# Tumorous Lesions of the Small Bowel

# 29

Hwang Choi

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Small bowel tumors are relatively rare. They can be classified as epithelial, mesenchymal, lymphoproliferative, or metastatic tumors. Endoscopic differential diagnosis usually starts with observation of the overlying epithelial layer. Neuroendocrine tumors, stromal tumors, hamartomas, lipomas, lymphangiomas, and inflammatory fibroid polyps are usually covered with normal epithelial layers. Stromal tumors, neuroendocrine tumors, and inflammatory fibroid polyps are usually firm, while most hamartomas, lipomas, and lymphangiomas are relatively soft. Yellowish adipose tissue in lipomas and clear fluid in lymphangiomas can be seen though the overlying mucosa. Stromal tumors and neuroendocrine tumors often present with surface ulcers. Unlike the above subepithelial lesions, the surface epithelium of adenoma and adenocarcinoma can be well discriminated from the surrounding normal mucosa. The epithelial layer of lymphomas and metastatic tumors also changes when tumor cells infiltrate the mucosal layer. Malignant lymphoma and gastrointestinal stromal tumors are thought to be the most common tumors in the small bowel.

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## 29.1 Introduction

Small bowel tumors are relatively rare, comprising 3–6 % of all primary gastrointestinal tumors. Approximately 60–75 % of small bowel tumors are benign [1].

The diagnosis of small bowel tumors is frequently delayed or even established incidentally because most are asymptomatic. Their common clinical symptoms are related to obstruction and bleeding. Small bowel tumors are the second most common cause of small intestinal bleeding following vascular lesions and are the leading cause of bleeding in patients below 50 years old. Another reason that small bowel tumors have often failed to be diagnosed early is that small bowel exploration was not feasible until recently. However, the development of capsule endoscopy and device-assisted endoscopy (e.g., balloon-assisted enteroscopy, spiral enteroscopy) launched a new era of diagnosis and treatment of small bowel tumors.

Capsule endoscopy is superior to device-assisted endoscopy in terms of noninvasiveness and convenience for total enteroscopy. However, the incidence of capsule retention can reach 9.7–25 % in patients with small bowel tumors, which is higher than the incidence in all patients receiving capsule endoscopy. The degree of concordance between capsule endoscopy and double-balloon endoscopy is 0.46 for small bowel tumors, which is lower than for vascular or inflammatory lesions because capsule endoscopy-based diagnosis is based only on gross findings and many small bowel tumors are located in the subepithelial layer.

Small bowel tumors are classified as epithelial, mesenchymal, lymphoproliferative, or metastatic (Table 29.1) [2]. Malignant lymphoma and gastrointestinal stromal tumors (GISTs) are thought to be the most common tumors in the small bowel, although the incidence may vary according to diagnostic modalities and countries [3, 4].

**Table 29.1** Classification of small bowel tumors

| | Cell type | Benign | Malignant |
|---|---|---|---|
| Epithelial | Glandular | Adenoma | Adenocarcinoma |
| | Neuroendocrine | Well-differentiated neuroendocrine tumor (benign carcinoid) | Well-differentiated neuroendocrine carcinoma (malignant carcinoid) |
| | | | Poorly differentiated neuroendocrine carcinoma |
| Mesenchymal | Vascular | Hemangioma | Angiosarcoma |
| | | Lymphangioma | Kaposi's sarcoma |
| | Adipocyte | Lipoma | Liposarcoma |
| | Interstitial cell of Cajal | GIST | GIST |
| | Smooth muscle cell | Leiomyoma | Leiomyosarcoma |
| | | Hamartoma | |
| | Nerve cell | Schwannoma | Malignant schwannoma |
| | | Neurofibroma | |
| Lymphoproliferative | B cell | | B cell lymphoma |
| | T cell | | T cell lymphoma |
| Metastatic | | | Direct invasion or distant metastasis |

*GIST* gastrointestinal stromal tumor

## 29.2 Tumors of the Small Bowel

### 29.2.1 Adenoma and Adenocarcinoma

Adenomas and adenocarcinomas are usually located in the proximal small bowel. Approximately 70 % of adenomas and adenocarcinomas occur in the duodenum or jejunum. Adenomas are classified histologically as tubular, tubulovillous, or villous. Endoscopic biopsy is recommended, and endoscopic mucosal resection can also be tried in select cases.

Small bowel adenomas may be associated with familial adenomatous polyposis, and the polyps may be multiple or laterally spreading (Fig. 29.1). Diagnosis of sporadic small bowel adenomas is very rare because the tumors seldom cause clinical symptoms, and screening enteroscopy is not usually performed. They are frequently flat and occasionally laterally spreading (Fig. 29.2).

