Duodenal Tumors

16

Jun Haeng Lee and Woo Chul Chung

Contents

16.1 Duodenal Polyps

Duodenal polyps are usually found incidentally in up to 2–4% of patients. The majority of patients are asymptomatic. Symptoms related to the duodenal polyps can be nonspecific discomfort, abdominal pain, obstruction, intussusception, and bleeding. The histological subtype of polyps is sometimes difficult to determine by endoscopic appearance alone, and biopsy is necessary.

They are divided them into nonneoplastic and neoplastic lesions. Nonneoplastic lesions are inflammatory hyperplastic polyps, gastric heterotopia, ectopic pancreas, and Brunner's gland hyperplasia/hamartoma. Adenomas, gastrointestinal stromal tumors (GISTs), neuroendocrine tumors, and metastatic cancer are neoplastic lesions. In previous report, the polyps of more than 10 mm or polyps in the second portion of the duodenum should be carefully observed and evaluated by histological examination for malignant potentials [1].

J. H. Lee
Department of Medicine, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, South Korea

W. C. Chung (✉)
Department of Internal Medicine, St. Vincent's Hospital, College of Medicine, The Catholic University of Korea, Suwon, Gyeonggi-Do, South Korea
e-mail: jwchulkr@catholic.ac.kr

H. J. Chun et al. (eds.), *Clinical Gastrointestinal Endoscopy*, https://doi.org/10.1007/978-981-10-4995-8_16

16.1.1 Inflammatory Hyperplastic Polyps

Inflammatory hyperplastic polyps (Figs. 16.1 and 16.2) are the most common histologic type. At endoscopy, inflammatory hyperplastic polyps are small, sessile polyps. They are located at the bulbar and postbulbar duodenum. They are small and quite commonly multiple. In the endoscopy, the surface may be normal or erythematous, eroded, or ulcerated.

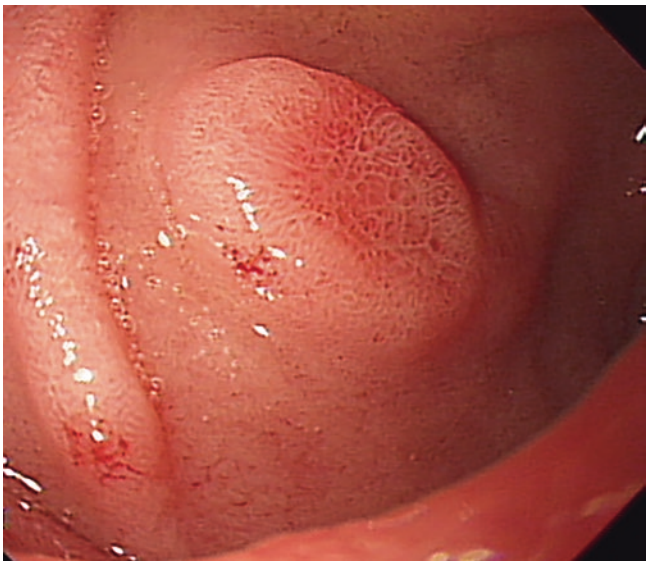

Fig. 16.1 Inflammatory hyperplastic polyp of the duodenum. About 10 mm-sized sessile polyp is observed in the duodenum

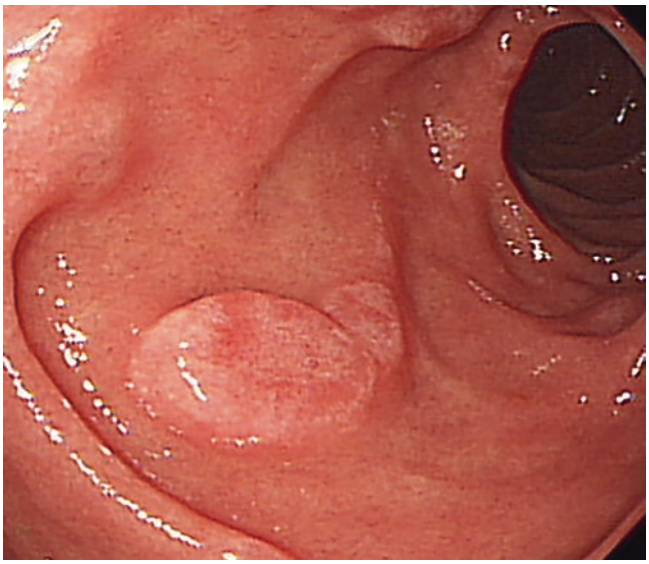

Fig. 16.2 Inflammatory hyperplastic polyp of the duodenum. In the duodenal bulb, polyps smaller than 10 mm do not need biopsy or treatment. Hyperplastic polyps of the duodenum seem to occur most frequently in patients with peptic ulcer disease

16.1.2 Gastric Heterotopia

Gastric heterotopia is usually located in the duodenal bulb. It is divided into congenital heterotopic gastric mucosa and acquired metaplastic gastric surface epithelium. The former may be single sessile polyp (Fig. 16.3), and the latter acquired forms are multiple variable-sized flat elevated lesions with unclear margin (Figs. 16.4 and 16.5). The surface may be hyperemic and reticular. Historically, the relationship between gastric heterotopia and peptic ulcer disease has been proposed without relevant evidences. Lymphoid hyperplasia is typically multiple small pale sessile polyps in the duodenal bulb (Fig. 16.6).

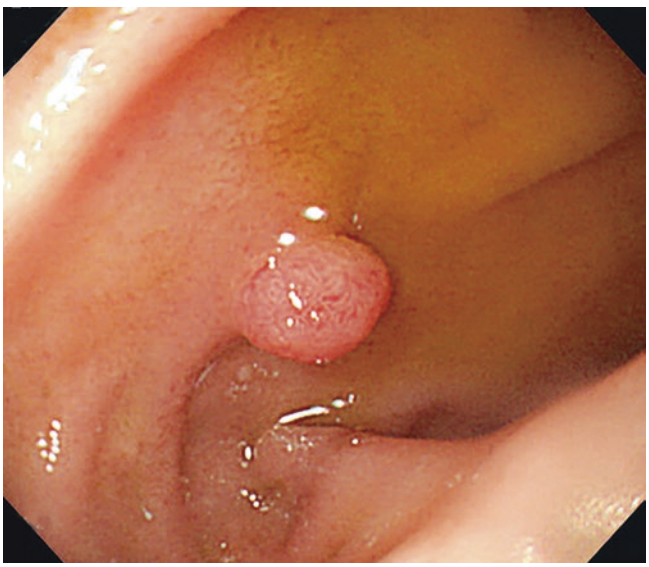

Fig. 16.3 Heterotopic gastric mucosa. A 5 mm-sized polyp in the duodenal bulb is observed. On histologic examination, there is a heterotopic gastric tissue which consists of well-formed body-type glands above muscularis mucosa

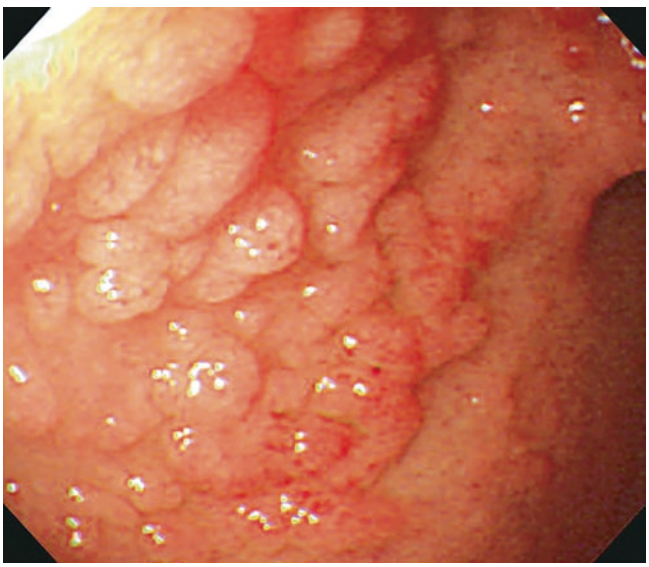

Fig. 16.5 Gastric metaplasia of the duodenum. Multiple nodular mucosal elevated lesions of duodenal bulb are seen at bulb

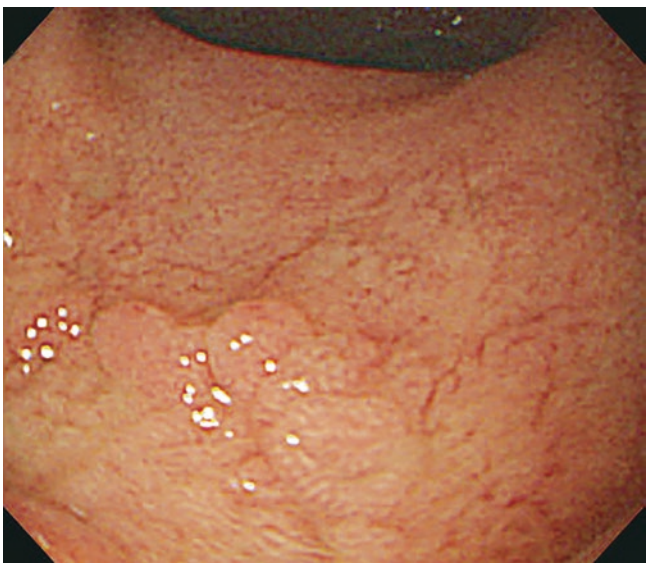

Fig. 16.4 Gastric metaplasia of the duodenum. The nodular mucosal elevated lesion is observed through the pylorus, and the color shows no difference from that of normal duodenal mucosa

