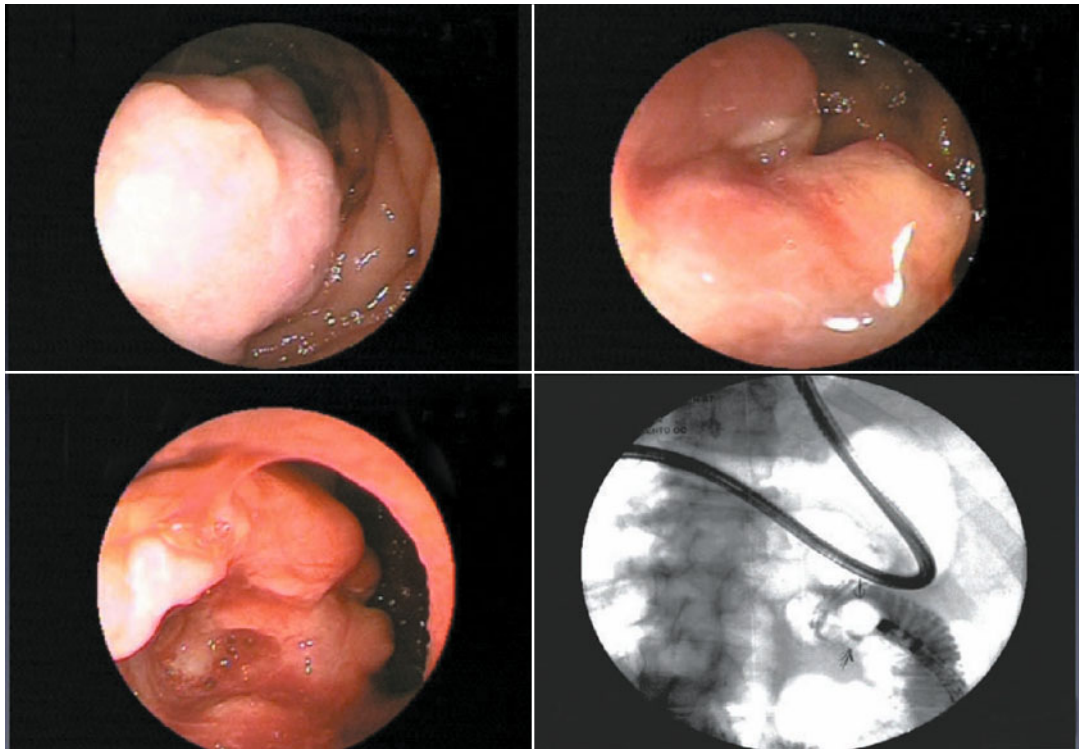

Fig. 10.1 Double-balloon enteroscopy shows a broad-based ulcerative lesion of the first jejunal portion

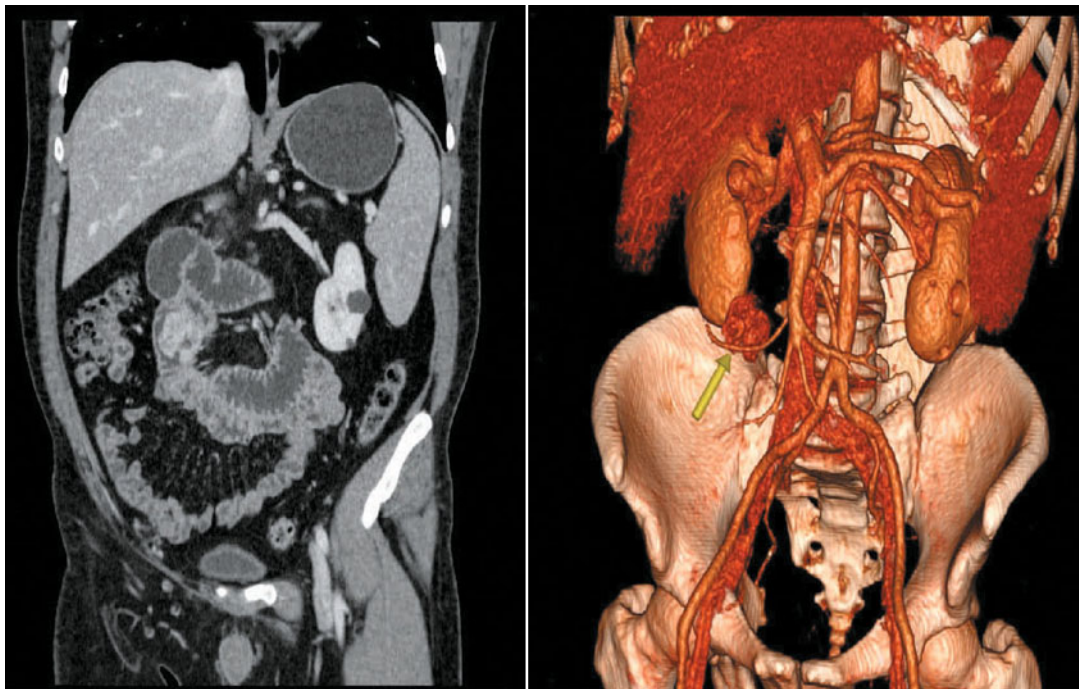

Fig. 10.2 CT scan of the abdomen, showing an abdominal mass

400 mg/die. The patient has been disease free during 3 years of follow-up.

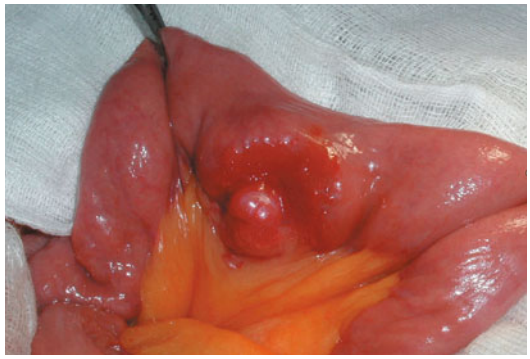

Fig. 10.3 Isolated jejunal loop at open surgery. The neoplastic mesenchymal lesion is clearly visible

Open Issues

Gastrointestinal bleeding is a commonly seen problem in clinical practice. Upper and lower endoscopy, including ileoscopy, shows unremarkable findings in 3–5% of patients [5]. The recent introduction of new imaging modalities, such as CE and DBE, can help the clinician to better identify the source of gastrointestinal bleeding, whether overt or occult. Okazaki showed that in over 1,044 cases of acute overt gastrointestinal bleeding, 13 (1.7%) represented small bowel bleeding, which in rare cases is due to small bowel tumors (SBTs) [6]. These constitue a rare clinical condition, accounting for only 1–3% of all primary gastrointestinal tumors. Among SBTs, GISTs are the most fre-

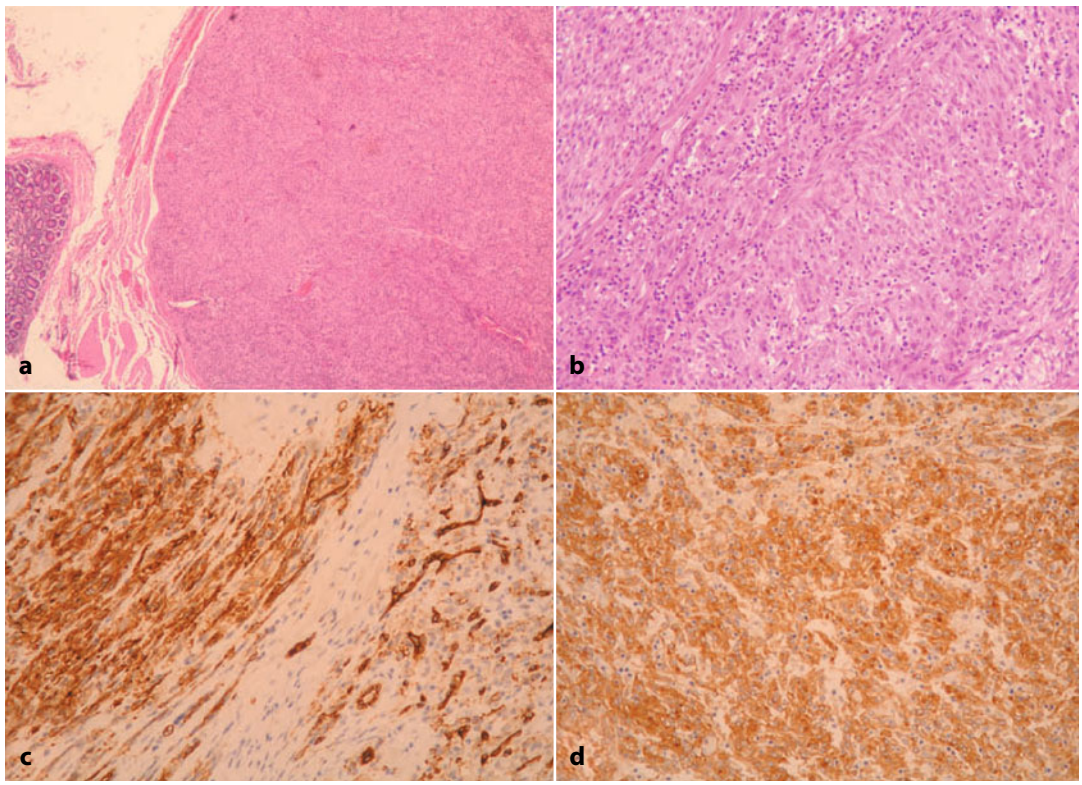

Fig. 10.4 a Gastrointestinal stromal tumor (GIST) extending to the muscularis mucosae, with spindle cells (H&E; original magnification 10×). **b** Moderate cellular atypia, without mitotic activity, and scattered small lymphocytes (H&E; original magnification 20×). **c** CD34 staining of the tumor (CD34 staining; original magnification 20×); **d** CD117/c-Kit immunostaining: strong cytoplasmic positivity of the neoplastic cells (CD117 staining; original magnification 20×)

quently occurring mesenchymal tumors and their most common location is the stomach and small bowel [7]. The accuracy rate of CE, DBE, and radiology for these tumors as reported in the international literature is controversial, with many authors suggesting that there are no differences between the methods. However, Nakatani, based on a series of 12 small bowel GISTS, recently reported that the detection rates of DBE, CE, and CT are 92%, 60% and 67%, respectively [8, 9]. The discrepancy may be due to the gross appearance of these tumors, which can present with intraluminal, intramural, or extramural growth [10]. In our patient, the tumor was missed after two CE procedures, even if a prompt endoscopic investigation would have been useful to compare their accuracy. DBE was effective in diagnosing the tumor and in allowing its histological characterization, thus confirming enteroscopy as an accurate method to obtain a final diagnosis in patients with small bowel GISTs.

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Multiple Choice Questionnaire

1) What is the incidence of small bowel tumors as a percentage of all neoplasms of the digestive tract?

a. 5–10%
b. 10–15%
c. 1–3%
d. 15–20%

2) True or false: Capsule endoscopy can always be performed along the digestive tract.

a. yes
b. no
c. yes, only if we use the patency system

3) Which GISTs are associated with a higher risk of malignancies?

a. GISTS are a benign disease
b. those with a mitotic index of > 10 mitosis per 50 HPF
c. those with a mitotic index of > 5 mitoses per 50 HPF
d. those with a mitotic index of > 50 mitoses per 50 HPF

1.c – 2.c – 3.b

Case 10.2
Crucial Additional Value of Enteroscopy in the Diagnosis, Assessment, and Further Management of Small Bowel Crohn's Disease

Maria Elena Riccioni, Alessandra Bizzotto, Franco Scaldaferri, Viviana Gerardi, Laura Maria Minordi, Vincenzo Arena, Antonio Gasbarrini and Guido Costamagna

Background

The diagnosis of Crohn's disease is challenging as it combines clinical and laboratory clues with endoscopic, radiological and histological findings [1, 2]. Consequently, the diagnosis is often delayed or the disease is misdiagnosed, in particular in the case of mainly or exclusively ileal or jejunal involvement [3]. Enteroscopy enables deep intubation, with endoscopic visualization of the small bowel and otherwise complicated or unobtainable tissue sampling. In a secondary-tertiary center, based on a strong clinical suspicion of inflammatory bowel disease (IBD) with laboratory findings consistent with Crohn's disease, radiology could be the first step in making a diagnosis, followed directly by enteroscopy if there are clear signs of jejunal or ileal localization. This is a "top down" strategy in the diagnosis of Crohn's disease, that might be effective in patients in whom the diagnosis is delayed.