Adenocarcinoma of the small intestine is similar to colon cancer. It grows circumferentially or exophytically and frequently presents with ulcerations (Fig. 29.3). Obstructive symptoms usually occur very late because the food materials are liquid in the small bowel. Small bowel adenocarcinoma may be associated with small bowel Crohn's disease or celiac disease.

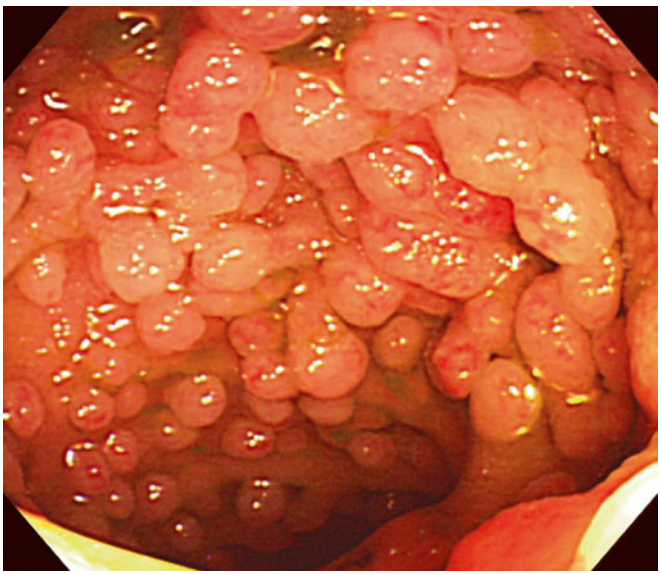

**Fig. 29.1** Numerous ileal adenomatous polyps in a patient with familial adenomatous polyposis

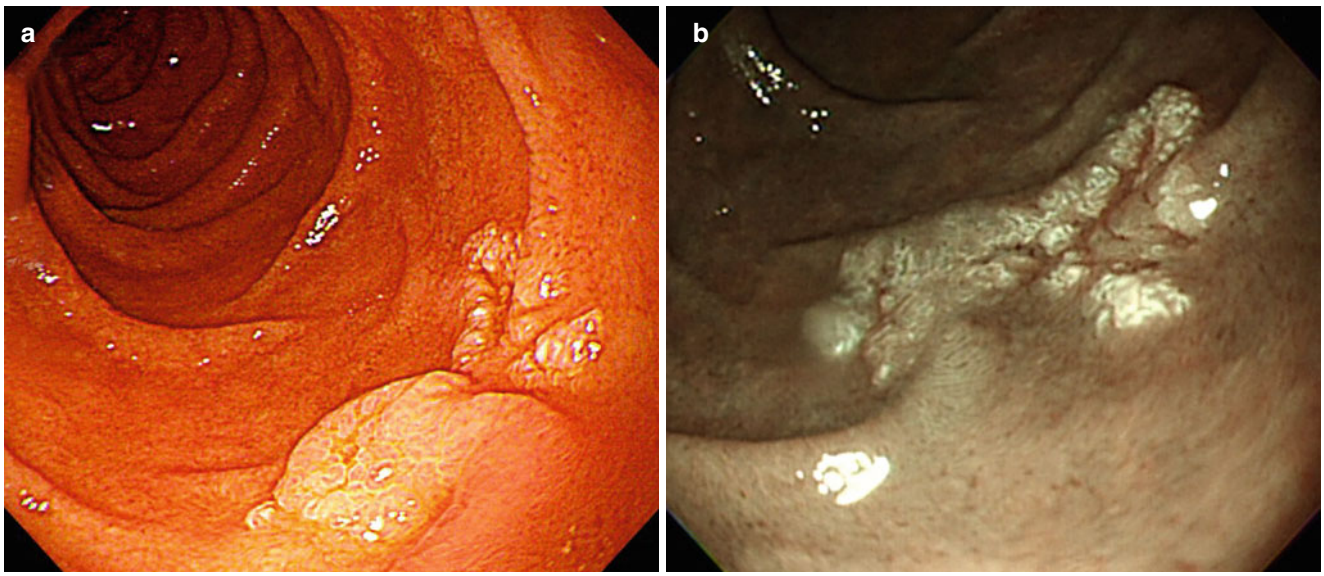


**Fig. 29.2** A sporadic adenoma of the proximal jejunum. The tumor is frequently flat, laterally spreading, and pale in color. (**a**) White-light endoscopy. (**b**) NBI endoscopy