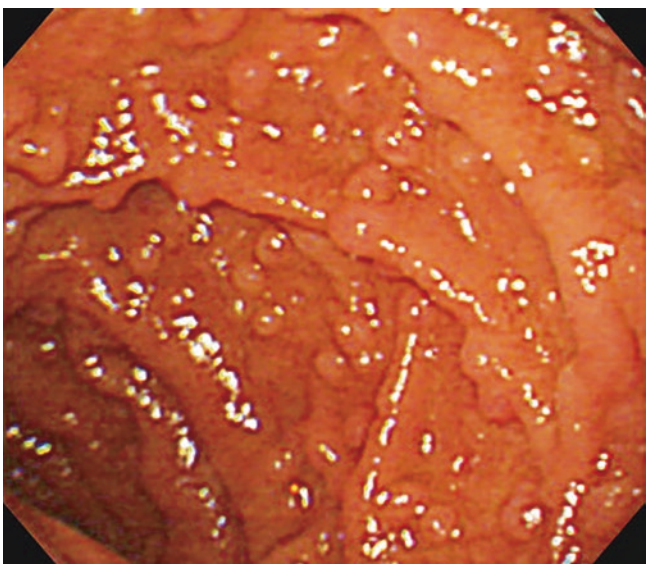

Fig. 16.6 Lymphoid hyperplasia. Endoscopy reveals diffuse nodularity in the post-bulb of the duodenum. The duodenal biopsy shows mucosal lymph node aggregates

16.1.3 Ectopic Pancreas

Ectopic pancreas (Figs. 16.7 and 16.8) is a pancreatic tissue outside the boundaries of the pancreas and may manifest as a submucosal mass. It can be misinterpreted as another submucosal tumor such as GISTs. Currently, endoscopic ultrasound (EUS) can differentiate from other types of submucosal tumors (Fig. 16.9).

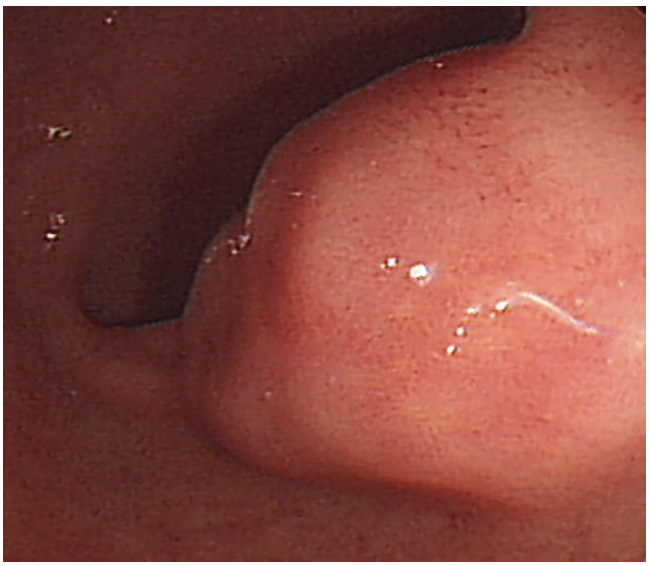

Fig. 16.7 Ectopic pancreas of the duodenum. Ectopic pancreas was usually diagnosed as a subepithelial tumor which was firm and slightly irregular

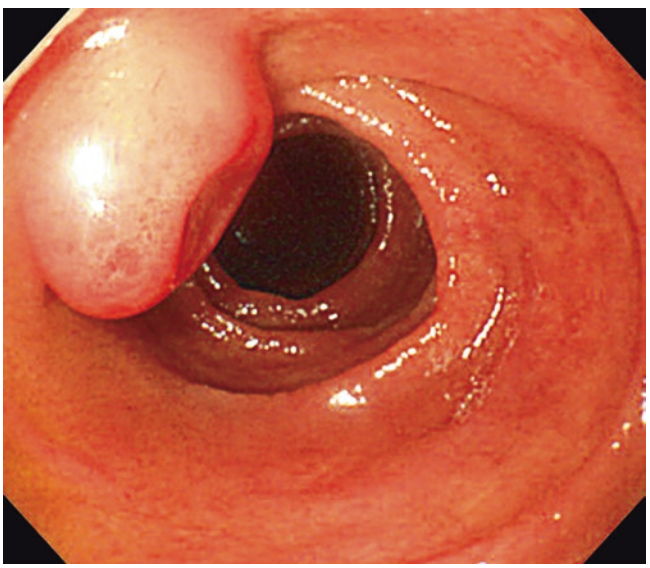

Fig. 16.8 Ectopic pancreas of the duodenum. The mucosa over the lesion may have a central depression or dimpling

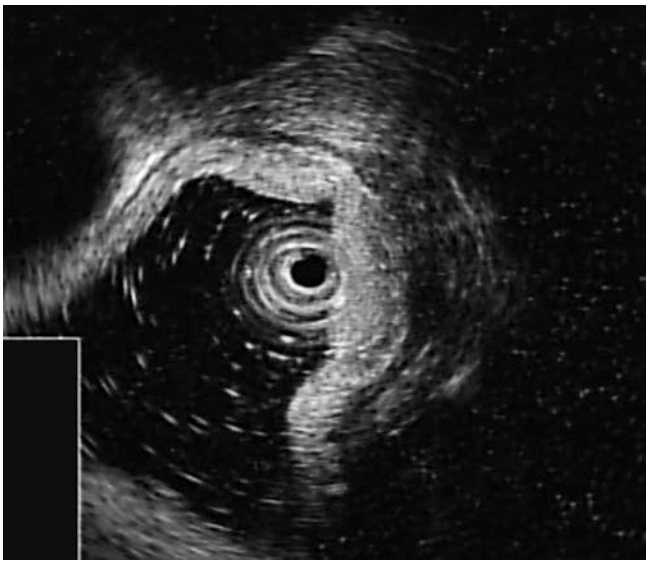

Fig. 16.9 EUS finding of ectopic pancreas. The characteristic EUS image of lesion is hypoechogenic with heteroechogenic structure. The lesion is located within the hyperechogenic submucosa (the third echo-endoscopic layer) or in the submucosal and muscularis propria (the third and fourth layers)

16.1.4 Brunner's Gland Hyperplasia

Brunner's gland hyperplasia and hamartoma are infrequently encountered polypoid nodules and masses in the proximal duodenum. Brunner's gland hyperplasia (Fig. 16.10) is a benign tumor of the duodenum, and it has a main physiological function of secreting an alkaline-based mucus. Brunner's gland hamartoma (Fig. 16.11) is a submucosal mass having a pedicle.

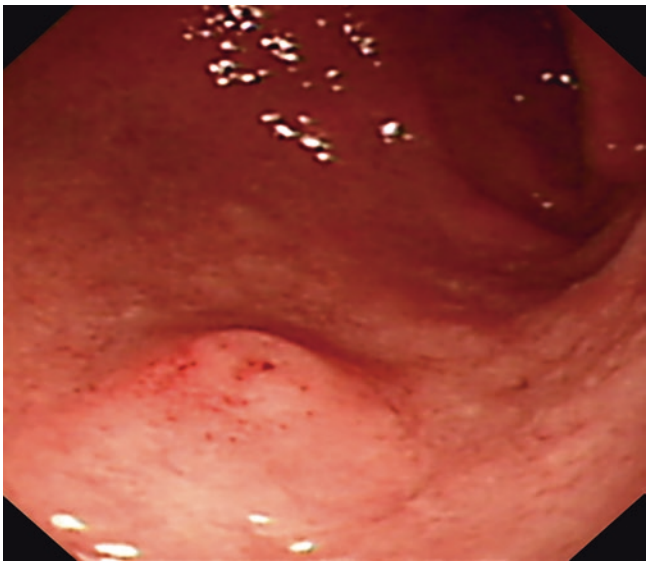

Fig. 16.10 Brunner's gland hyperplasia. Gastroduodenoscopic finding shows a sessile or elevated, umbilicated lesion in the duodenal bulb

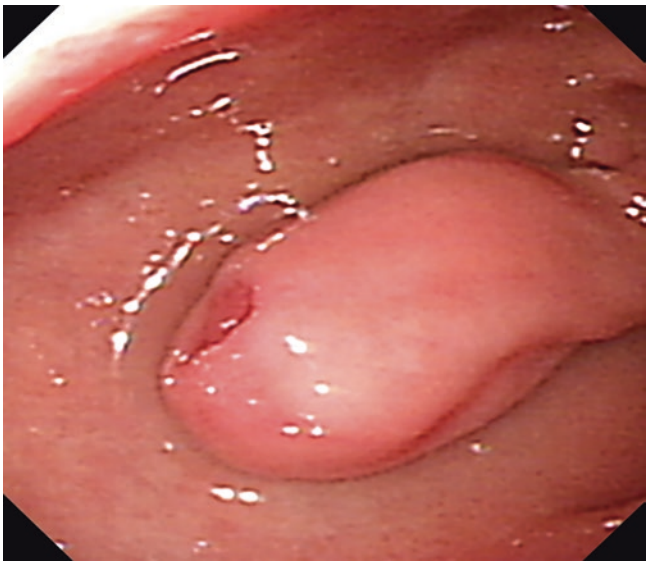

Fig. 16.11 Brunner's gland hamartoma. Gastroduodenoscopy shows a pedunculated polyp measuring approximately 25 mm in the duodenal bulb. Biopsy specimen is usually negative due to the submucosal nature of the tumor

16.2 Duodenal Adenomas/ Adenocarcinomas

16.2.1 Duodenal Adenomas

Duodenal adenomas are rare conditions. Approximately 40% of them are sporadic, and the remaining 60% are associated in patients with familial adenomatous polyposis (FAP) [2]. They are thought to progress to duodenal adenocarcinomas with accumulation of genetic mutations. Duodenal adenomas are geographic-shaped flat elevated lesions (Fig. 16.12) or semi-pedunculated polyps (Fig. 16.13).