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Clinical Presentation

A 34-year-old male was referred to our tertiary gastroenterology unit after a not fully conclusive diagnostic work-up and a therapeutic attempt based on the suspicion of IBD. The patient reported that during the last 5 years he had suffered from slight persistent evening fever associated with mainly mesogastric chronic abdominal pain and weight loss (10 kg in 2 years). Clinical history did not reveal obstructive symptoms neither did the patient describe other symptoms such as bloody or mucous diarrhea. He had undergone esophagogastroduodenoscopy, abdominal ultrasound investigation and stool tests at another hospital. Laboratory tests showed a slightly increased CRP and alpha-1-acid glycoprotein without anemia and or leukocytosis. According to the results of two colonoscopies showing, in histological specimens only, chronic active inflammation, cryptitis and mucosal edema, the patient was initially diagnosed with ulcerative colitis. He was therefore started on cycles of corticosteroid therapy (prednisone) plus mesalazine with partial benefit for 3–4 years. He was admitted to our unit after a clinical relapse (severe abdominal pain, further weight loss) with the onset of mucous diarrhea. An MR-enterography, strongly suggestive for Crohn's disease, depicted a stratified jejunum and proximal

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ileal wall thickening with five ileal strictures (ranging from 2–2.5 cm to 9 cm), discontinuous pre-stenotic luminal dilations, mesenteric fibro-fatty proliferation with associated "comb-sign", and mesenteric lymphadenopathy without evidence of fistulas, abscesses, or fluid collections (Figs.10.5, 10.6). The patient was scheduled for anterograde

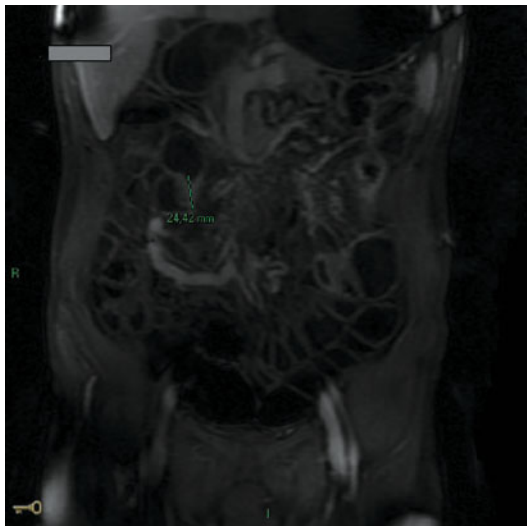

Fig. 10.5 MR-enterography: small bowel wall thickening, mesenteric fibro-fatty proliferation with associated "comb-sign", luminal dilations

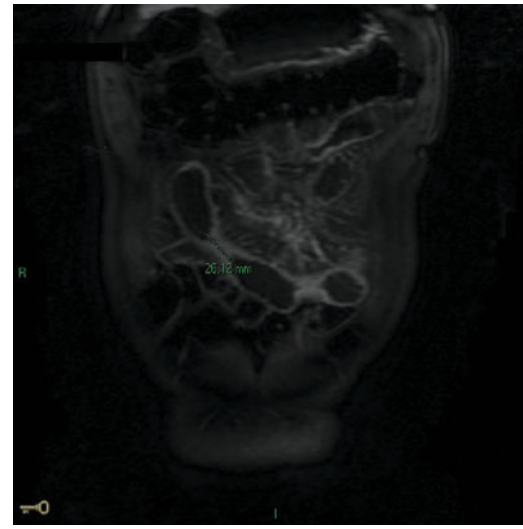

Fig. 10.6 MR-enterography: small bowel wall thickening with ileal strictures, discontinuous pre-stenotic luminal dilations, mesenteric fibro-fatty proliferation with associated "comb-sign", mesenteric lymphadenopathy

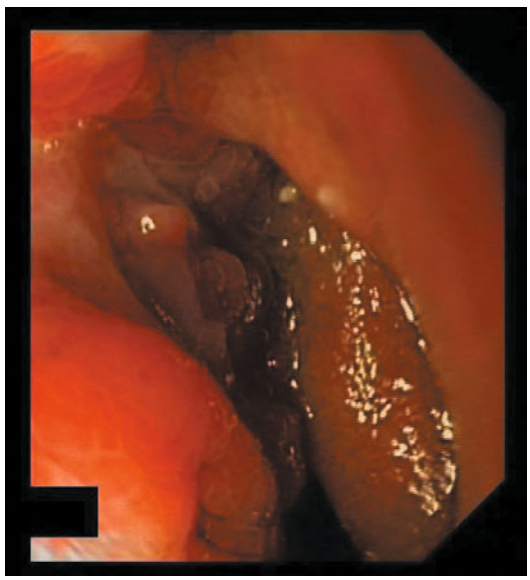

Fig. 10.7 Enteroscopy: small bowel deep serpiginous irregular ulcerations together with mucosal hyperemia, edema and luminal substenosis

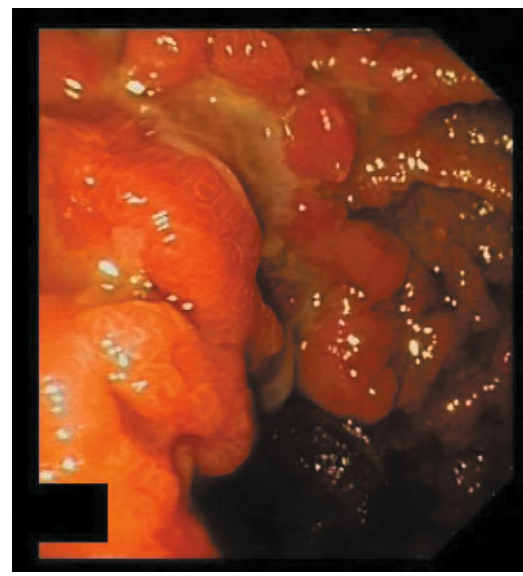

Fig. 10.8 Enteroscopy: small bowel deep serpiginous irregular ulcerations together with mucosal hyperemia and edema

enteroscopy with biopsies. Push and pull enteroscopy with an oral approach using the single-balloon enteroscope (SIF-Q180, Olympus Optical, Tokyo, Japan) was successfully performed with the patient under general anesthesia with endotracheal intubation according to our anesthesiology standards. The procedure was fairly easy to perform as far as the proximal jejunum. However, deeper intubation failed because of perivisceral adhesions. Endoscopic imaging confirmed the presence of multiple, patchy, deep serpiginous and irregular ulcerations, with fibrin deposits accompanied by mucosal hyperemia, edema, and luminal substenosis (Figs. 10.7, 10.8). Multiple targeted biopsies were performed under fluoroscopy control, reducing the risk of perforation. Finally, fluoroscopic assessment at the end of the procedure proved the absence of bowel leaks. Histology showed extensive inflammation with granulation tissue and confirmed the suspected diagnosis (Fig. 10.9). Taken together, the clinical, endoscopic, radiological, and histology elements showed that the patient had small bowel Crohn's disease. His therapy was accordingly adjusted and a tailored therapy, based on azathioprine and high-dose corticosteroids, was started, yielding complete symptomatic relief even after the steroids were later discontinued.

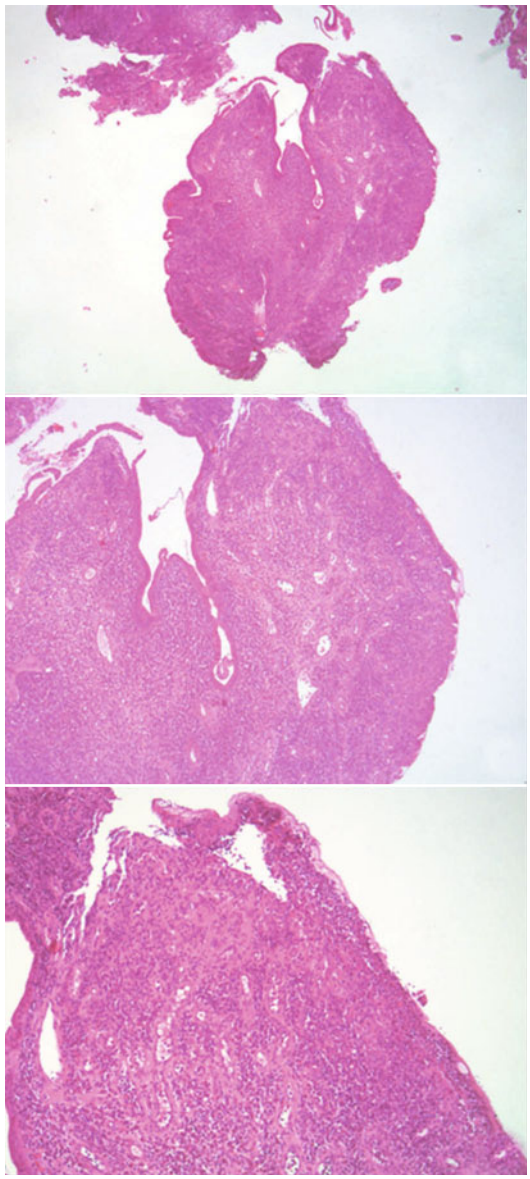

Fig. 10.9 Photomicrograph of the biopsy specimen showing extensive inflammation with granulation tissue

Open Issues

Radiology and endoscopy are complementary procedures in the diagnosis, assessment, and further management of Crohn's disease, with a good correlation in defining the clinical activity of the disease [4]. Especially during the diagnostic phase, more than one technique is desirable. Biopsies should be performed in all segments, regardless of the area of endoscopically confirmed disease activity. Extensive small intestinal disease imposes a strict follow-up. Current clinical guidelines do not mandate the timing of follow-up. Whether CT or MR-enterography is superior to ileoscopy in follow-up remains to be further evaluated, although the endoscopic assessment of "mucosal healing" in Crohn's disease is an intriguing goal in its management [1, 5]. Capsule endoscopy, although apparently accurate, is hampered in its application in patients with suspected or known stenosing Crohn's disease.

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Multiple Choice Questionnaire

1) The diagnosis of Crohn's disease is made by
a. histology
b. endoscopy
c. radiology
d. clinical picture
e. all of the above

2) The most frequent localization of Crohn's disease is
a. perianal
b. colonic
c. ileo-colonic
d. duodenal
e. jejunal

3) First-level examinations for the diagnosis of Crohn's disease consist of
a. stool culture
b. hemoglobin and inflammatory markers
c. ileocolonoscopy
d. radiological imaging of the ileum
e. all of the above

4) Ileoscopy can be performed by
a. double-balloon enteroscopy
b. push and pull enteroscopy
c. capsule endoscopy
d. none of the above
e. all of the above

5) CT and MRI enterography consist of
a. abdominal imaging with liquid distension of intestinal loops
b. abdominal imaging after the anal injection of barium
c. abdominal imaging after the anal injection of air
d. none of the above
e. all of the above

1.e – 2.c – 3.e – 4.e – 5.a

Case 11.1
Heterotopic Pancreas Presenting as a Jejunal Nodule in a Young Patient with Breast Cancer

Antonio Crucitti, Pasquina M. C. Tomaiuolo, Andrea Mazzari and Ugo Grossi

Background

Heterotopic pancreas (HP) is defined as the presence of pancreatic tissue in an aberrant location, without any vascular or anatomical continuity with the normal pancreas. HP is a rare condition occurring in 0.11%-0.21% at autopsy, with a male to female ratio of 3:1 and a peak age incidence between the 4th and 6th decades of life. Up to 90% of HPs are found in the upper gastrointestinal tract, involving the stomach, duodenum or jejunum. We report a case of HP presenting as a jejunal nodule found in a young girl during the staging of breast cancer.

Clinical Presentation

A 28-year-old Caucasian woman was referred to our department for a 4-cm palpable lump at the junction of the upper and lower outer quadrants of the right breast. She also complained of vague abdominal discomfort which had occurred intermittently for the past 2 years. Concomitant mammography and ultrasound examinations along with MRI of the breast and thoracoabdominal CT were used for clinical tumor staging. The CT scan showed a small nodular area in the distal jejunal loops (Fig. 11.1). The patient underwent nipple-sparing right mastectomy with immediate tissue expander breast reconstruction and sentinel node biopsy, followed by axillary lymph node dissection. Histopathological examination showed an infiltrating ductal carcinoma.

Shortly thereafter, an 18.1-mm nodule on the serosal surface of the distal jejunum was identified at magnetic resonance enterography (MRE); the lesion showed strong

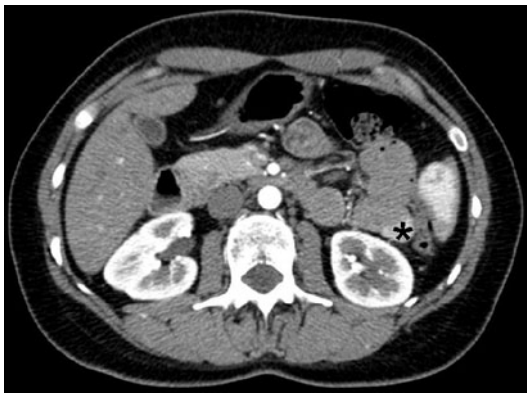

Fig. 11.1 Arterial phase CT (computed tomography) imaging showing a nodular area of the jejunum (*asterisk*)

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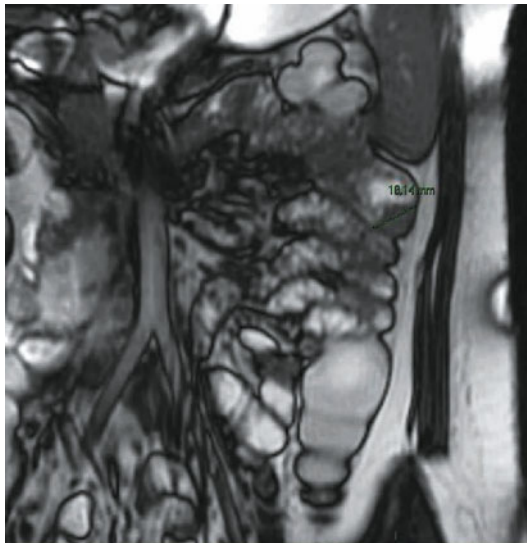

Fig. 11.2 MRE (magnetic resonance enterography) showing an 18.1-mm jejunal nodule

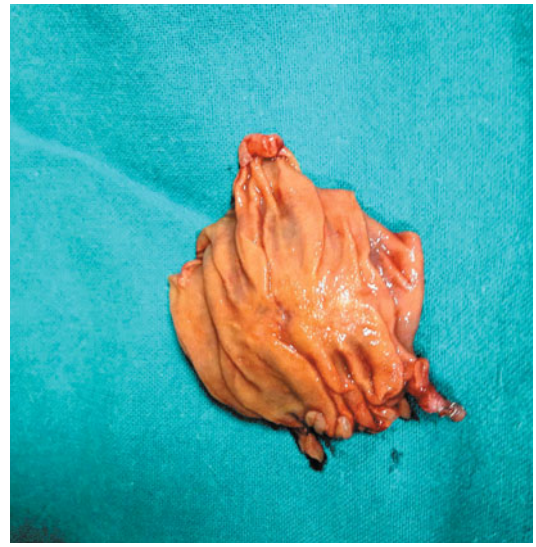

Fig. 11.4 Photograph of the cut, resected specimen shows an intact mucosa

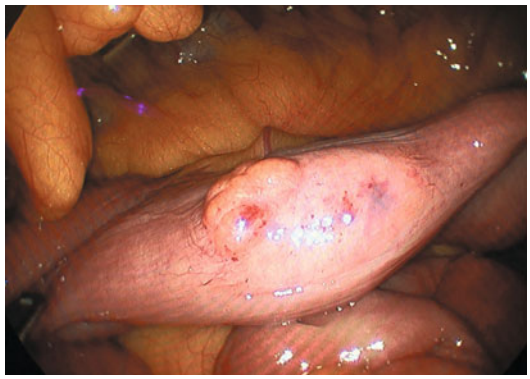

Fig. 11.3 Jejunal nodule at laparoscopic exploration

enhancement during the arterial phase, with persistent enhancement during the portal venous phase, suggestive of a gastrointestinal stromal tumor (Fig. 11.2). Due to the persistence of abdominal symptoms and the suspected jejunal tumor, the patient underwent laparoscopy. A 2-cm exophytic, mammillated, yellowish, subserosal nodule of the distal jejunum was confirmed at exploration of the abdominal cavity. Laparoscopic jejunal resection was therefore performed (Figs. 11.3, 11.4). Histopathological examination revealed a heterotopic pancreas. The postoperative period was uneventful.

