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Gastrointestinal Radiology

Primary Ileal Adenocarcinoma Simulating Crohn's Disease

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Abstract. An unusual case of primary adenocarcinoma of the terminal ileum with a clinical and radiographic appearance simulating Crohn's disease is presented. This example further supports the concept that terminal ileal disease should not immediately be equated with regional enteritis.

Key words: Crohn's disease, simulators – Ileum, adenocarcinoma.

Primary adenocarcinoma of the small intestine is a very uncommon tumor. Crucial in its detection is a high index of suspicion. In recent years the diagnosis of Crohn's disease has assumed considerable popularity. Physicians have become aware of many diseases which Crohn's disease may mimic; however, the converse has not been true. An unusual case of primary adenocarcinoma of the terminal ileum with a clinical and radiographic appearance simulating Crohn's disease is presented.

Case Report

A 64-year-old white male presented to his private physician in June 1978 with the acute onset of postprandial vomiting associated with 6 to 8 watery bowel movements per day. There was no prior history of gastrointestinal disease, and no evidence of gastrointestinal bleeding. Physical examination was unremarkable and the patient was presumed to have acute viral gastroenteritis. Because of persistent symptoms, the patient was hospitalized. Sigmoidoscopy with biopsy was unremarkable, but an upper gastrointestinal series with small bowel follow-through (Fig. 1) revealed a long segment of irregular narrowing in the terminal ileum with probable sinus tracts (arrowhead) and cecal compression felt to be consistent with Crohn's disease. The patient was treated with steroids and sulfasalazine with minimal response. He was discharged, but had multiple readmissions for small bowel obstruction.

In September 1978 he was admitted to the Northport Veterans Administration Medical Center with a right lower quadrant abdominal mass and pain. He suffered three episodes of small bowel obstruction, with one episode complicated by septic shock. Vigorous antibiotic, steroid, and decompression therapy was instituted. The patient then developed a right pulmonary embolism and was treated with heparin. Steroids were tapered and then discontinued. Hyperalimentation was begun because of deteriorated nutritional status and inability to feed orally. *Serratia marcescens* sepsis complicated hyperalimentation therapy. Gentamicin was begun and the patient was stabilized.

On the 89th hospital day, a laparotomy was performed. The terminal 20 cm of ileum appeared chronically inflamed and thickened and felt fibrotic and hard. Because of the appearance of this area and the fact that the remainder of the abdomen appeared normal, the surgeon was convinced that this represented Crohn's disease and he performed an ileocecal resection involving 40 cm of bowel with an end-to-end anastomosis. Gross pathological examination (Fig. 2) revealed bowel wall thickening to 1 cm in depth with bosselated, thickened mucosa and no prominent ulceration, beginning 26 cm from the proximal margin and extending to, but not beyond, the ileocecal valve. Microscopically (Fig. 3), there was deep penetration to the serosal fat by poorly formed malignant glands composed of highly anaplastic polygonal cells, many of which were "signet ring" types. Abundant mucin production by the cells was shown by both PAS and mucicarmine stains. Regional lymph nodes and the resection margins were free of disease; however, there was tumor extension into the periappendiceal fat. Bowel wall fibrosis, granulomas, and lymphoid infiltrates were conspicuously absent. Thus there was no evidence of Crohn's disease and the few lymphoid aggregates seen were believed secondary to the tumor.

Six months after surgery, the patient returned with a 1-month history of increasingly severe nausea and vomiting. Physical examination revealed a right lower quadrant abdominal mass and abdominal X-rays showed a distal small bowel obstruction. At exploratory laparotomy, tumor recurrence was found at the anastomosis as well as in multiple small bowel and mesenteric implants.