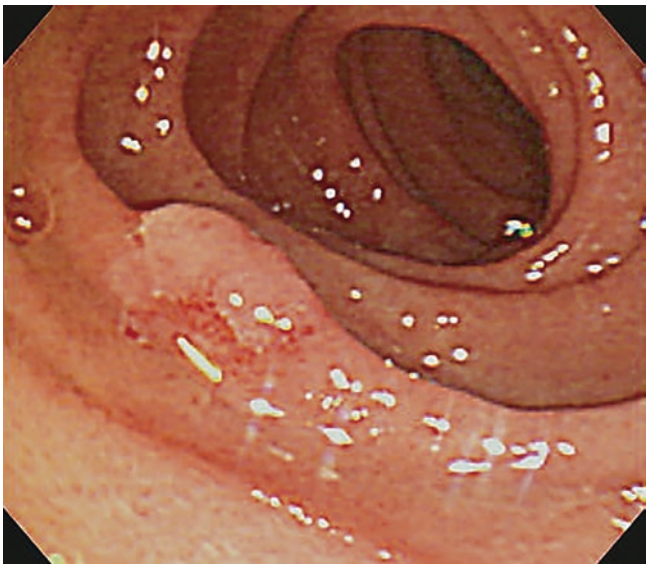

Fig. 16.12 Duodenal adenoma. There is a 12 mm-sized superficial elevated type (IIa) lesion in the second portion of the duodenum

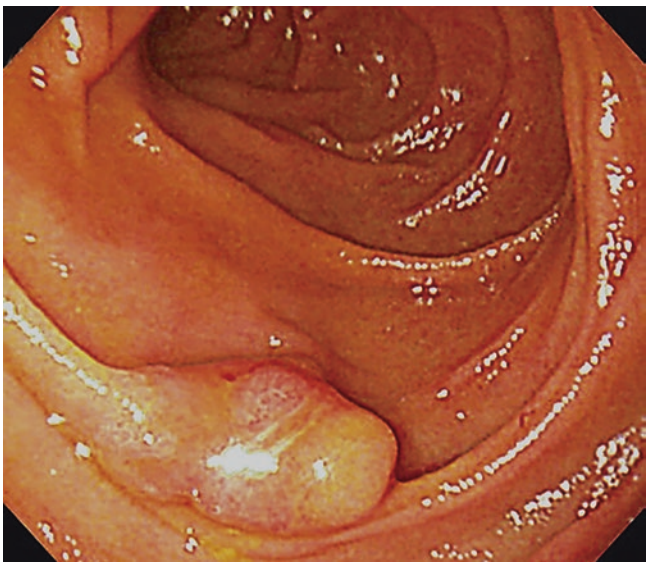

Fig. 16.13 Duodenal adenoma. A 14 mm-sized sessile, centrally depressed polyp is found in the second portion of the duodenum

16.2.2 Duodenal Adenocarcinomas

Adenocarcinomas are the most common malignant tumors in the duodenum but account for less than 0.4% of all gastrointestinal cancers. The symptoms of duodenal adenocarcinomas are usually pain, bleeding, and biliary obstruction; symptoms of intestinal obstruction are possible but uncommon [3]. The endoscopic appearance of adenocarcinomas is not specific and cannot be differentiated from lymphoma or leiomyosarcoma. The lesions are usually nodular (Figs. 16.14 and 16.15), flat wall thickening, (Fig. 16.16) and fungating (Fig. 16.17). Also, it may be ulcerated or obstructed (Figs. 16.18 and 16.19). Duodenal adenocarcinomas are most often confined to the second or third portion of the duodenum. Some cases with adenocarcinomas of the third or fourth part of the duodenum cannot be found in the routine upper endoscopy (Fig. 16.20). In that situation, push enteroscopy or upper endoscopy with colonoscope may be useful (Fig. 16.21).

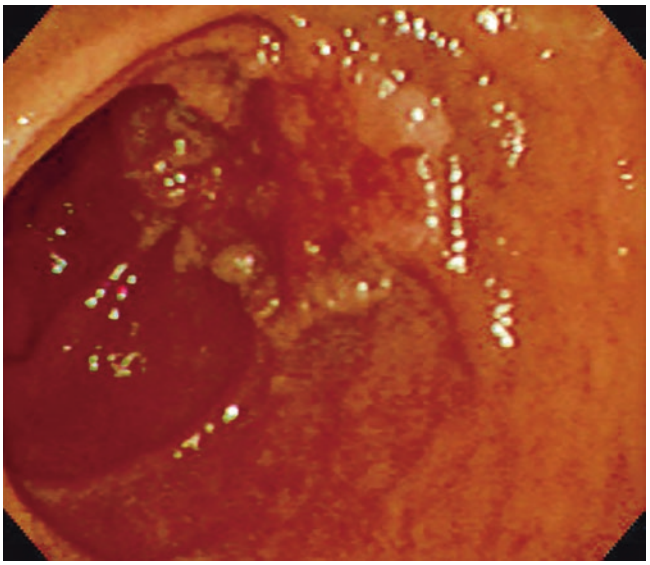

Fig. 16.14 Duodenal adenocarcinoma. A large laterally spreading sporadic duodenal adenoma is seen in the second portion of the duodenum

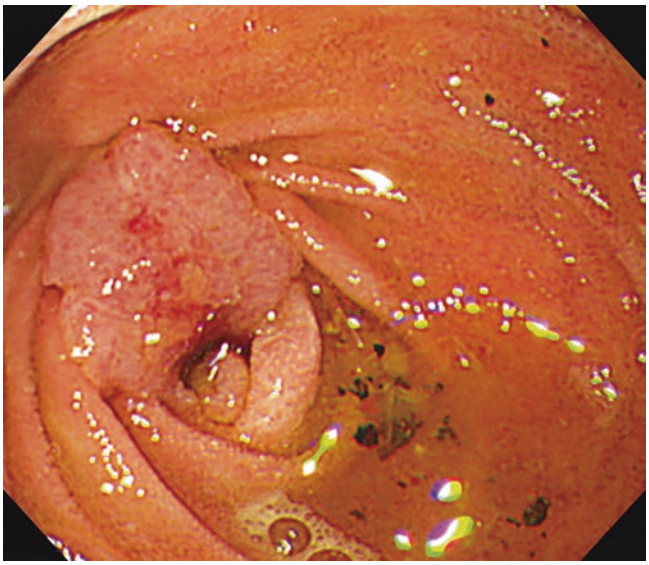

Fig. 16.16 Duodenal adenocarcinoma. A flat and thickened area of the mucosa is seen in the second portion of duodenum, and it results in significant luminal narrowing

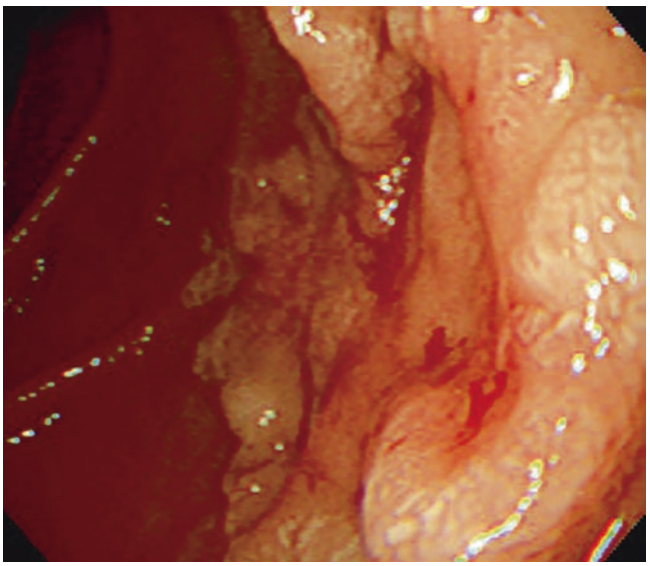

Fig. 16.15 Duodenal adenocarcinoma. A huge sessile and nodular lesion is seen in the second portion of the duodenum

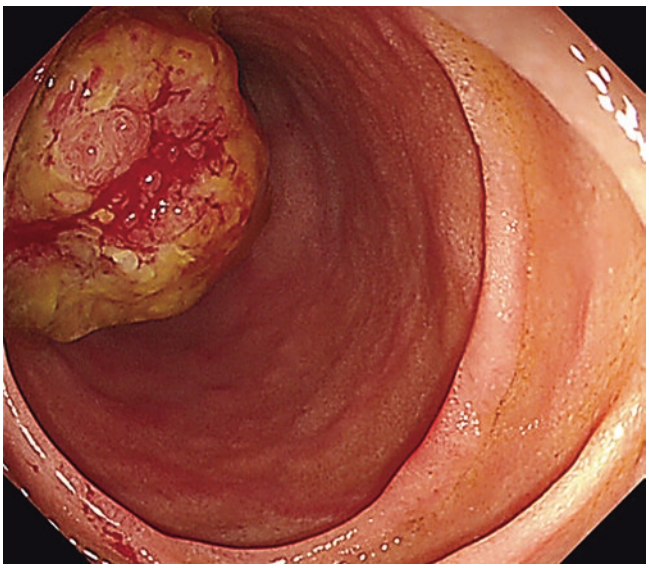

Fig. 16.17 Duodenal adenocarcinoma. A huge fungating mass with luminal narrowing is found in the second and third portions of the duodenum