Open Issues

Heterotopic pancreas (HP) was first reported in 1727 by Jean Schultz and histologically confirmed in 1859 by Julius Klob [1, 2]. HP most often represents an incidental finding, although in up to 40% of cases it becomes symptomatic, especially when located in the stomach and larger than 1.5 cm [3].

Although rare, several complications and clinical manifestations have been reported, including acute and chronic pancreatitis, gastrointestinal bleeding, and malignant transformation [3-7]. For these reasons, HP can pose a diagnostic and therapeutic challenge, especially in oncologic patients in whom it may mimic disease relapse.

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Multiple Choice Questionnaire

1) What is the reported incidence of heterotopic pancreas at autopsy?

a. less than 1%
b. 2–8%
c. 8–10%
d. 10–15%
e. 20–25%

2) What is the percentage of asymptomatic HP?

a. 5%
b. 10%
c. 20%
d. 40%
e. 60%

3) Where is HP most frequently found?

a. stomach and duodenum
b. jejunum
c. ileum
d. colon
e. rectum

1.a – 2.e – 3.a

Case 11.2
Unusual Evolution of a Clinical Case of Crohn's Disease in a Patient with Multiple Surgeries and Multiple Fistulas

Annibale D'Annibale, Maria C. Di Paolo, Graziano Pernazza, Vito Pende, Giorgio Lucandri, Paolo Mazzocchi and Giovanni Alfano

Background

The clinical course of Crohn's disease (CD) is often unpredictable. Severe forms of the disease still require surgery in nearly 80% of these patients. Moreover, one third will require reoperation [1], with rates ranging from 20 to 25% at 5 years and 34 to 39% at 10 years [2, 3]. Many risk factors for repeated surgery have been hypothesized in the international literature. Among these, young age (below 16 years), stricturing behavior, intra-abdominal abscess, emergency surgery, and delayed diagnosis after surgery are recognized as the most relevant [4]. In the following we describe a severe case of CD with a difficult clinical course.

Clinical Presentation

A 40-year-old man came to our attention with a diagnosis of long standing CD not responding to medical treatment, even after corticosteroids. Lower endoscopy showed a stenosis of the ileocecal valve with a large flat lesion located just above the cecal region, confirmed by abdominal magnetic resonance (Fig. 11.5). The patient underwent a robotic right colectomy with an intracorporeal, isoperistaltic, handsewn double-layer, side-to-side, ileotransversal anastomosis (Fig. 11.6). Histology on the resected specimen confirmed an inflammatory bowel disease with the features of CD and an adenomatous lesion with a high-

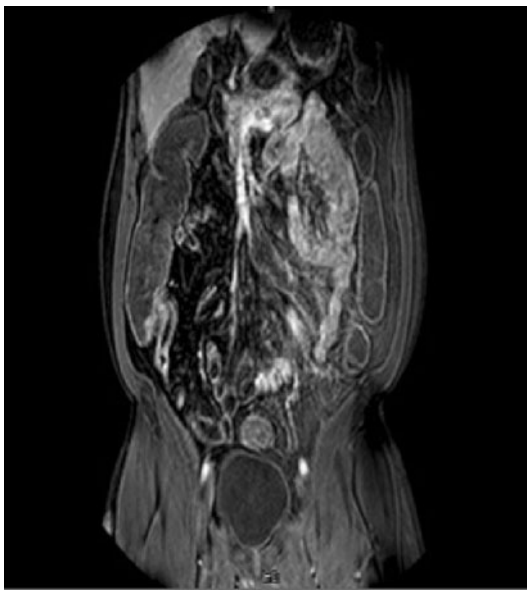

Fig. 11.5 Preoperative abdominal magnetic resonance

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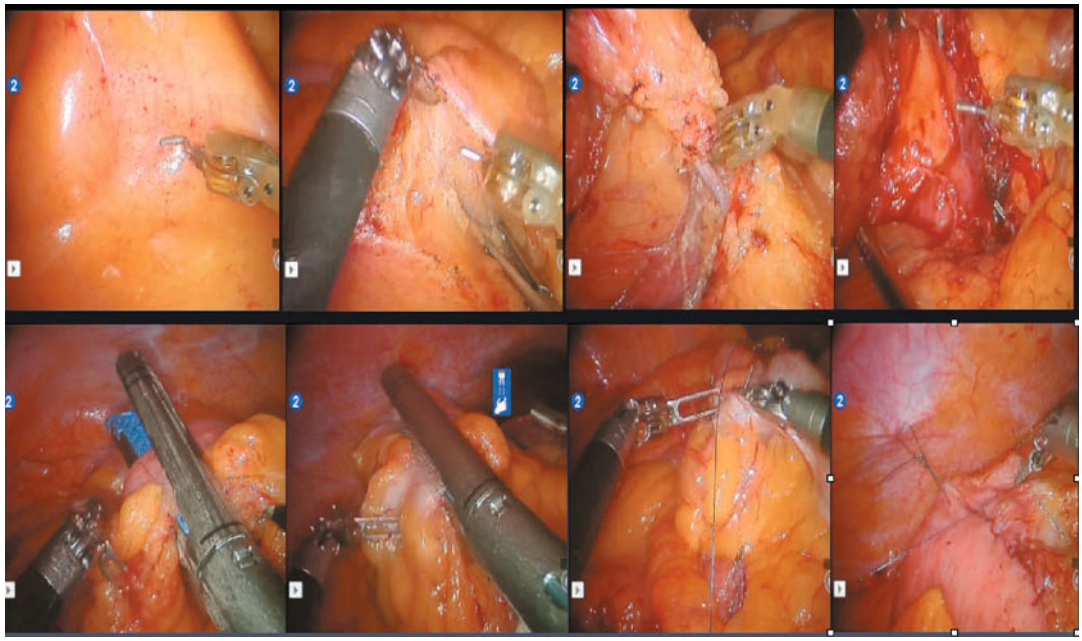

Fig. 11.6 Robotic right colectomy

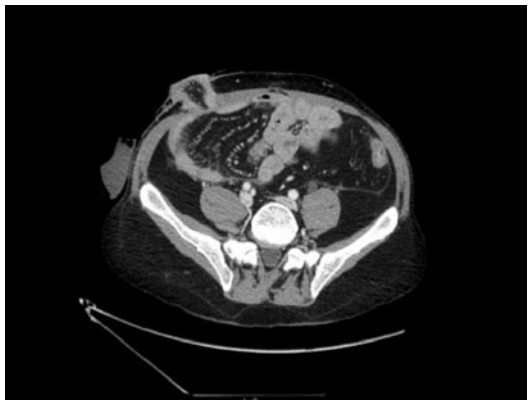

Fig. 11.7 Abdominal CT-scan on the 9th postoperative day (after the first operation)

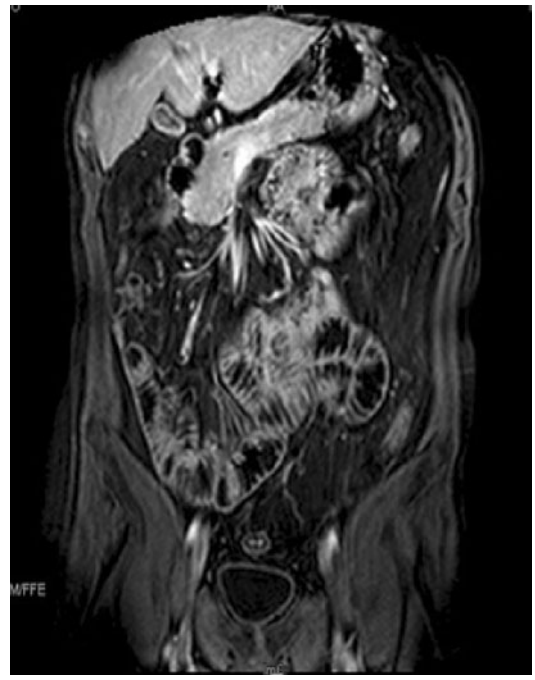

Fig. 11.8 Abdominal magnetic resonance at the end of the surgical procedures

grade dysplasia. On the second postoperative day, the patient had fever (38°) and the day after he developed an acute abdomen, with right lower quadrant peritonism and leakage of fecaloid material from the abdominal drainages. An emergency laparoscopy was performed. It revealed a fistulous tract located on the posterior wall of the colon, outside of the anastomosis, at the level of the lower end of the colonic stump. The conspicuous abdominal leakage and the overdistension of the transverse colon prompted us to perform

an open surgery with a new ileocolic anastomosis and a protective ileostomy in the right iliac fossa. In the early postoperative period, the patient had intermittent fever (up to 39°), with evidence of a small amount of pleural effusion at CT scan of the chest. After medical treatment with steroids, he was finally discharged. Five months later, after the integrity of the anastomosis had been evaluated and the presence of fistulas excluded, immediately after ileostomy closure, the patient had an early acute abdomen with evidence of pneumoperitoneum, as seen on the abdominal CT-scan. A second emergency laparotomy showed a normal ileocolic anastomosis with an abscess cavity and multiple fistulous tracts located proximally to the ileocolic anastomosis. A surgical toilette was then performed with a new lateral ileostomy. In the postoperative period, another perianastomotic abscess was evident at abdominal CT scan but it was successfully treated by medical therapy. The patient was discharged after one month and was disease free during one year of clinical follow-up.

Open Issues

Surgery still represents a fundamental aspect in the management of CD. In fact, 80% of the patients with CD undergo surgery at disease onset [5]. Many surgical procedures such as strictureplasties or intestinal resections are now available, even if they are rarely curative [6]. In the era of biologics, the request for surgery is still important, ranging between 25% and 33%, as opposed to 27% and 61% within 5 years after diagnosis during the period when biologics were not an option [7]. The rate of repeated surgery is still very high, accounting for one third of the patients with CD. In a recent review by the Korean IBD study group on the risk factors responsible for reoperation, it was shown that the different surgical techniques, such as strictureplasties or intestinal resections, did not influence the need for a second or third operation, unlike other reports [8, 9]. The introduction of minimally invasive procedures such as single port laparoscopic surgery or robotic surgery, seemed to guarantee minimum trauma for the patient, but the specific long-term complications of inflammatory bowel disease (IBD), such as recurrence of stenoses, are not likely to be influenced by the technique used in the primary operation [10] to access the abdomen. In particular robotic surgery allows an anastomosis to be made without the use of a stapler, as in open surgery (handsewn and double-layer sutures). The robotic approach was used in the treatment of our patient with a severe form of CD, who nonetheless received a handsewn anastomosis, in order to guarantee the best result possible. Still, the clinical course was complicated and the patient needed two more operations, confirming the unpredictable nature of the disease. It should be noted that the sites of the two relapses were distant from the anastomosis site. Moreover, the successful final course of the patient seems to support the use of robotic surgery for the treatment of IBD even if nowadays these surgical patients have complex disease and undergo surgery after many years of corticosteroid or immunosuppressive therapy or after serious complications arise.

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Multiple Choice Questionnaire

1) In what percentage of patients with severe forms of Crohn's disease is surgery needed?

a. 5–10%
b. 15–20%
c. 25–30 %
d. 40–50%
e. nearly 80 %

2) The reported reoperation rate at 5 years in Crohn's disease is

a. 1–3%
b. 5–10%
c. 10–15%
d. 20–25%
e. 40–45%

3) Which risk factors are thought to be responsible for the need for repeated surgery in Crohn's disease?

a. young age
b. stricturing behavior
c. intra-abdominal abscess
d. emergency surgery
e. all of the above

1.e – 2.d – 3.e

Histology

Case 12
The Histopathology Tribune: A Detailed Report on Gut Diseases

Vincenzo Villanacci, Stefania Manenti, Marina Yarygina, Maxim Untesco, Raffaele Manta and Gabrio Bassotti

Background

The following provides an overview of the different pathological situations that involve the terminal ileum and in which the histopathological examination plays a crucial role in the correct diagnosis. All the cases presented are based on a strict collaboration with other specialists, following the concept of a multidisciplinary approach.