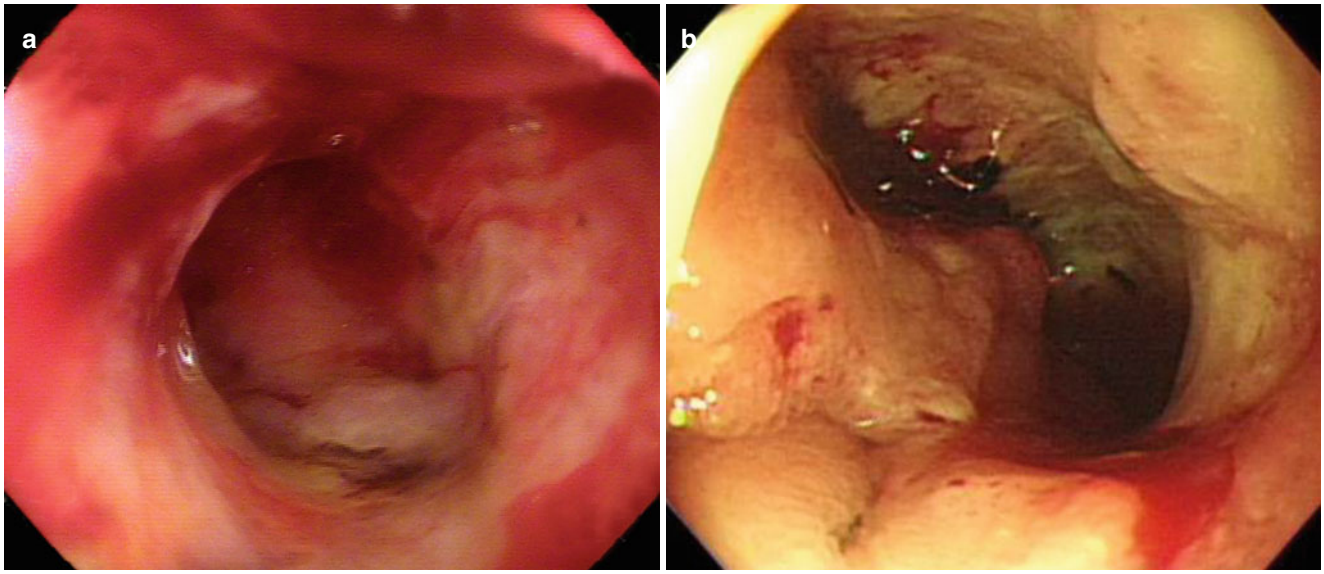


**Fig. 29.3** Adenocarcinoma of the jejunum. (**a**) Circumferential narrowing of the lumen by an ulceroinfiltrative mass is noted. (**b**) Luminal obstruction by a jejunal adenocarcinoma

### 29.2.2 Neuroendocrine Tumor (Carcinoid Tumor)

The small bowel (particularly the ileum) is the second most common location for neuroendocrine tumors next to the appendix. They usually appear as submucosal mass lesions, occasionally with ulcerations (Fig. 29.4). Although 86 % of small bowel carcinoids secrete serotonin, the typical carcinoid syndrome is rare. Three-fourths of primary small bowel carcinoids are <1.5 cm at the time of diagnosis, but approximately 30 % already have multifocal lesions at the time of diagnosis. Lesions less than 1 cm rarely metastasize.

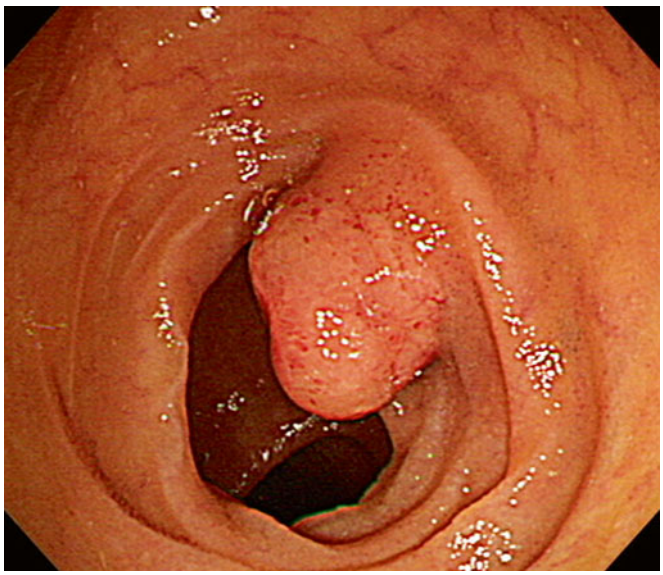

**Fig. 29.4** A neuroendocrine tumor in the terminal ileum. The tumor presents with the typical appearance of submucosal tumors. It has a surface erosion and a diameter of 1.5 cm. Invasion to the proper muscle layer was observed in the final pathology