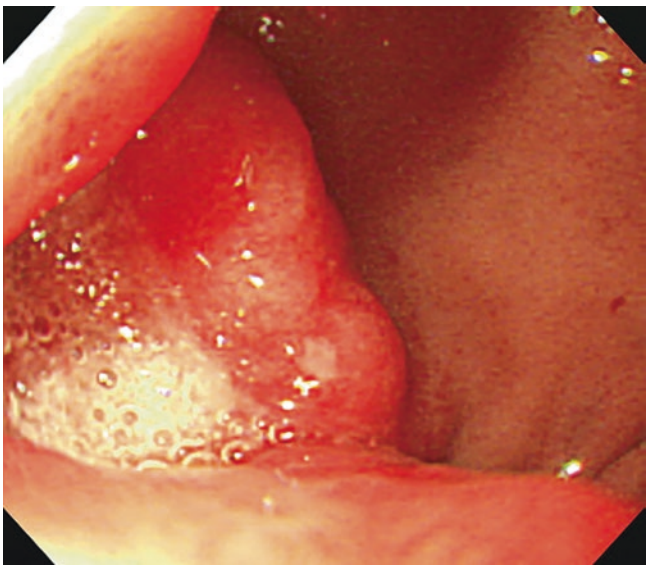

Fig. 16.18 Duodenal adenocarcinoma. Duodenal malignancy is strongly suspected when the mass is ulcerated or over 2 cm in size

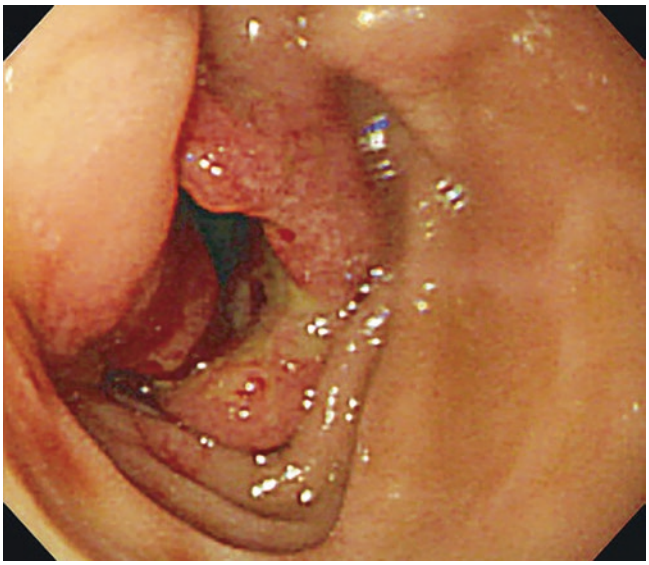

Fig. 16.19 Duodenal adenocarcinoma. A circumferential ulceration with wall thickening is observed in the second portion of duodenum, it is usually revealed by the obstruction symptoms

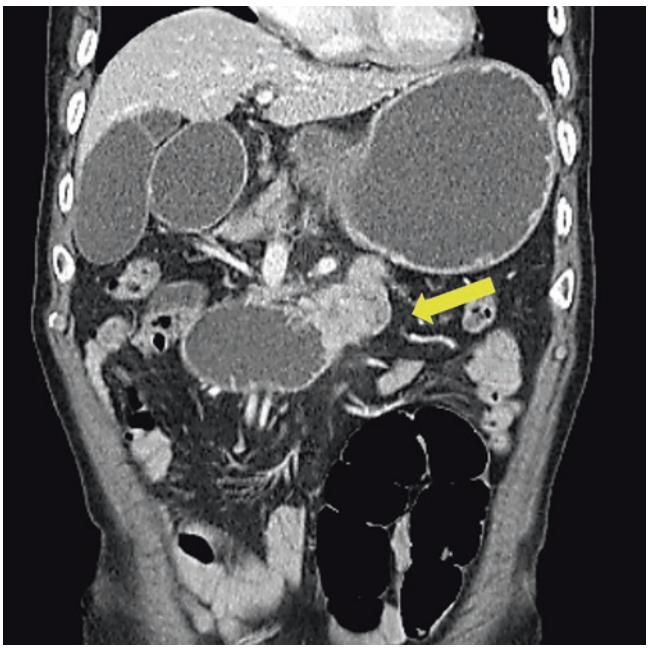

Fig. 16.20 Duodenal adenocarcinoma. Computed tomography shows infiltrative heterogeneous enhancing mass in the distal fourth portion of duodenum adjacent to the Treitz ligament (yellow arrow)

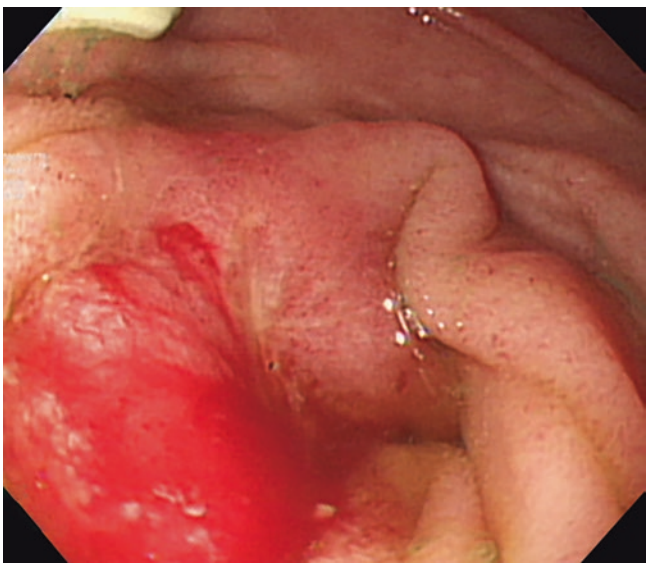

Fig. 16.21 Duodenal adenocarcinoma. Push enteroscopy shows an ulcerating mass with obstruction in the fourth part of the duodenum

16.3 Duodenal GISTs

GISTs are spindle cell tumors and arise in the smooth muscle pacemaker interstitial cell of Cajal. CD34 and CD117 (c-kit protein) are identified as their markers. Duodenal GISTs represent about 6–21% of surgically resected GISTs [4]. Clinical presentations may be gastrointestinal bleeding, pain, and rarely intestinal obstruction. The most common location is the second part of the duodenum. In endoscopy, the characteristics of the duodenal GISTs are the same with gastric GISTs. It is usually a mass with normal-looking surface (Fig. 16.22). There may be central ulcerations (Figs. 16.23 and 16.24). In some cases, it is difficult to differentiate from the more common duodenal adenocarcinoma (Fig. 16.25). On EUS, GIST is homogeneous and its echogenicity is similar to that of the normal proper muscle layer. Usually, it originates from the proper muscle layer (Fig. 16.26). Several cases of multiple GISTs were reported in a patient with neurofibromatosis type I (Fig. 16.27) [5].

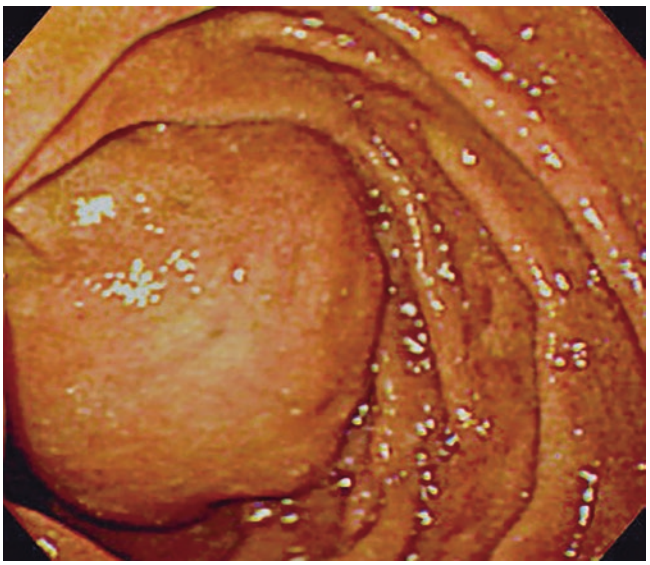

Fig. 16.22 Duodenal GIST. Huge subepithelial mass with smooth surface is seen in the second portion of the duodenum