The cases presented concern the following diseases:

- Endocrine tumors
- Crohn's disease
- Backwash ileitis and pouchitis
- Celiac disease
- Autoimmune enteropathy
- Drug damage
- Actinic Ileocolitis
- Vasculitis
- Infections

Endocrine Tumors

Clinical Presentation

Case 1

A 65-year-old man with abdominal discomfort in the right abdominal area and occasional episodes of diarrhea. Ultrasound examination revealed a thickened terminal ileum, raising the suspicion of Crohn's disease. Worsening of symptoms leading to occlusion mandated a CT scan, which revealed a nodular mass in the terminal ileum. The patient underwent laparoscopic resection of the terminal ileum and cecum. The histological examination revealed an endocrine tumor (Fig. 12.1).

Case 2

A 75-year-old woman with abdominal discomfort in the right abdominal area suspicious for inflammation of the appendix. An ileocolonoscopy showed nodules in the terminal ileum and ileocecal valve. Histology on biopsy samples revealed an endocrine tumor, confirmed after surgical resection (Fig. 12.2).

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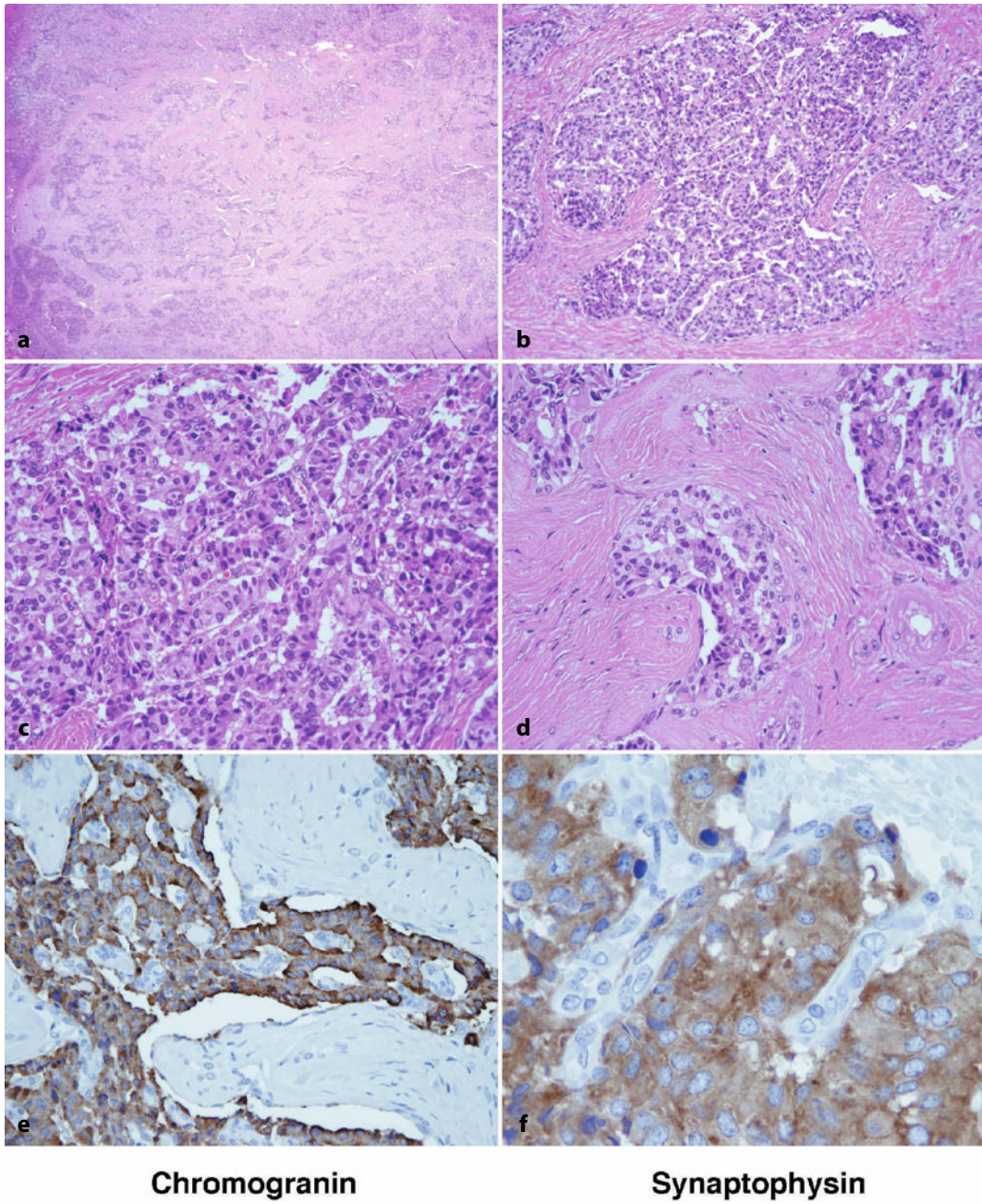

Fig. 12.1 Typical trabecular proliferation of endocrine cells. H&E **a** ×10, **b** ×20, **c** × 40 **d** ×40. **e**, **f** Immunohistochemistry with chromogranin and synaptophysin: **e** ×20, **f** ×40

Pape UF, Perren A, Niederle B, Gross D, Gress T, Costa F, Arnold R, Denecke T, Plöckinger U, Salazar R, Grossman A; Barcelona Consensus Conference participants (2012) ENETS Consensus Guidelines for the management of patients with neuroendocrine neoplasms from the jejuno-ileum and the appendix including goblet cell carcinomas. Neuroendocrinology 95(2):135-56

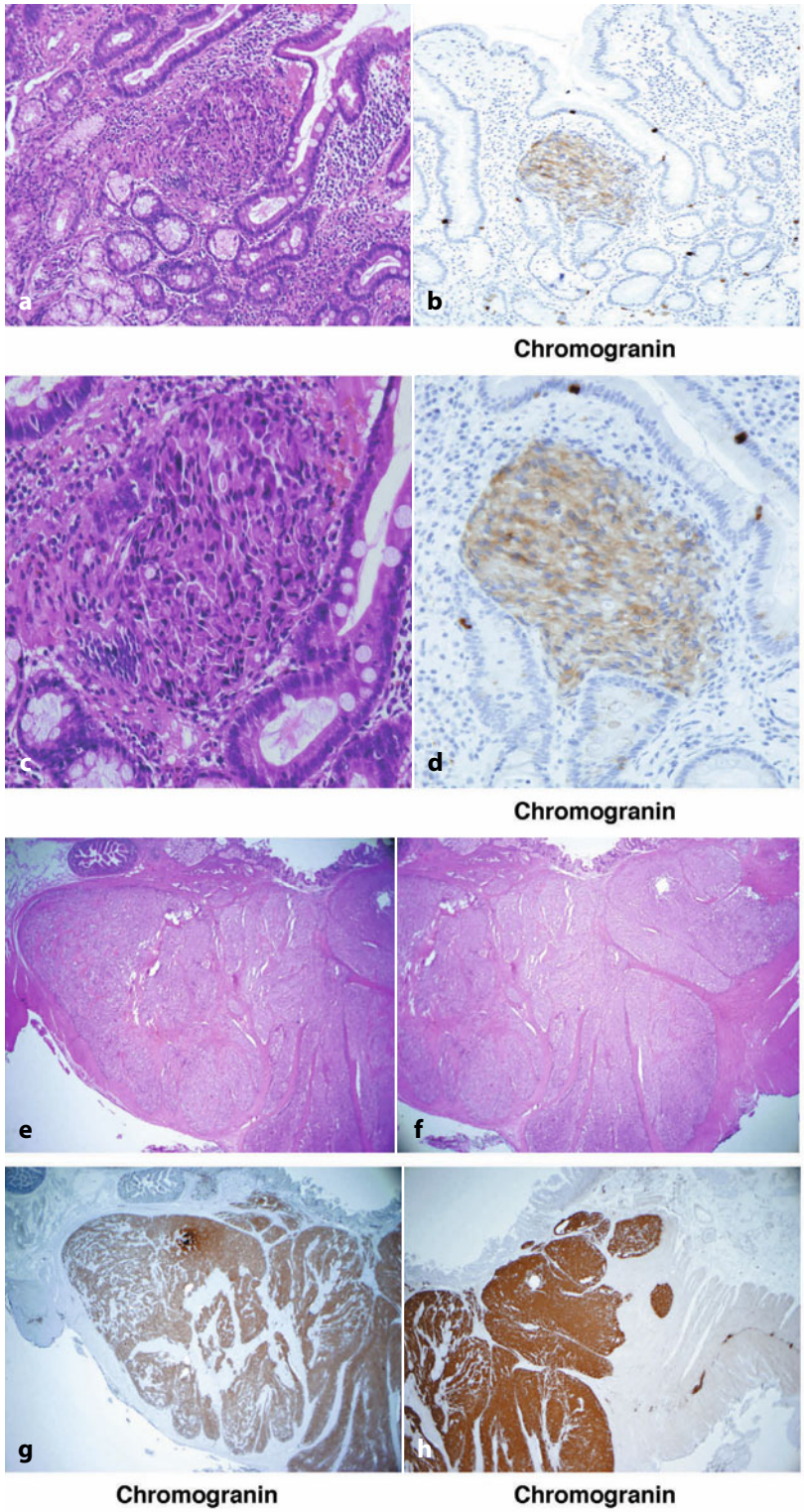

Fig. 12.2 a, c Ileal biopsies of small nodules of endocrine tumor. H&E **a** × 20, **c** × 40. **b, d** Immunohistochemistry with chromogranin ×20 (**b**), ×40 (**d**). **e, f** Surgical specimen with evidence of endocrine tumor; H&E ×5; **g, h** immunohistochemistry with chromogranin, ×5

Crohn's Disease

Clinical Presentation

An 8-year-old female with abdominal cramping and diarrhea. An ileocolonoscopy revealed aphthous ulcers in the right colon and terminal ileum. Histology on biopsies confirmed the diagnosis of Crohn's disease (Fig. 12.3).

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Backwash Ileitis and Pouchitis

Clinical Presentation

Case 1

An 18-year-old female with bloody diarrhea and abdominal pain of 3 months duration. Colonoscopy showed ulcerative pancolitis with hyperemia in the terminal ileum. Biopsies revealed a backwash ileitis (Fig. 12.4a, b).

Case 2

A 32-year-old male with a 12-year history of ulcerative colitis. He underwent surgery (total colectomy with pouch reconstruction) after developing toxic megacolon. Subsequently, he had diarrhea and abdominal pain. Endoscopy revealed pouchitis, confirmed by biopsy samples (Fig. 12.4c, d).

Suggested Reading

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Celiac Disease

Clinical Presentation

Case 1

A 22-year-old female with a family history positive for celiac disease (mother). Sideropenic anemia was determined and she complained of diarrhea and abdominal discomfort. At ileocolonoscopy, the colonic mucosa was normal but the terminal ileum revealed focal areas of hyperemia. Biopsies revealed low-grade atrophy of the villi and pathologically increased T lymphocytes (Fig. 12.5).

Case 2

A 56-year-old male who was diagnosed with celiac disease at the age of 42. During the last 2 years, he lacked a clinical and histological

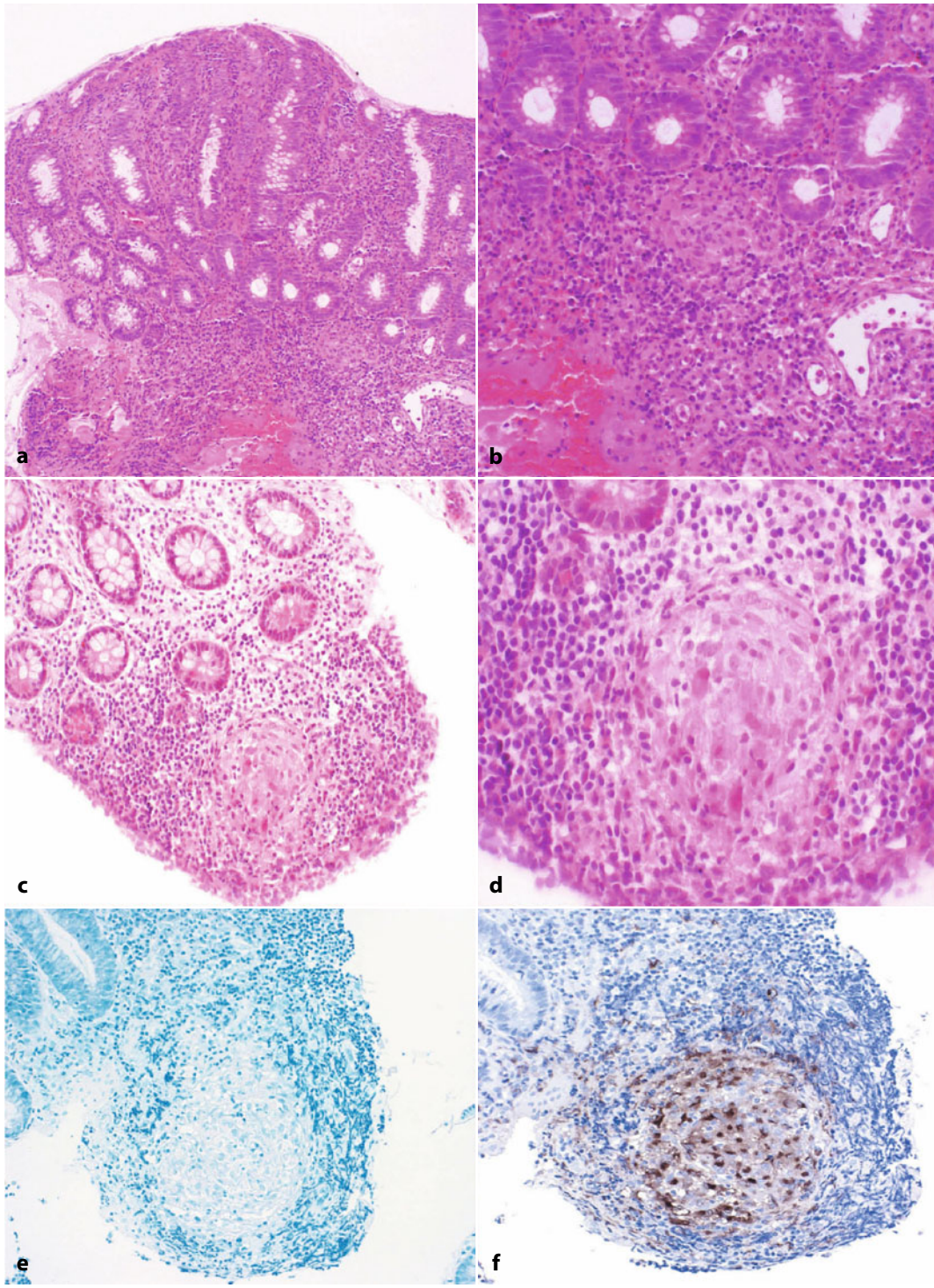

Fig. 12.3 a Inflammatory infiltration of the ileum with penetration of the submucosa; H&E ×10. **b–e** Typical granulomas: H&E **b**, **c** ×20, **d** ×40; **e** Ziehl-Neelsen ×20; **f** CD68 ×20