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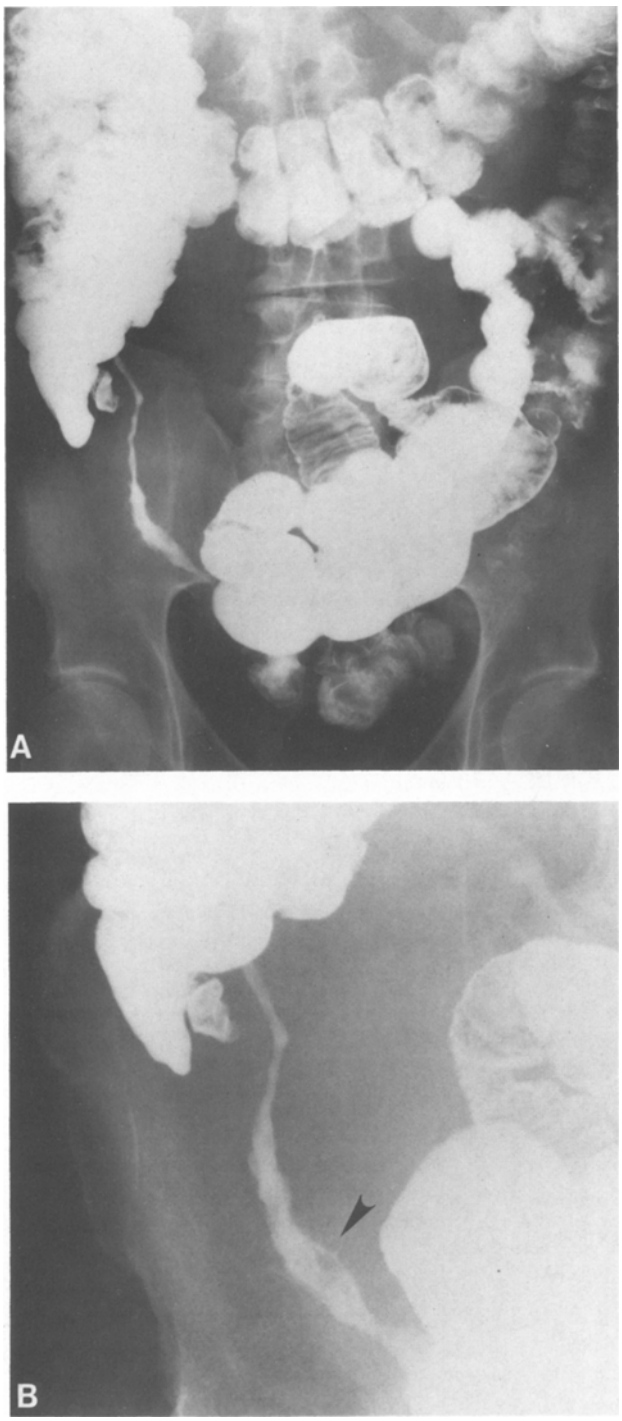

Fig. 1 A Small bowel series delayed film showing a long segment of irregular narrowing in the terminal ileum with extrinsic compression on the medial aspect of the cecum. **B** Enlarged view of ileocecal region also reveals probable sinus tracts (*arrowhead*).

Discussion

Neoplasms are relatively rare in the small intestine. The incidence of benign and malignant tumors as well as the distribution of tumors vary depending on whether a clinical, surgical, or autopsy series is considered [1]. Primary adenocarcinoma of the ileum occurs with equal frequency in males and females, and its peak incidence is in the seventh decade [1]. The most common signs and symptoms of small bowel tumors include obstruction, hemorrhage, a palpable abdominal mass, and perforation [2]. Symptoms are far more common in patients with malignant tumors, who most commonly complain of abdominal pain and weight loss [3]. Bridge and Perzin [1] found that in patients with primary adenocarcinoma of the ileum partial or complete small bowel obstruction was the most common (82%) presenting symptom. In addition, 73% complained of lower abdominal pain which was usually crampy and occasionally postprandial.

In the review by Bridge and Perzin [1], 9 of the 11 patients with ileal adenocarcinoma had intestinal roentgenograms showing either obstruction or mucosal changes suggesting tumor. To our knowledge, no patient with primary ileal adenocarcinoma has been previously described with radiographic changes simulating stenosing Crohn's disease of the terminal ileum. Our patient's clinical and radiographic presentation emphasizes the ability of this rare small intestinal tumor to mimic the more common inflammatory bowel disease. Chang et al. [4] recently presented six cases with different disorders (endometriosis, adhesions, pancreatic pseudocyst, carcinoid tumor, and intra-abdominal metastases) mimicking regional enteritis, and enumerated other considerations in the radiographic differential diagnosis of Crohn's disease — including lymphosarcoma, ischemic enteritis, radiation enteritis, tuberculosis, actinomycosis, Meckel's diverticulitis, typhoid, perforated cecal carcinoma, anisakiasis, and yersiniosis. They stressed that terminal ileal disease should not immediately be equated with regional enteritis. This case reiterates the importance of this concept.

Although obstruction is a frequently encountered complication of Crohn's disease, most patients present with recurrent episodes over many years. In this patient, with no prior history of inflammatory bowel disease, repeated episodes of obstruction over a relatively short period of time should have made us more suspicious of small bowel tumor.

Radiographically, the long area of irregular narrowing as well as the evidence of probably small sinus tracts were most suggestive of Crohn's disease. Although no sinus tracts were found at pathologic evaluation, it should be noted that 4 months had elapsed since radiographic examination. Even though 38 cases of adenocarcinoma complicating Crohn's disease have been reported [5], there was no evidence of Crohn's disease on pathological examination of this case.