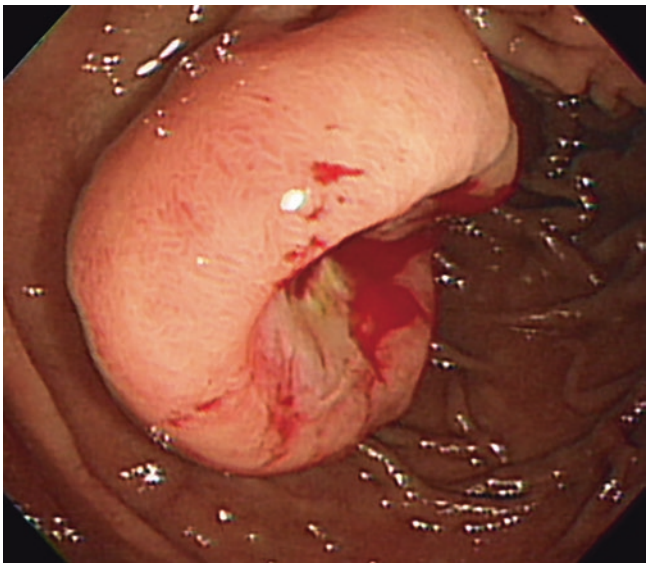

Fig. 16.24 Duodenal GIST. Most common symptom is gastrointestinal bleeding

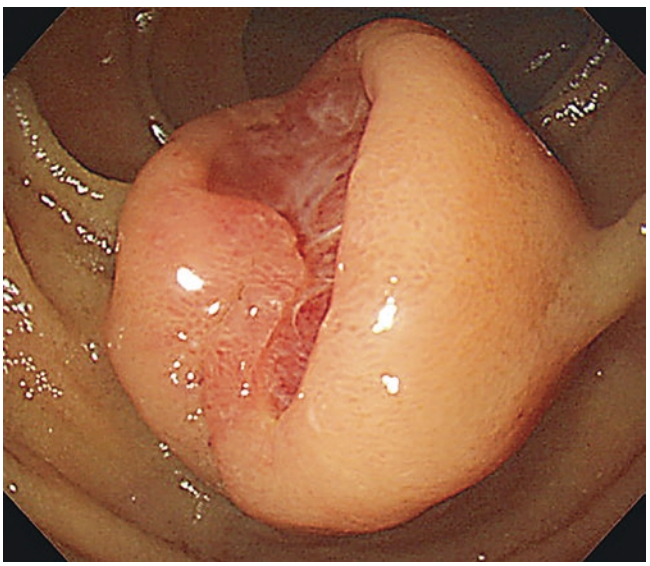

Fig. 16.23 Duodenal GIST. It often has central ulceration

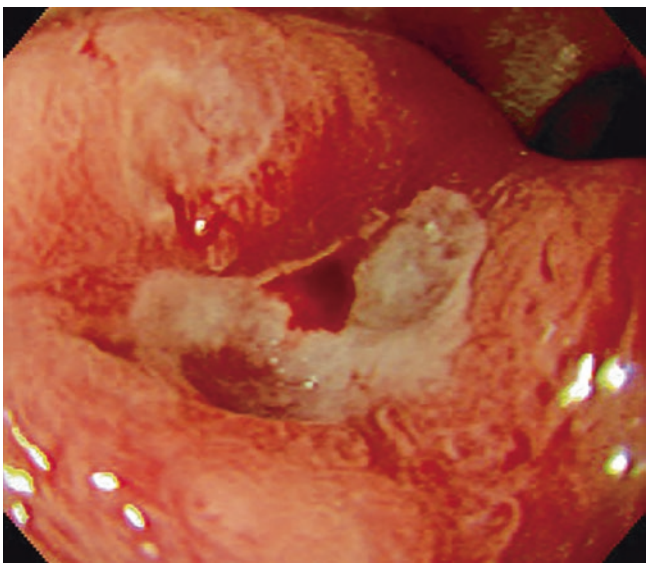

Fig. 16.25 Duodenal adenocarcinoma. It is difficult to differentiate between duodenal GIST and adenocarcinoma

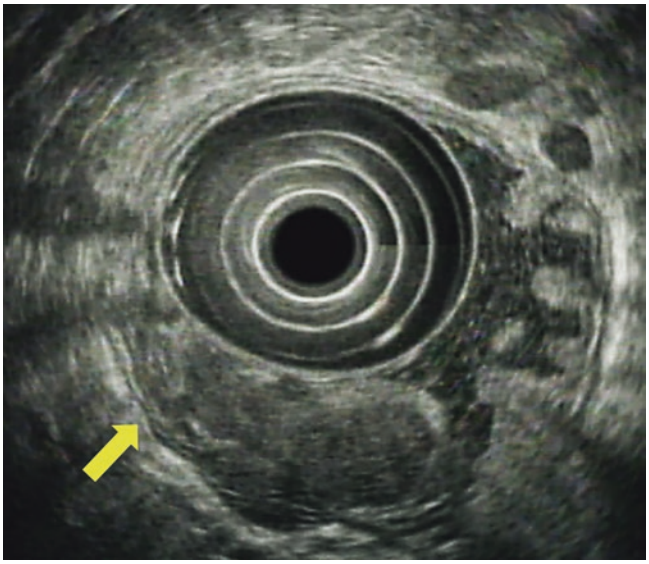

Fig. 16.26 EUS finding of duodenal GIST. Hypoechoic mass is contiguous with the fourth hypoechoic layer of the wall, which corresponds to the muscularis propria (yellow arrow)

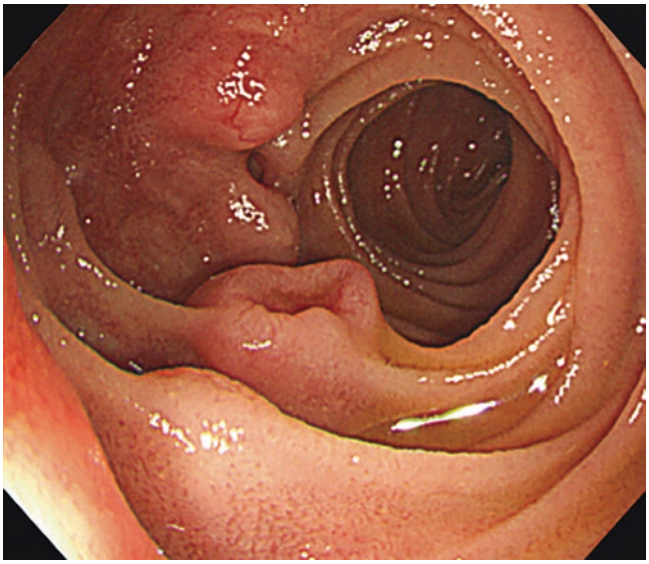

Fig. 16.27 Multiple duodenal GIST in a patient with neurofibromatosis type I. Gastroduodenoscopy revealed multiple polypoid formation with central ulceration of the second portion of the duodenum

16.4 Duodenal Neuroendocrine Tumor (NET)

Recently, small duodenal NETs are increasingly recognized with the more widespread use of upper gastrointestinal endoscopy. They are more common in men. Some of the NETs are functional tumors like gastrinomas or somatostatinomas. Gastrinomas tend to be smaller than somatostatinomas. Duodenal NETs are typically small polypoid lesions with smooth slightly yellow overlying mucosa (Figs. 16.28 and 16.29). Forceps biopsy is very effective for histological diagnosis. There may be top ulcerations (Fig. 16.30). On EUS, duodenal NET has homogeneous iso-echogenicity between muscularis mucosa and submucosal layer (Fig. 16.31).

The standard treatment for small duodenal NETs is endoscopic resection, but the rate of perforation may be very high up to 30%. Given the risks associated with EMR and the likely favorable natural history of small duodenal NETs, conservative management with close follow-up may represent a viable alternative to endoscopic treatment, especially in patients with a high risk of perioperative complications [6].

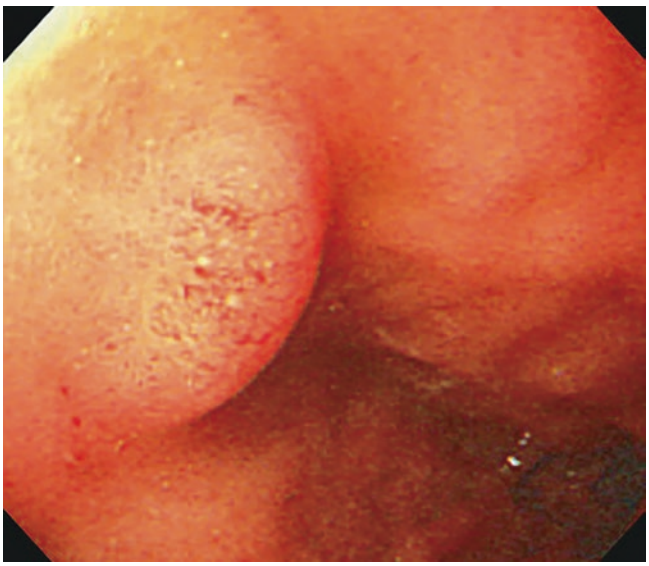

Fig. 16.28 Duodenal NET. A yellowish sessile polyp smaller than 10 mm is seen in the duodenal bulb

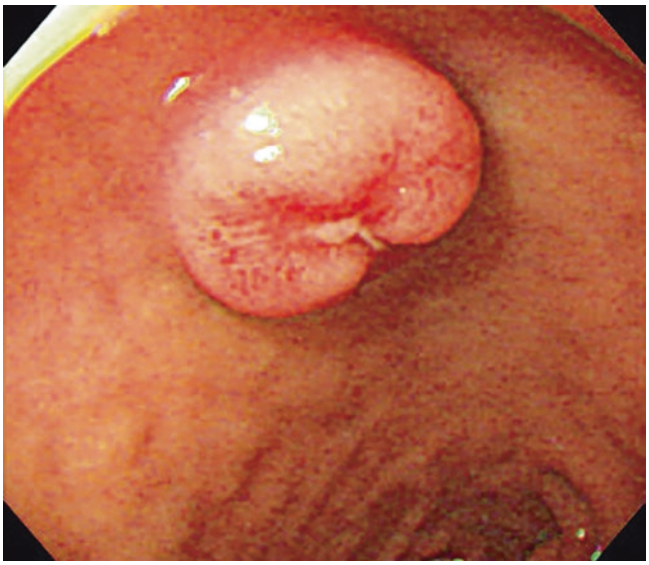

Fig. 16.30 Duodenal NET. Sometimes, it has central ulceration

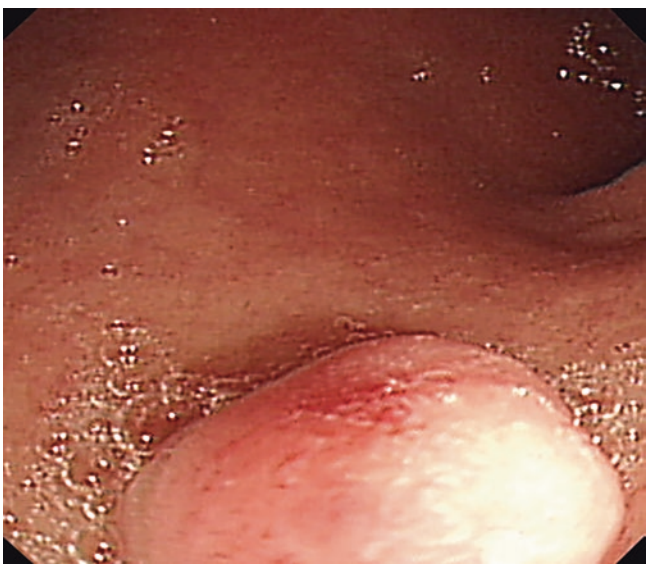

Fig. 16.29 Duodenal NET. An elevated lesion accompanied by an irregularly shaped erythematous central depression is observed in the duodenal bulb