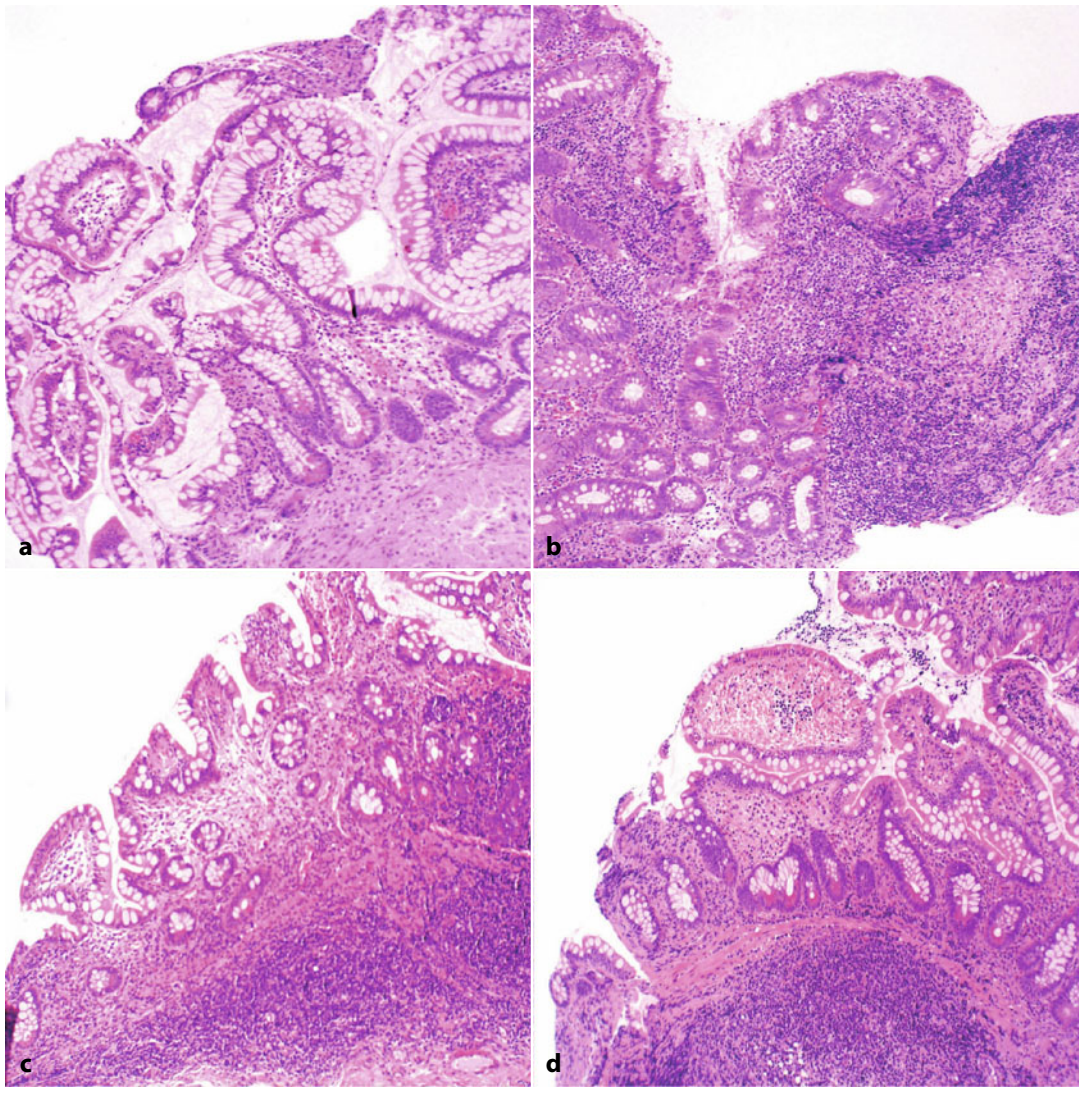

Fig. 12.4 a, **b** Inflammatory infiltrate of the ileum, with lymphoid follicles. Architectural distortion of the crypts and crypt abscess are both absent; H&E ×20. **c**, **d** Pouch inflammatory infiltrate with lymphoid follicles. No architectural distortion of crypts, superficial erosions; H&E ×10

response to treatment with a gluten-free diet (Fig. 12.6).

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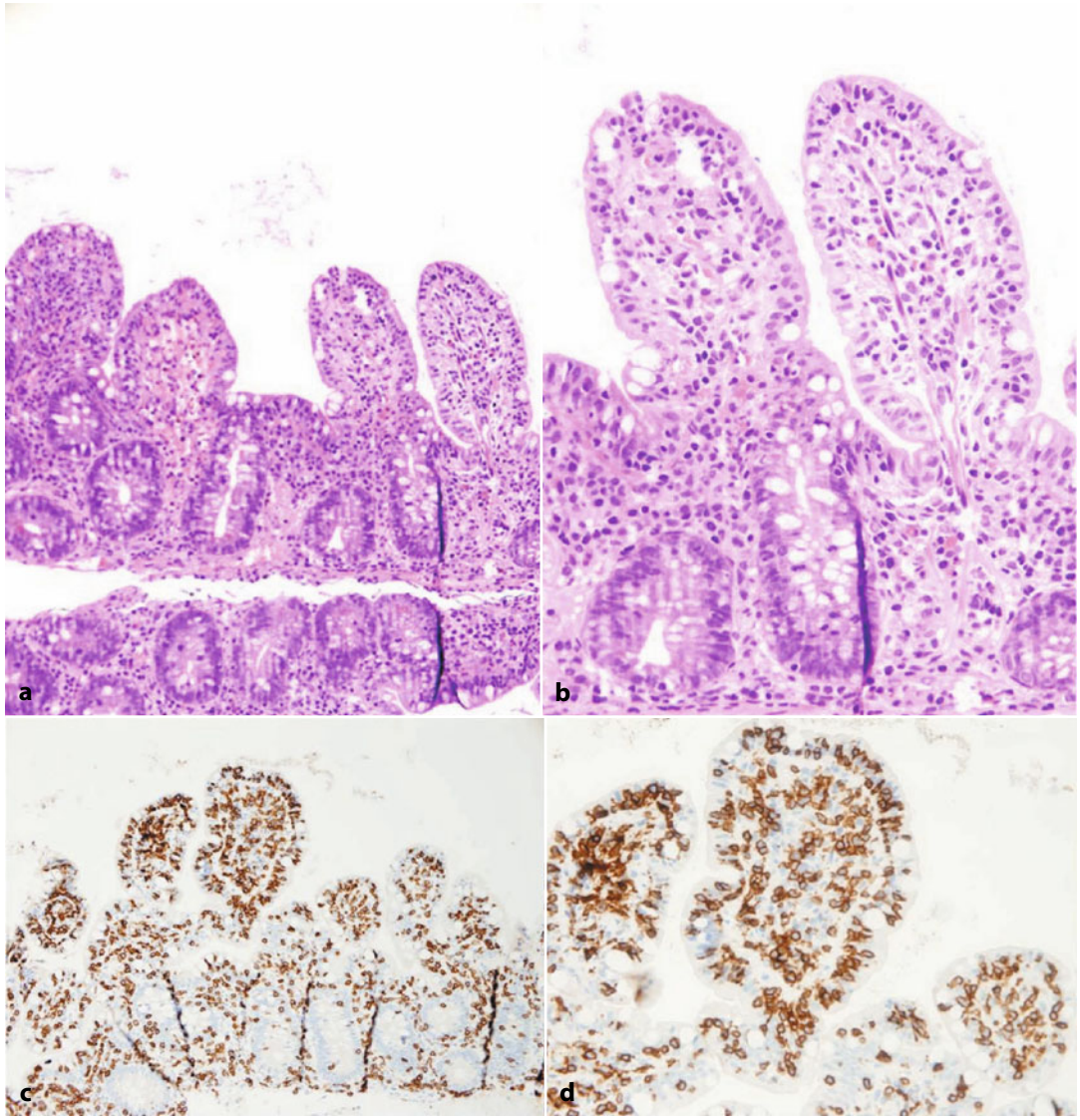

Fig. 12.5 **a**, **b** The terminal ileum shows low-grade atrophy of the villi; H&E **a** ×10, **b** ×20. **c**, **d** Pathological increase of T lymphocytes; immunostain for CD3 **c** ×10, **d** ×20

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Autoimmune Enteropathy

Clinical Presentation

A 3-year-old female with intractable diarrhea of 2 months. After common infective and

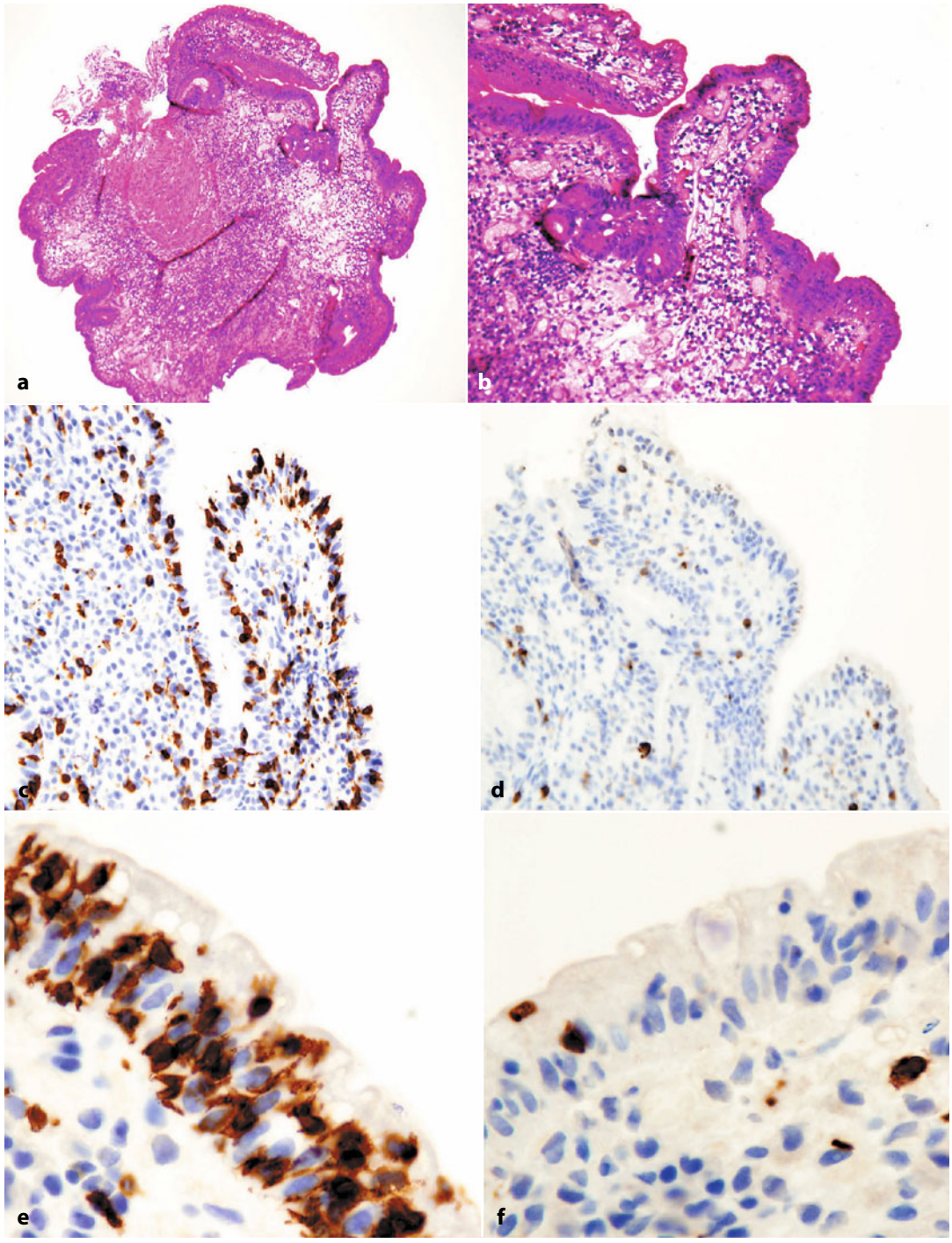

Fig. 12.6 **a**, **b** The terminal ileum shows low-grade atrophy of the villi; H&E **a** ×10, **b** ×20. **c** **e** T lymphocytes positive for immunostaining with CD3 **c** ×20, **e** x100. CD8-negative **d** ×20, **f** ×100