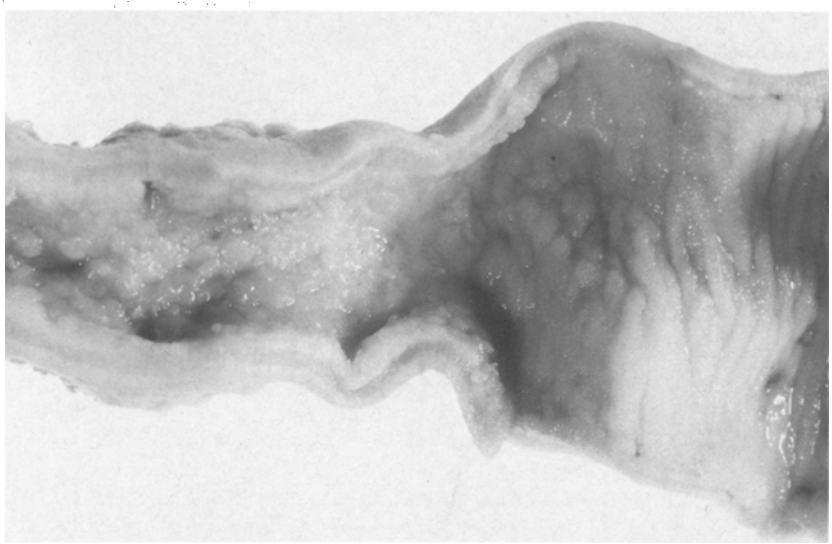

Fig. 2. Gross pathological specimen shows prominent submucosal thickening, mamillated mucosal surface, and luminal narrowing. Findings simulate those of Crohn's disease

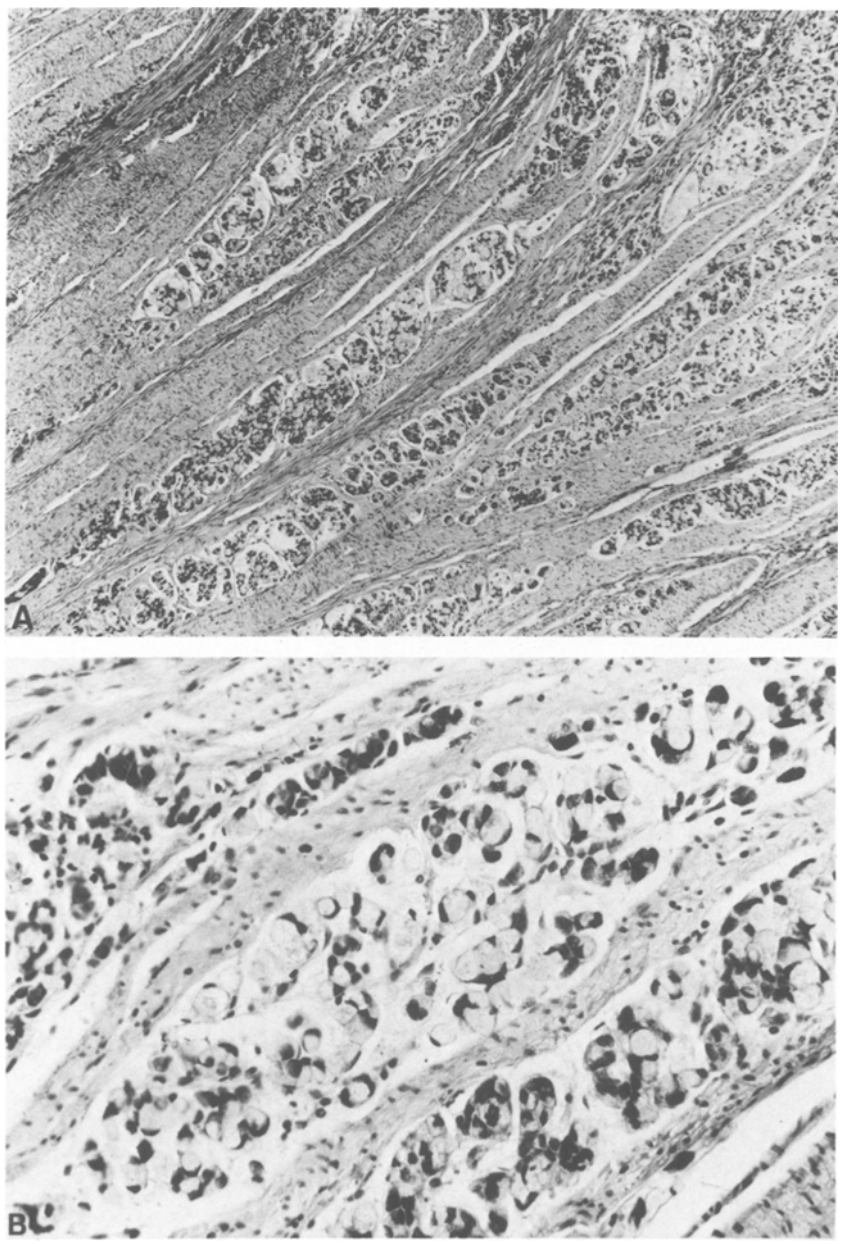

Fig. 3. A Photomicrograph (46×) showing tumor diffusely infiltrating the muscularis. **B** Photomicrograph (117×) showing the mucin compressing the nuclei to form a signet ring pattern

Bridge and Perzin [1] could not correlate prognosis with either the histologic grade or the gross appearance of the tumor; however, prognosis did correlate with depth of tumor invasion and tumor size. In addition, mesenteric lymph node metastasis almost certainly portends a fatal outcome.

Darling and Welch [2] found a dismal 5% five-year survival among 20 patients with jejunal or ileal carcinoma. Most other series generally report better 5-year survival rates ranging between 22% and 28% [1, 3, 6]. This may be improved with curative rather than palliative or bypass surgery [1].

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