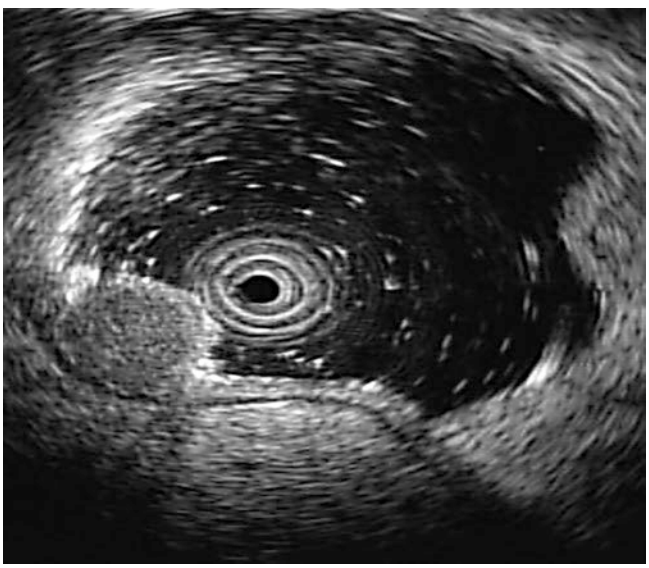

Fig. 16.31 EUS finding of duodenal NET. Well-defined hypoechoic tumor confines to the submucosal hyperechoic layer, and the underlying hypoechoic muscularis propria is intact

16.5 Other Duodenal Subepithelial Tumors

There are many different kinds of polyps in the duodenum, such as submucosal cysts (Fig. 16.32), lipoma (Figs. 16.33 and 16.34), lymphangioma (Fig. 16.35), and others.

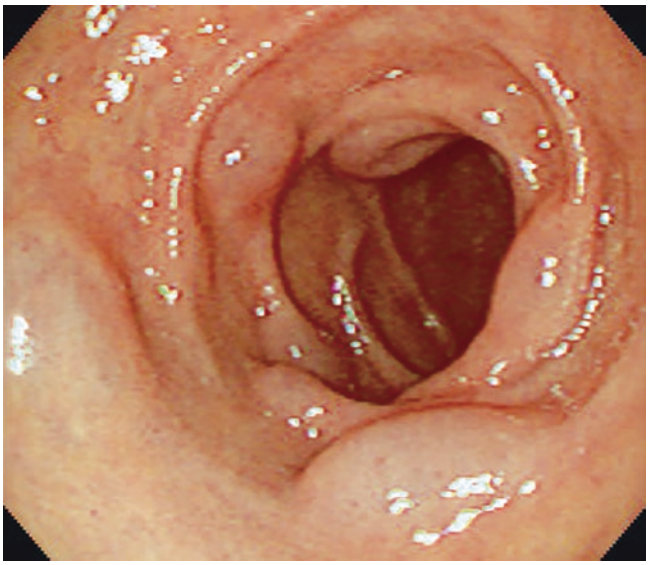

Fig. 16.32 Duodenal submucosal cysts. There are multiple semitransparent lesions with smooth surface in the duodenal bulb

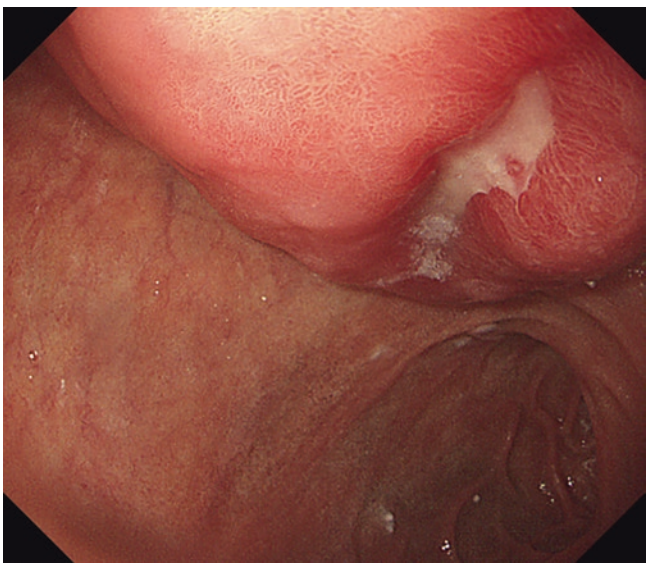

Fig. 16.33 Duodenal lipoma. There is a giant duodenal lipoma presenting with gastrointestinal bleeding in the duodenal bulb. It has subepithelial property with surface ulceration

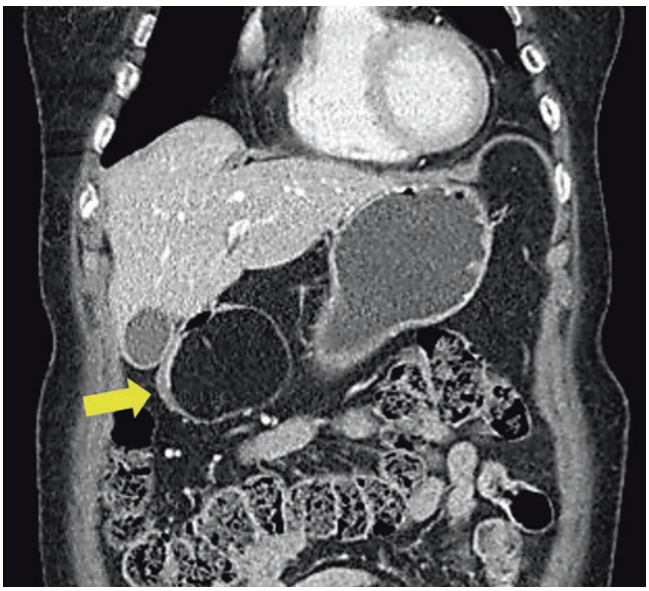

Fig. 16.34 Computed tomographic finding of duodenal lipoma (yellow arrow). It shows an overall hypodense homogeneous low signal lesion

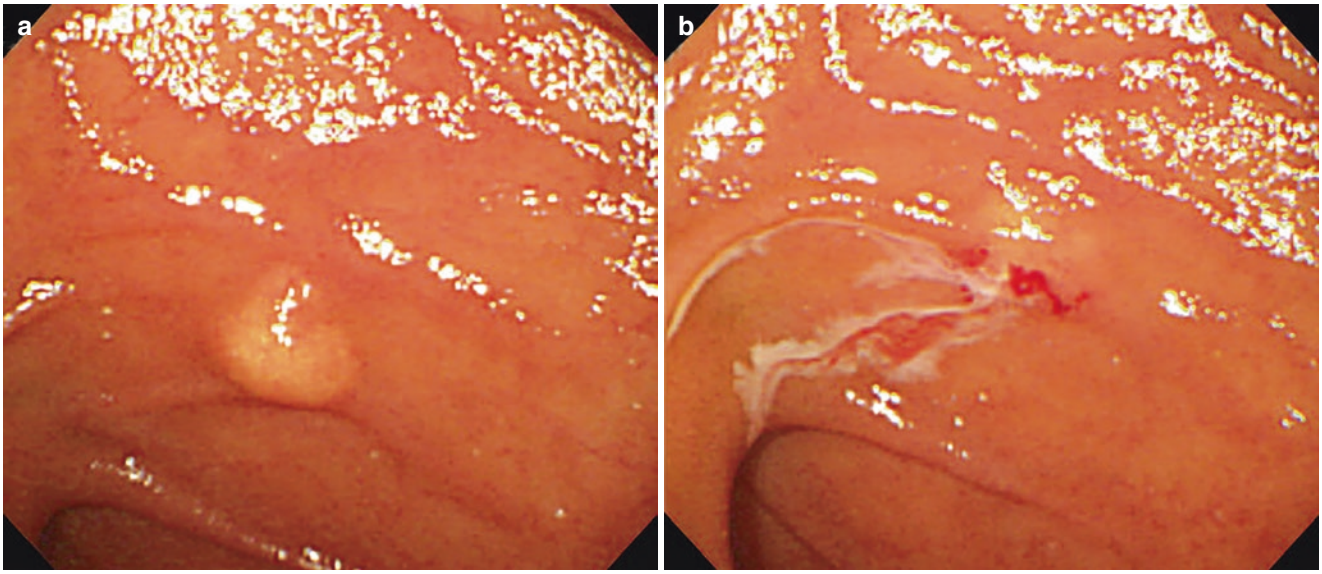

Fig. 16.35 Duodenal lymphangioma. (**a**) It is a single, about 10 mm, polyp-like lesion in the second portion of the duodenum. (**b**) When it is grasped by biopsy forceps, milky liquid flows into the lumen

16.6 Duodenal Lymphomas

The most common site of extranodal lymphoma is the gastrointestinal tract. Gastrointestinal lymphomas make up approximately one-third to one-half of extranodal lymphomas and approximately 1% of all gastrointestinal neoplasms. The stomach (50–60%) is the most common site of gastrointestinal lymphomas, followed by the small intestine (20–30%) and the colon (10–20%). The ileum is the most common site of small bowel lymphomas, followed by the jejunum and then the duodenum. Duodenal lymphomas make up only about 5% of gastrointestinal lymphomas. In endoscopy, duodenal lymphomas may have different appearances. They usually apear as large masses, which may be exophytic, polypoid or ulcerated.

The most common histological type of duodenal lymphoma is diffuse large B cell lymphoma (DLBCL), which afflicts relatively young patients, is more likely to present with disseminated disease, and is more likely to require surgery (Figs. 16.36 and 16.37). MALT lymphoma (Fig. 16.38) and follicular lymphomas (Fig. 16.39 and 16.40) are usually seen in older patients. Mantle cell lymphomas (MCL) (Fig. 16.41) and T-/NK-cell lymphomas (Fig. 16.42) are rare but have worst prognosis.