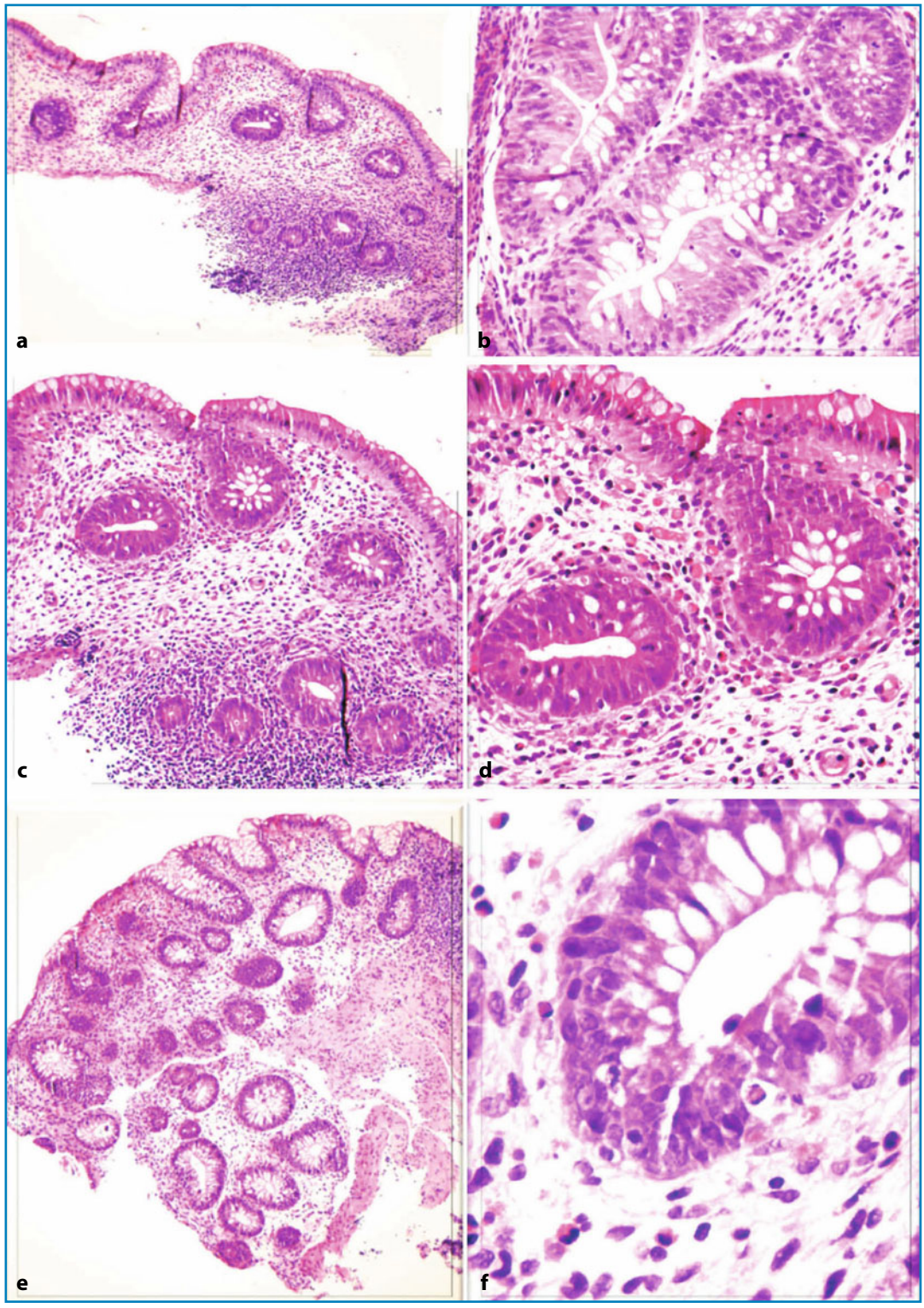

Fig. 12.7 a–f Marked atrophy and hyperplasia of the crypts; H&E **a** ×10, **b** ×40, **c** ×20, **d** x 40, **e** ×10. Apoptotic body in a crypt, **f** ×100

other diarrheal conditions were excluded, colonoscopy with biopsy sampling was carried out (Fig. 12.7).

Suggested Reading

Fenoglio Preiser C (2008) Gastrointestinal pathology. An atlas and text, 3rd edn. Lippincott Williams & Wilkins

Gentile NM, Murray JA, Pardi DS (2012) Autoimmune enteropathy: a review and update of clinical management. Curr Gastroenterol Rep 14:380-5

Greenson JK (2010) Diagnostic pathology gastrointestinal AMIRSYS

Drug Damage

Clinical Presentation

A 76-year-old female with a history of depression treated with sertraline. Due to her high cholesterol levels, she was also treated with cholestyramine. After a short period of treatment, bloody diarrhea with severe abdominal pain appeared. Based on her worsening clinical condition, she underwent surgery, with resection of 20 cm of small intestine (Fig. 12.8).

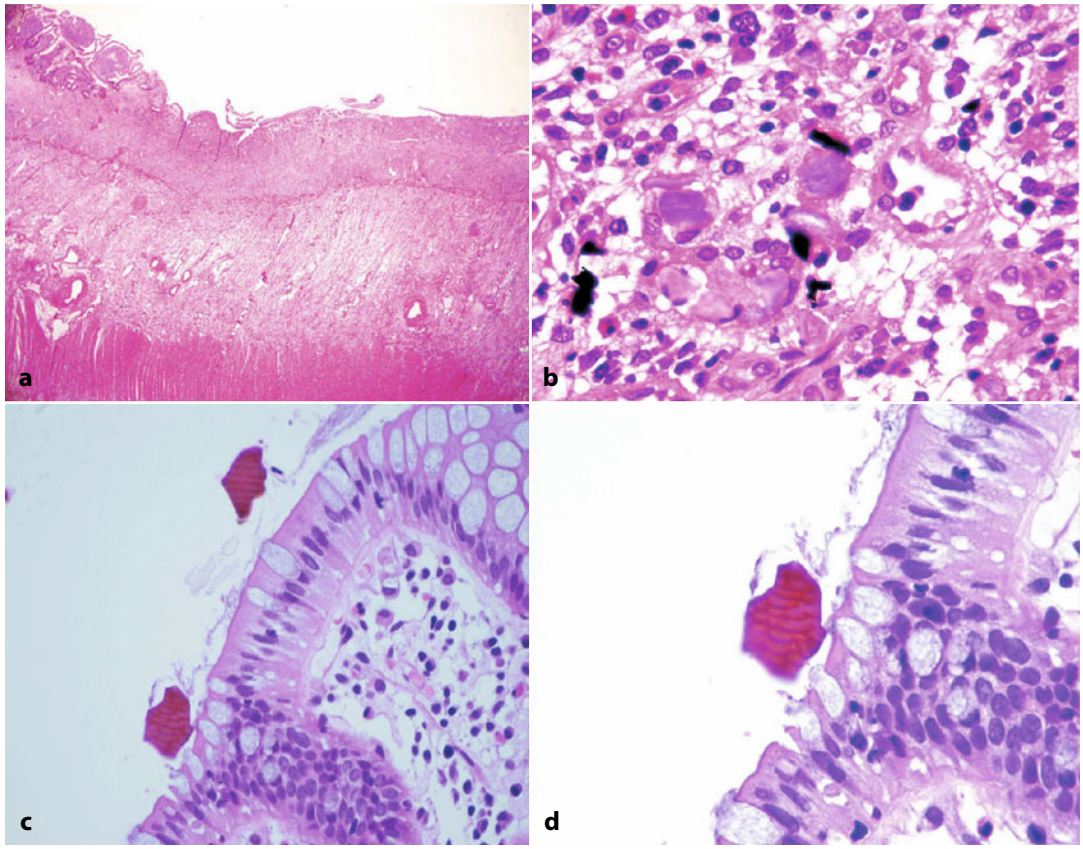

Fig. 12.8 Surgical specimen. **a** Diffuse coagulative necrosis of the small intestine; H&E ×10. **b** Questran crystals in the lamina propria; H&E ×40. **c**, **d** Questran crystals on the superficial epithelium H&E **c** ×20, **d** ×40

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Villanacci V, Casella G, Bassotti G (2011) The spectrum of drug-related colitides: important entities, though frequently overlooked. Dig Liver Dis 43(7):523-8

Actinic Ileocolitis

Clinical Presentation

A 66-year-old female with a history of ovarian tumor treated by radiotherapy. Thereafter, the patient developed diarrhea and mucosal prolapse in the rectum. Histological biopsies at ileocolonoscopy revealed actinic ileocolitis with fibrosis in the lamina propria and dilated vessels with hyaline walls (Fig. 12.9).

Suggested Reading

1. Greenson JK (2010) Diagnostic Pathology Gastrointestinal AMIRSYS
2. Fenoglio Preiser C (2008) Gastrointestinal Pathology. An atlas and text, 3 edn. Lippincott Williams & Wilkins

Vasculitis

Clinical Presentation

A 3-month-old male with bloody diarrhea. Laparoscopic biopsies revealed an ischemic mucosa of the small intestine involving a segment of 5 cm. Histology revealed the typical picture of vasculitis (Fig. 12.10).

Suggested Reading

Fenoglio Preiser C (2008) Gastrointestinal Pathology. An atlas and text, 3 edn. Lippincott Williams & Wilkins

Greenson JK (2010) Diagnostic Pathology Gastrointestinal AMIRSYS

Infections

Clinical Presentation

Case 1

A 24-year-old HIV-positive male with a history of bloody diarrhea. Ileocolonoscopy showed ulcers in the terminal ileum. At histology, biopsies revealed a *Mycobacterium avium intracellulare* infection (Fig. 12.11).

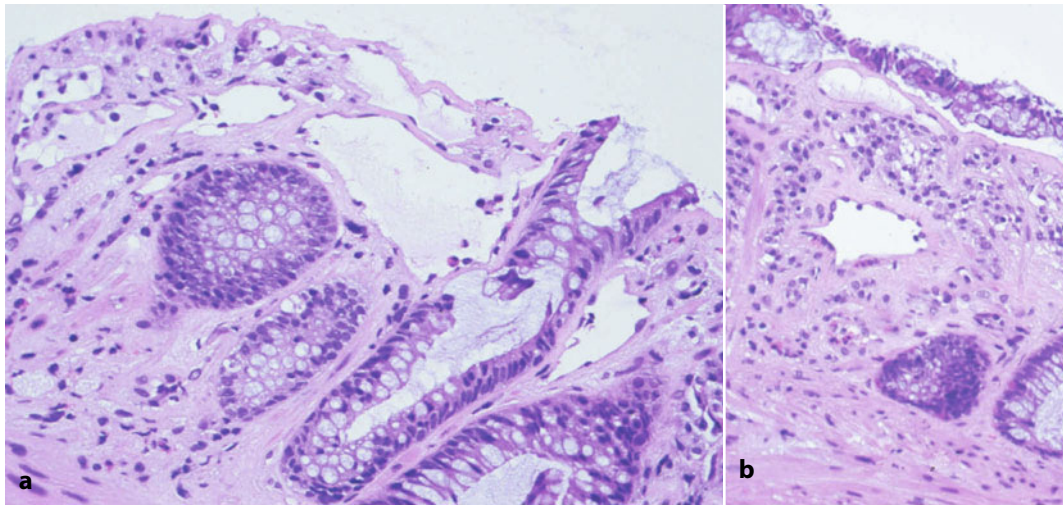

Fig. 12.9 a, **b** Diffuse fibrosis of the lamina propria and dilated vessels with hyaline walls; H&E × 20

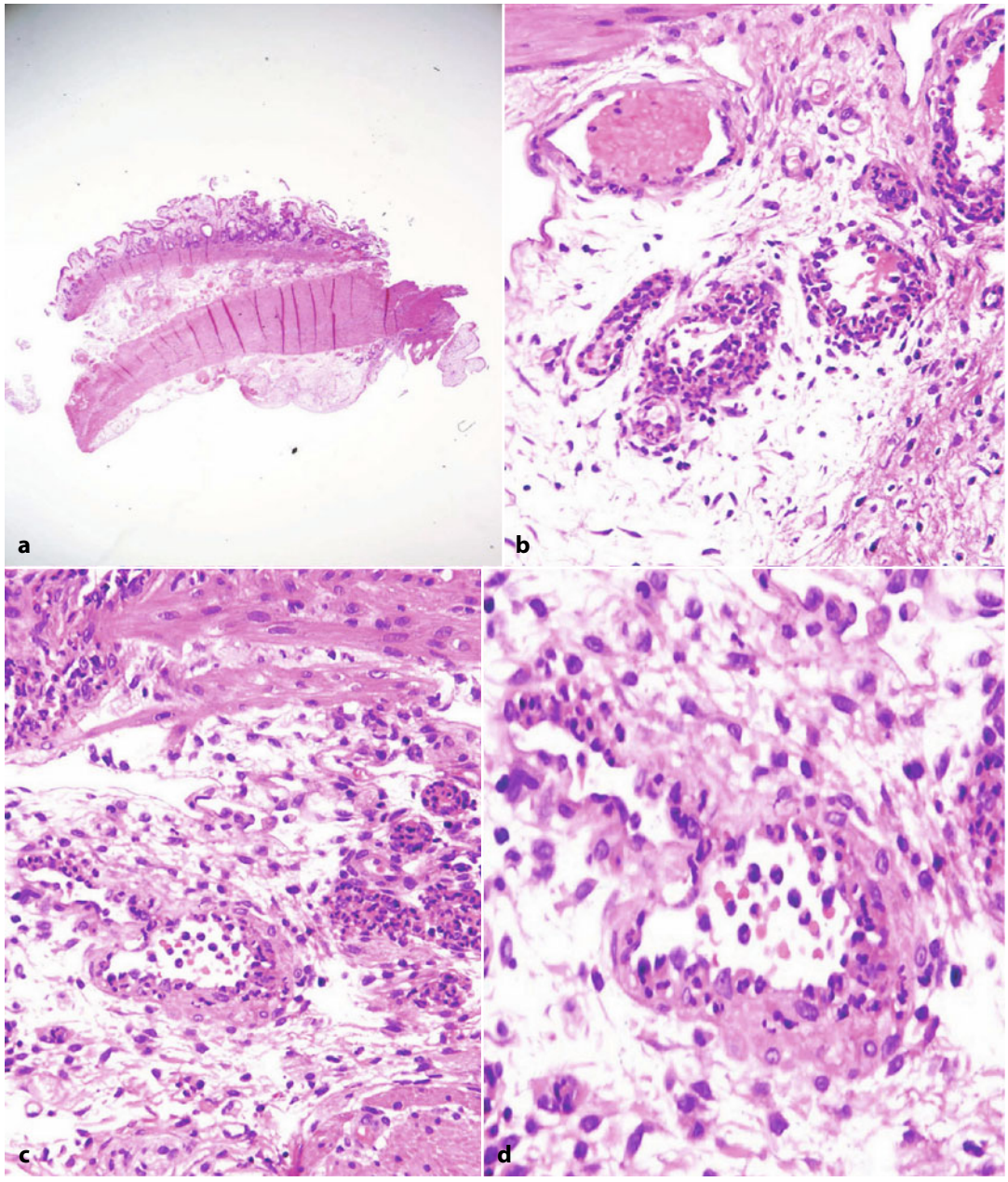

Fig. 12.10 Diffuse infiltration of the vessel walls of the submucosa by an inflammatory infiltrate composed of eosinophils. H&E **a** x5, **b**, **c** ×20 **d** ×40

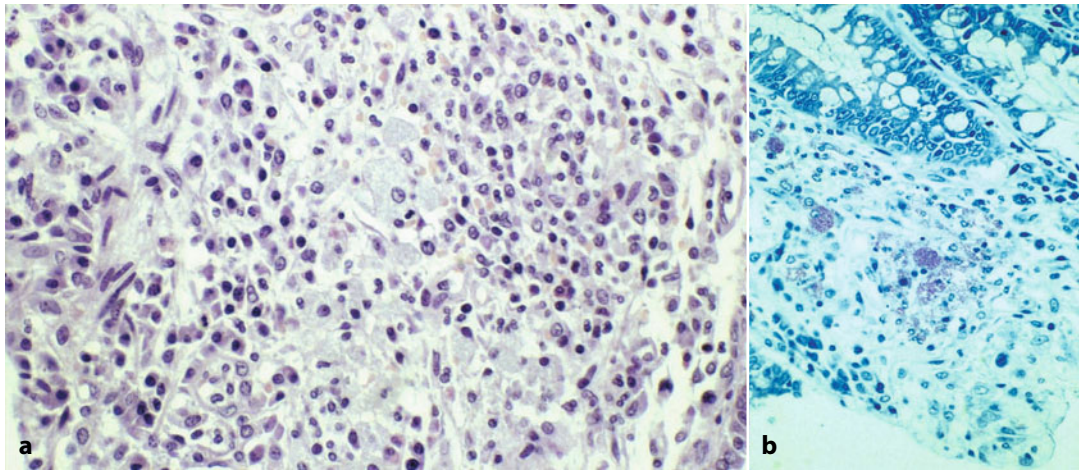

Fig. 12.11 a Diffuse inflammatory infiltrate of the lamina propria, showing a few histiocytes; H&E ×20. **b** Ziehl Neelsen stain revealed numerous bacilli; ×20

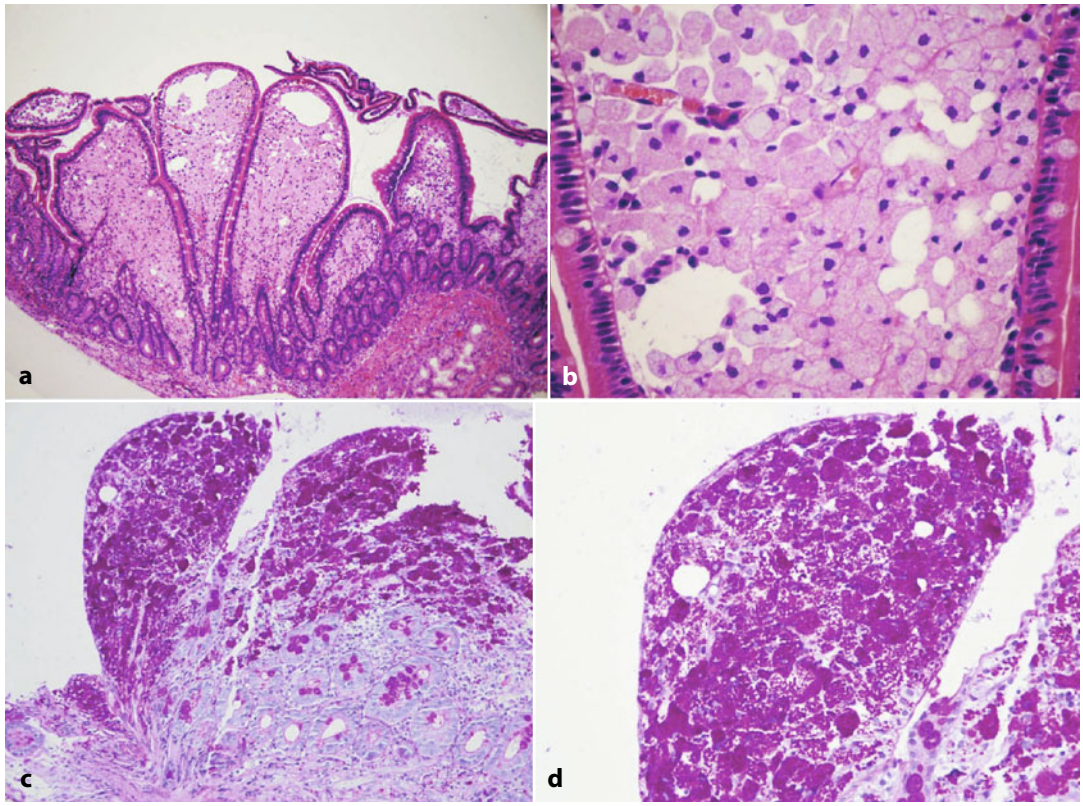

Fig. 12.12 a, **b** Diffuse infiltrate of the lamina propria by histiocytes, with occasional dilated lymphatic vessels; H&E **a** ×10, **b** ×40. **c**, **d** PAS diastase positivity of the histiocytes; **c** ×20, **d** ×40

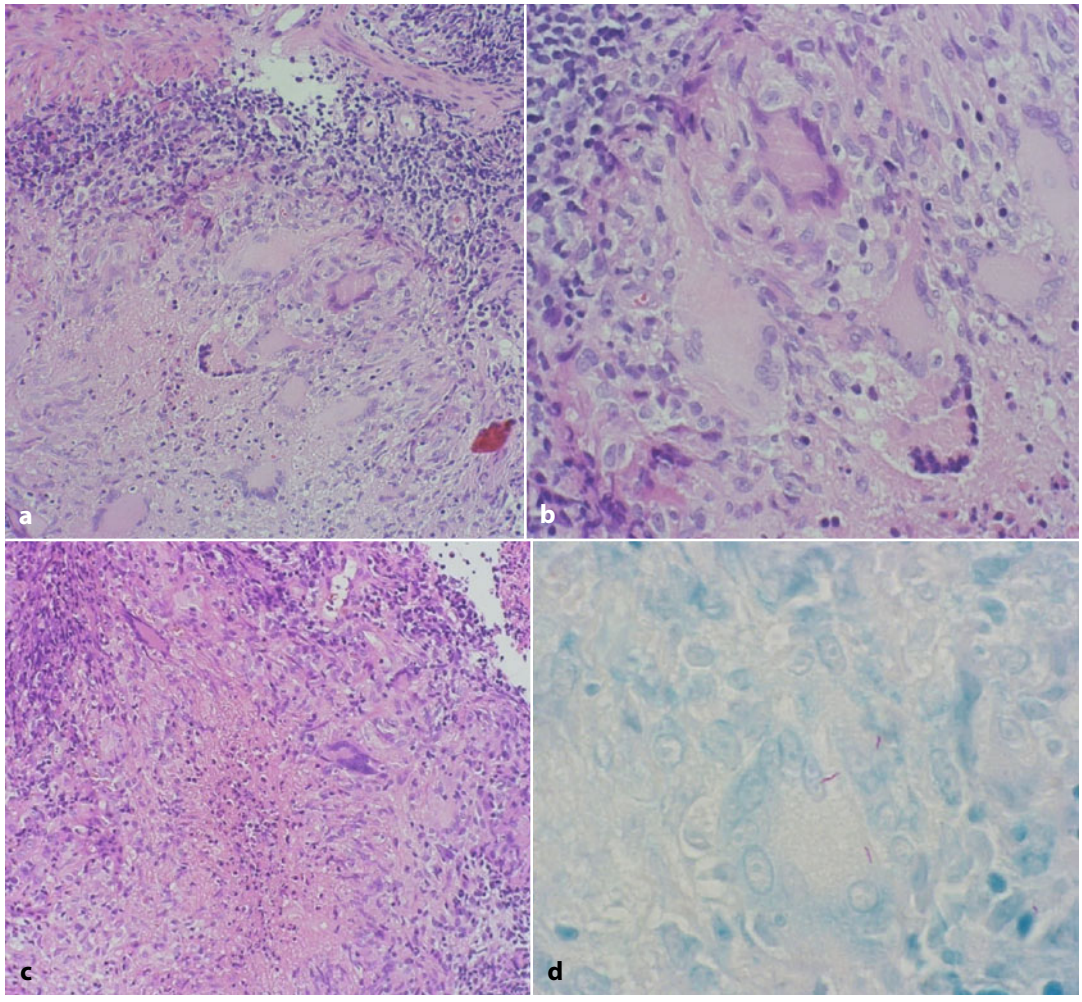

Fig. 12.13 a–c Numerous granulomas in the lamina propria; H&E **a** x 10, **b** ×40, **c** ×20. **d** Ziehl-Neelsen positivity for bacilli (alcohol/acid-fast); ×40

Case 2

A 54-year-old male with a history of diarrhea. Ileocolonoscopy showed villous atrophy in the terminal ileum. At histology, biopsies revealed the typical features of Whipple disease (Fig. 12.12).

Case 3

A 44-year-old male with a history of diarrhea. Ileocolonoscopy showed a nodular mass suspicious of tumor in the cecum. On histology, a granuloma positive for Ziehl-Neelsen staining for bacilli (alcohol/acid-fast) was determined. The diagnosis was tuberculosis (Fig. 12.13).

Suggested Reading

Fenoglio Preiser C (2008) Gastrointestinal Pathology. An atlas and text, 3 edn. Lippincott Williams & Wilkins

Greenson JK (2010) Diagnostic Pathology Gastrointestinal AMIRSYS

Miscellaneous

Case 13.1
A Fat Jejunum

Gianfranco Tappero

Background

Submucosal tumors are rare endoscopic findings and their diagnostic and therapeutic management is a subject of debate [1]. Neoplastic disease in the gut accounts for 0.4% of all digestive malignancies and is mainly represented by carcinoid tumors [2]. Benign diseases are even rarer and their treatment is largely unexplored. This is especially the case for lipomatous neoplasms, which have a very low malignant potential. The two most common sites of lipomas are the subcutaneous tissue and/or the subserosa of the intestinal tract [3]. Herein we present a very interesting case of this rare condition.

Clinical Presentation

A 73-year-old man came to our emergency department because of sudden vomiting, followed by overt hematemesis and melena. Blood tests showed severe anemia (Hb 6.6 gr/dl) with hypovolemic syndrome. After hemodynamic stabilization, he underwent urgent upper endoscopy, which revealed a long, yellowish, pedunculated submucosal tumor in the proximal jejunum, with a linear breakage at the top and active bleeding (Figs. 13.1, 13.2). The bleeding was immediately stopped by injecting diluted adrenaline 1:10,000 at the base of the tumor, which was then removed by electrosurgical snare polypectomy, without complications (Fig. 13.3). The specimen showed a smooth, soft and elastic surface (cushion sign), while yellow fat tissue oozed from the base (Figs. 13.4, 13.5). The tumor measured 72 × 20 mm. A definitive diagnosis of lipoma was based on the histopathology, which revealed a typical enteric mucosa surrounding an adipose submucosal mass filled with blood vessels (Fig. 13.6). The patient had no further episodes of gastrointestinal hemorrhage and rapidly recovered, as confirmed at follow-ups of 1 month and 6 years.