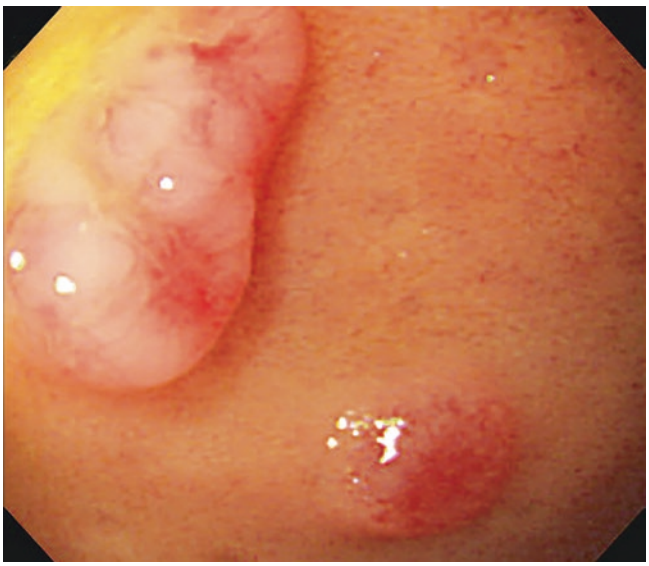

Fig. 16.36 Diffuse large B cell lymphoma of the duodenum. Gastroduodenoscopy reveals multiple edematous swelling of the mucosa at duodenal bulb

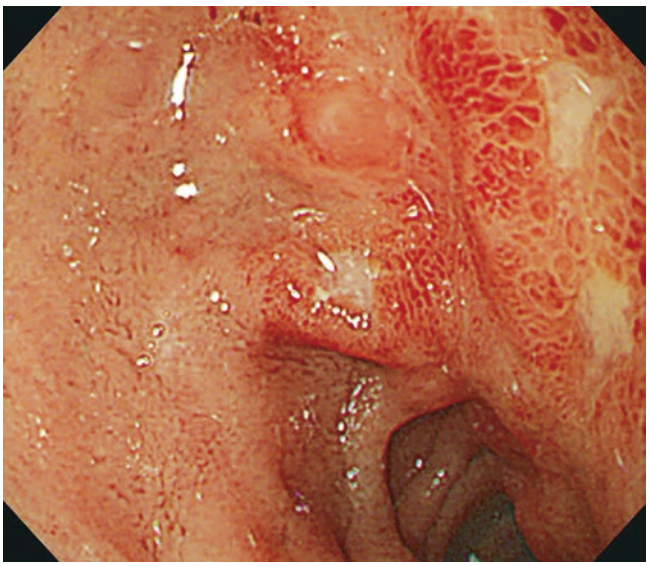

Fig. 16.38 MALT lymphoma of the duodenum. Multiple shallow ulcerations with edematous mucosa are seen with fold fusion on the duodenum blub (near superior duodenal angle)

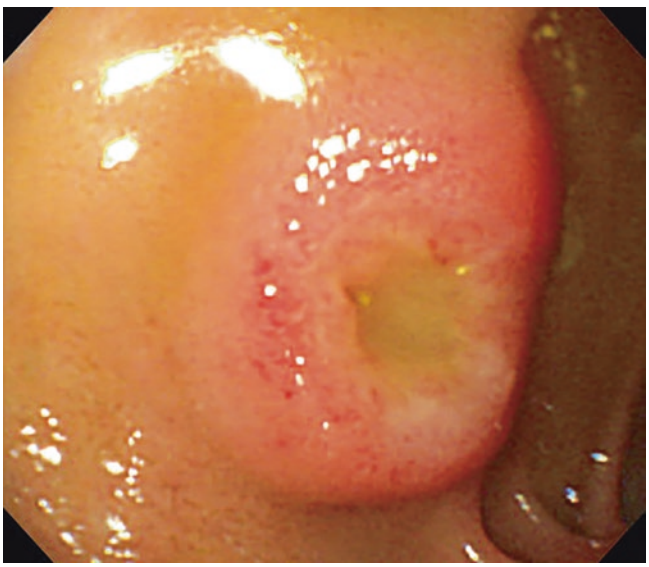

Fig. 16.37 Diffuse large B cell lymphoma of the duodenum. Sometimes, a spontaneously bleeding polyp with ulceration is observed at the duodenal bulb

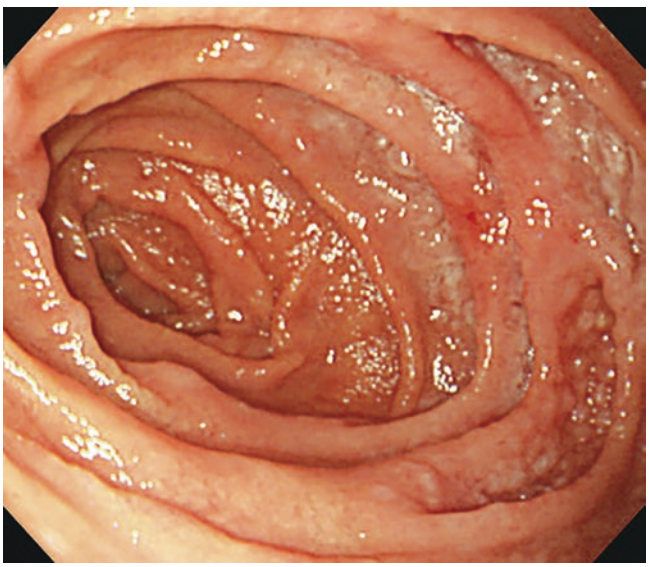

Fig. 16.39 Follicular lymphoma of the duodenum. Nodular mucosa with plaque-like lesions present in the second and third part of the duodenum

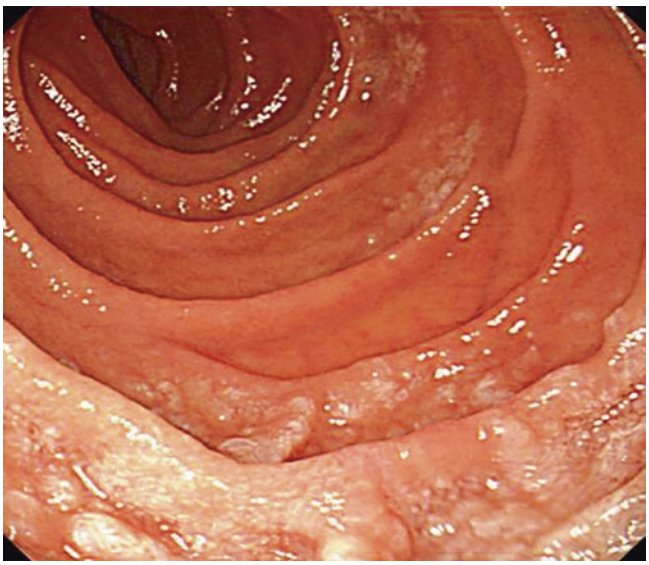

Fig. 16.40 Follicular lymphoma of the duodenum. The duodenal lesions are observed as multiple whitish granules or nodules

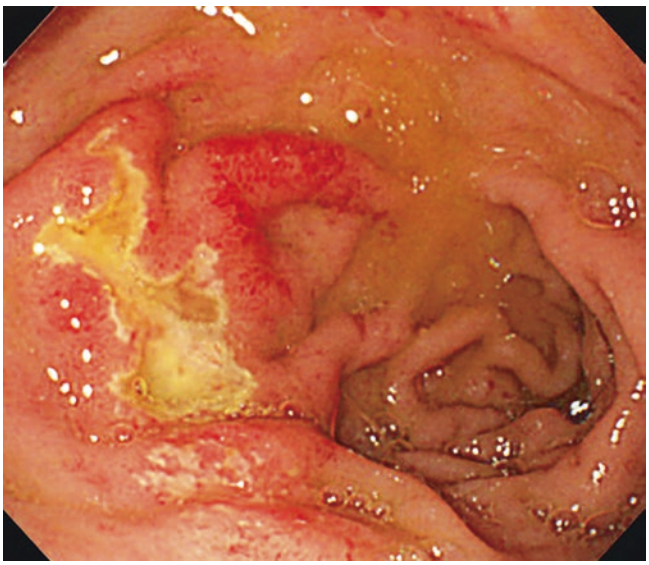

Fig. 16.41 MCL of the duodenum. The endoscopic findings in GI MCL are variable. Ulcero-infiltrative lesion is seen in the duodenal bulb

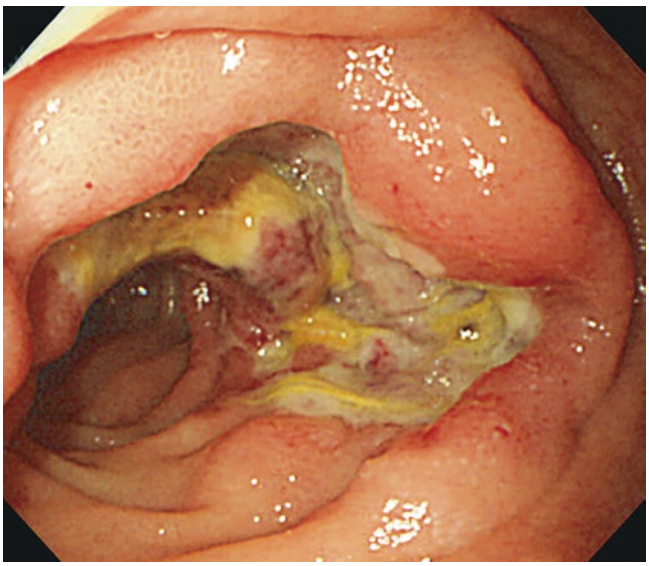

Fig. 16.42 T-/NK-cell lymphoma of the duodenum. Primary gastrointestinal NK−/T-cell lymphoma was endoscopically characterized by superficial/erosive, ulcerative, or ulcero-infiltrative lesions without fungating mass

16.7 Duodenal Involvement of Other Malignancies

Various types of malignancies of liver (Fig. 16.43) and pancreas can directly invade the duodenal wall, which can cause bleeding or obstruction (Fig. 16.44). Duodenal metastases from lung adenocarcinoma are extremely uncommon. Endoscopic image reveals multiple polypoid-like lesions with superficial erosion/ulcer (Fig. 16.45).

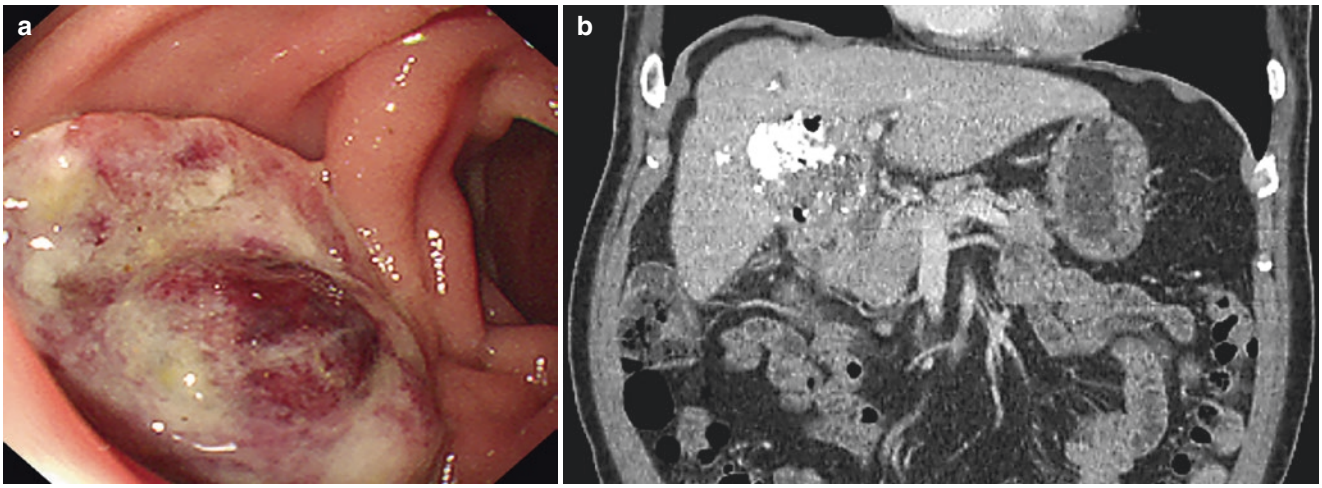

Fig. 16.43 Duodenal involvement of hepatocellular carcinoma. (**a**) A large duodenal ulcerating mass with ulcer and adherent blood clots is observed in patient with hepatocellular carcinoma. (**b**). Computed tomographic image of duodenal involvement of hepatocellular carcinoma. The majority of cases are direct tumor invasion

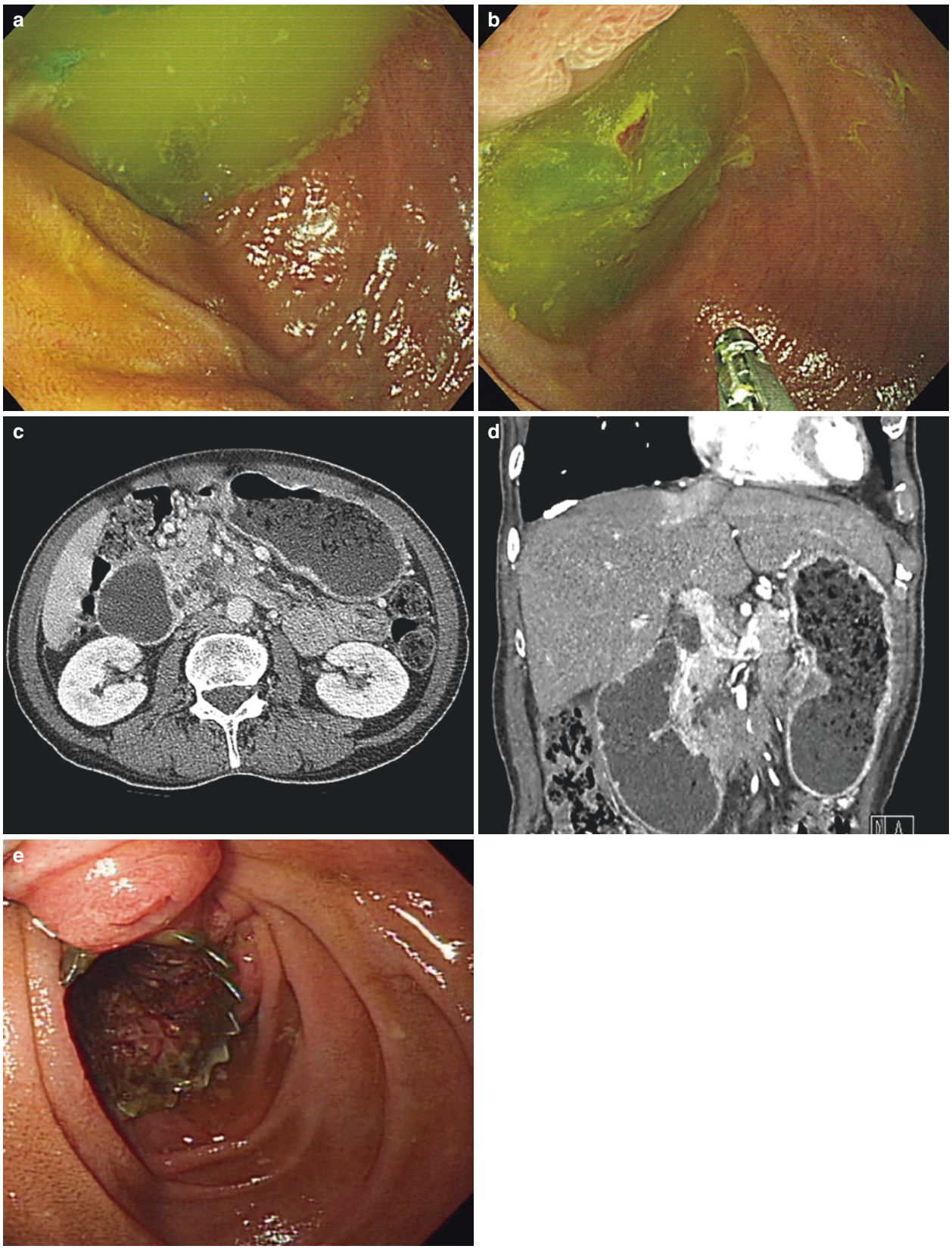

Fig. 16.44 Duodenal involvement of pancreatic cancer. (**a, b**) As pancreatic tumors enlarge, direct duodenal invasion and obstruction will happen. There is a bile pool with food materials in the duodenum. (**c, d**). The computed tomographic finding of duodenal involvement of pancreatic cancer. Tumor located in the pancreatic head and uncinate process tends to obstruct the third parts of the duodenum. Proximal part of the duodenum is marked dilated. (**e**) The delivery of expandable metallic stent relieves gastroduodenal obstruction

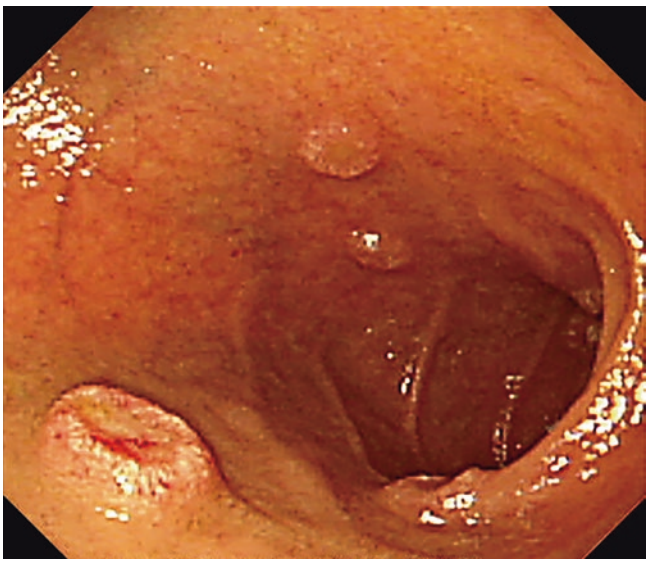

Fig. 16.45 Duodenal metastases from lung adenocarcinoma. Multiple polyp-like lesions with superficial ulcers of the duodenal bulb are seen in patients with lung adenocarcinoma

References

1. Jung SH, Chung WC, Kim EJ, et al. Evaluation of non-ampullary duodenal polyps: Comparison of non-neoplastic and neoplastic lesions. World J Gastroenterol. 2010;16:5474–80.
2. Johnson MD, Mackey R, Brown N, Church J, Burke C, Walsh RM. Outcome based on management for duodenal adenomas: sporadic versus familial disease. J Gastrointest Surg. 2010;14:229–35.
3. Hung FC, Kuo CM, Chuah SK, et al. Clinical analysis of primary duodenal adenocarcinoma: an 11-year experience. J Gastroenterol Hepatol. 2007;22:724–8.
4. Johnston FM, Kneuertz PJ, Cameron JL, et al. Presentation and management of gastrointestinal stromal tumors of the duodenum: a multi-institutional analysis. Ann Surg Oncol. 2012;19:3351–60.
5. Kang DY, Park CK, Choi JS, et al. Multiple gastrointestinal stromal tumors: clinicopathologic and genetic analysis of 12 patients. Am J Surg Pathol. 2007;31:224–32.
6. Min BH, Kim ER, Lee JH, et al. Management strategy for small duodenal carcinoid tumors: does conservative management with close follow-up represent an alternative to endoscopic treatment? Digestion. 2013;87:247–53.