Open Issues

Lipomas of the colon are rare and usually present in older individuals. Normally, these tumors are benign and slowly growing. They are either incidentally found at endoscopy or because they become evident by gastrointestinal bleeding, anemia, intussusception, and bowel obstruction. Endoscopy allows their treatment via different modalities, although many of these tumors are

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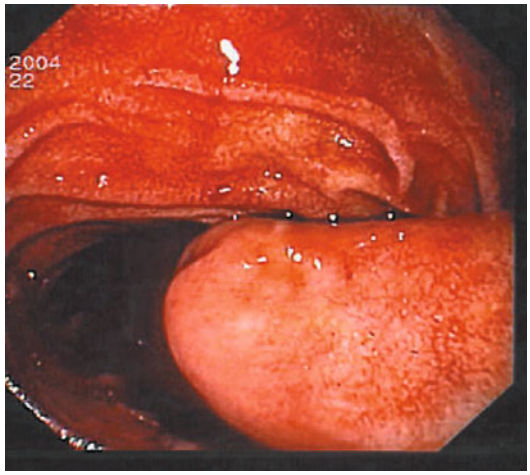

Fig. 13.1 Endoscopic view of a large pedunculated lipoma with evident bleeding

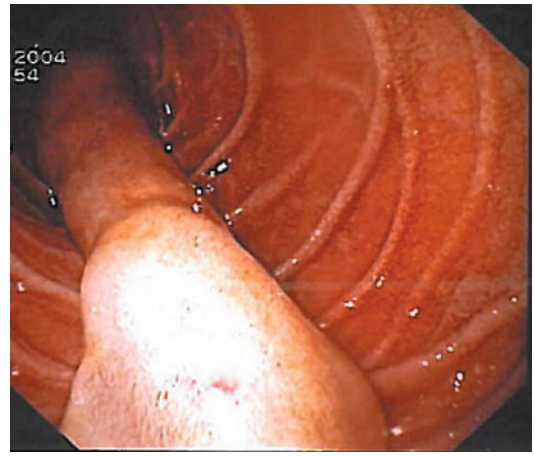

Fig. 13.2 Endoscopic view of the long pedunculated lipoma after the hemostatic injection of diluted Adrenalin at the base, with stop bleeding

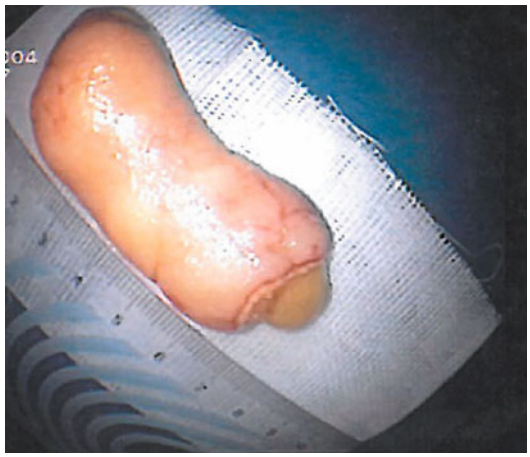

Fig. 13.3 The lipoma tissue was completely removed

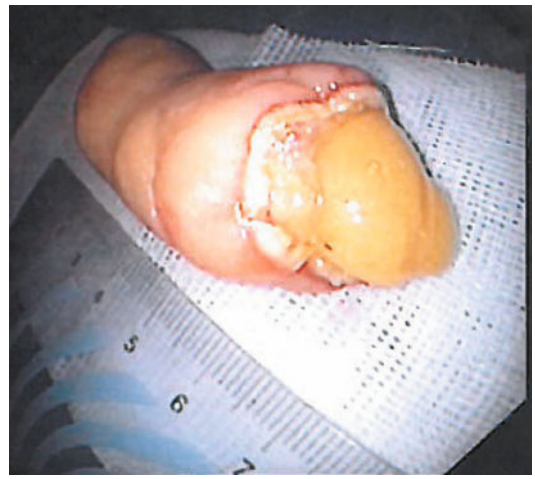

Fig. 13.4 The lipoma tissue was completely removed

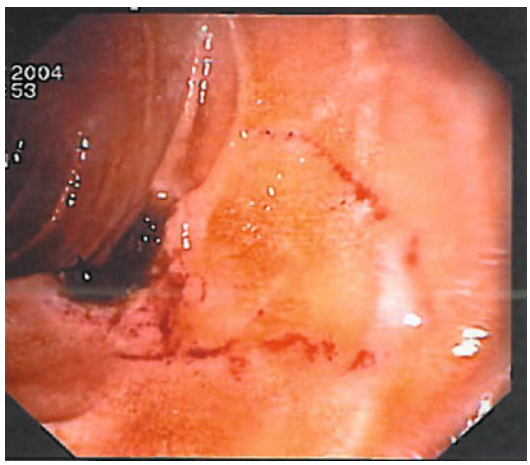

Fig. 13.5 Endoscopic view of the iatrogenic scar after resection

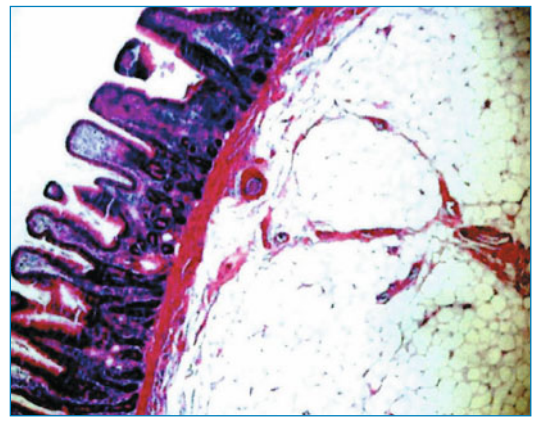

Fig. 13.6 Histological examination of the resected specimen: lipomatous tissue was clearly visible

resected surgically due to the risk of complications such as perforation and bleeding during therapeutic endoscopy [4, 5]. Our diagnosis of lipoma was at emergency endoscopy, performed due to the patient's severe bleeding, with immediate treatment. The tumor was successfully removed and subsequently confirmed on histopathologically to be a lipoma. After endoscopic removal, there were no complications. Our case seems to confirm that endoscopic treatment even of large lipomas can be safely performed with a high rate of success.

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Multiple Choice Questionnaire

1) Which is the incidence of small bowel submucosal tumors?
a. 0.4%
b. 1%
c. 2%
d. 3%

2) Which are the most common sites of lipomas?
a. submucosal tissue and subserosa of the intestine
b. colonic mucosa
c. abdominal wall

3) How many centimeters of terminal ileum can be visualized by ileoscopy?
a. no, never
b. no, only in case of larger lesions or in case of obstructive symptoms
c. yes, always

1.a – 2.a – 3.c

Case 13.2
A Rare Cause of Gastrointestinal Bleeding: A Paraprosthetic Aortojejunal Fistula

Paolo Trentino, Fabio Baldi and Sergio Coda

Background

Secondary aorto-enteric fistula (SAEF) is a rare but serious complication after the surgical or endovascular treatment of abdominal aortic aneurysm (AAA). Multiple mechanisms lead to SAEF formation. Infection and close adhesion between the graft and the bowel are considered the main predisposing factors. Graft infection occurs in 0.3–3.1% of AAA repairs, while 20–45% of infected patients will develop SAEF [1, 2].

Many diagnostic tests are available for SAEF, but they are not specific enough to universally diagnose the disease.

Herein, a rare case of paraprosthetic aortojejunal fistula is reported.

Clinical Presentation

In May 2010, a 65-year-old man was admitted as an emergency case at our institution because of lumbar pain and lower intermittent gastrointestinal bleeding. His past medical history revealed a previous duodenal ulcer and an infrarenal aorto-bi-iliac graft operation performed in 2003 because of an aneurysmal disease. At admission, his hemoglobin (Hb) was 8.4 g/dl, and due to recurrent melena and episodes of rectal bleeding, with bright red blood, he required red blood cell transfusions. An upper endoscopy up to the fourth duodenal portion was performed as the first diagnostic test, but it was negative. A colonoscopy with terminal ileal intubation revealed only slight bleeding coming from the upper portion of the GI tract.

A CT scan revealed a proximal aortic pseudoaneurysm at the level of the proximal graft anastomosis, with close adhesion of a jejunal loop. SAEF was suspected.

Following the CT scan results and based on the recurrent bleeding episodes, a **push enteroscopy** was performed. The jejunum was explored for about 50 cm distal to the ligament of Treitz. At this level, a decubitus of a small portion of the graft was identified in the jejunal lumen (Fig. 13.7).

At laparotomy, the pseudoaneurysm and the adhesion between the proximal graft anastomosis and a jejunal loop were identified (Fig. 13.8). The adhesion was carefully dissected, and the paraprosthetic aortojejunal decubitus was identified. A jejunal resection with an end to end jejunal anastomosis was performed (Fig. 13.9). The greater omentum

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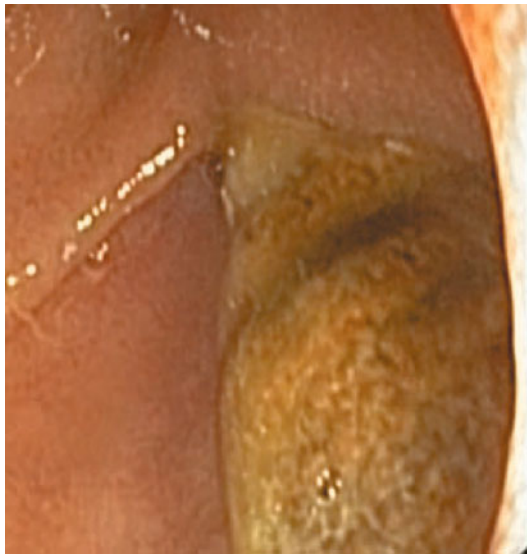

Fig. 13.7 Push enteroscopy: decubitus of the aortic graft in the jejunal lumen

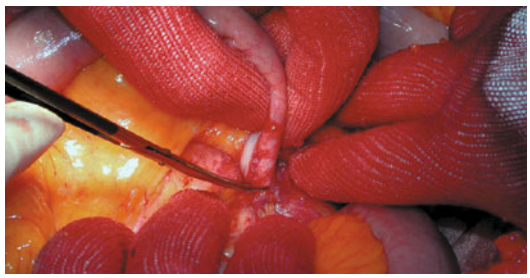

Fig. 13.8 Intraoperative finding: close adhesion of a jejunal loop to the aortic graft

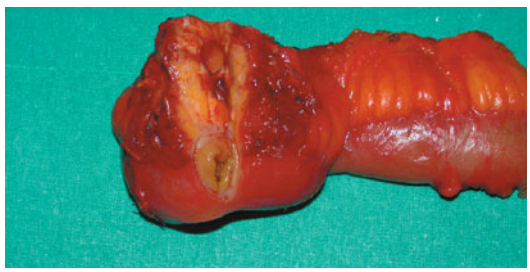

Fig. 13.9 Operative specimen: resected pseudoaneurysm and jejunal fistula

was interposed between the jejunum and the graft. The patient was then placed on a 2 week course of IV antibiotics and the postoperative course was uneventful. At 2 years follow-up, he is well, without any recurrence of the disease.

Open Issues

SAEF is a rare but serious complication after reconstructive therapy for aortic diseases. According to Vollmar and Kogel's classification [3], SAEF may occur as a true fistula between the aortic graft lumen and the enteric lumen (true SAEF) or as an ulceration of the bowel wall caused by chronic decubitus on the aortic graft (paraprosthetic SAEF). Gastrointestinal bleeding may be life threatening in the case of a true SAEF.

The pathogenesis of SAEF is multifactorial; local infection, bowel injury during surgical dissection, anastomotic pseudoaneurysm, and fibrous contact between the graft and the bowel have been often described.

CT scan and endoscopy are the most reliable tests for the early diagnosis. CT scan shows mainly indirect signs of SAEF (direct contact between the graft and the bowel, extravasation of fluids, pseudoaneurysm, air), while endoscopy is mandatory up to the fourth portion of the duodenum. In fact, 70–81% of SAEFs are localized in the distal portions of the duodenum, as the proximal graft anastomosis is mainly involved in the SAEF. If traditional endoscopy is negative, capsule endoscopy should be considered [6].

Unfortunately, however, these tests are not sufficiently specific to universally indicate a diagnosis. A reasonable suspicion of SAEF should therefore arise in every patient treated for aortic disease, as this may help to determine the appropriate tests in order to obtain the correct diagnosis.

SAEF is less commonly found in the jejunum (9–12%), ileum (6–7%), and colon (4–8%) [4, 5]; in these cases an enteroscopy may be preferable.

Early confirmation of both the presence of the fistula and its location may shorten the operative time and allow a tailored surgical procedure, therefore reducing postoperative morbidity and mortality.

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Multiple Choice Questionnaire

1) The diagnostic tests for gastrointestinal bleeding in patients already treated for aortic disease are

a. upper and lower GI endoscopy
b. CT scan followed by extended upper and lower GI endoscopy
c. CT scan
d. capsule endoscopy
e. upper and lower GI endoscopy + capsule endoscopy

2) A patient with a previous history of treated aortic disease complains of fever and lumbar pain. You prescribe

a. antibiotics therapy
b. analgesic and antibiotics therapy
c. CT scan followed by digestive endoscopy
d. digestive endoscopy
e. CT scan only

3) Which is the most probable pathogenesis of Secondary Aorto-Enteric Fistula (SAEF)?

a. local infection
b. bowel injury during surgical dissection
c. anastomotic pseudoaneurysm
d. fibrous contact between the graft and the bowel
e. all of them

1.b – 2.c – 3.e

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