### 29.2.3 Hamartoma

Hamartomatous polyps can be found in the small bowel, and they are usually pedunculated. The most common syndrome associated with hamartomatous polyposis is Peutz-Jeghers syndrome, which is an autosomal recessive condition characterized by benign multiple hamartomatous polyps of the gastrointestinal tract and pigmentations of the oral mucosa. A recent meta-analysis found up to a 13 % lifetime risk for the development of small intestinal cancer in patients with Peutz-Jeghers syndrome.

Hamartomas are frequently lobulated and covered with non-tumorous epithelium. Endoscopic resection of large polyps in the deep small bowel is possible by device-assisted enteroscopy or push enteroscopy (Fig. 29.5).

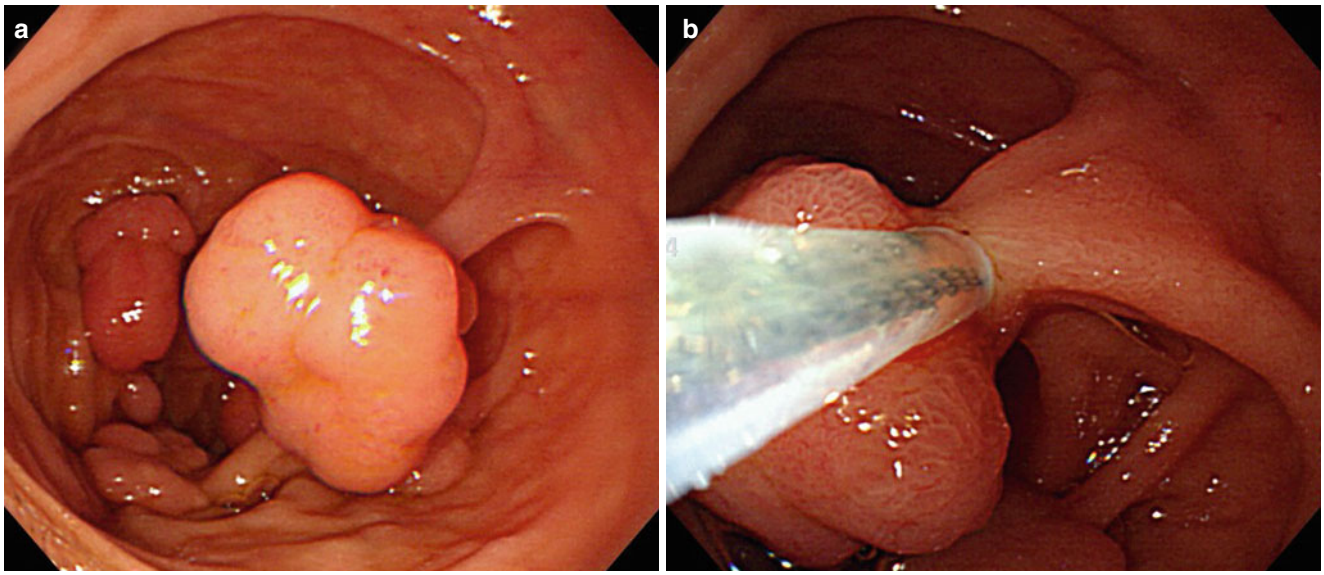


**Fig. 29.5** Multiple jejunal hamartomatous polyps in a patient with Peutz-Jeghers syndrome. (**a**) Polyps are varied in size and may be sessile or pedunculated. The head of the polyp is usually lobulated, but tumorous pit patterns are not observed. (**b**) The development of device-assisted enteroscopy enabled the performance of polypectomy without laparotomy

### 29.2.4 Stromal Tumors

GISTs are the most common stromal tumors in the small intestine. More than 85 % of tumors previously called leiomyoma or leiomyosarcoma are now known to be GISTs. GISTs originate from the interstitial cell of Cajal, the pacemaker cell in the myenteric plexus. GISTs appear as submucosal masses but sometimes grow as subserosal masses. GISTs are also the most common bleeding tumors in the small bowel. Bleeding usually occurs from central necrosis and ulceration (Fig. 29.6). Small bowel GISTs may grow to a large size without any clinical symptoms.

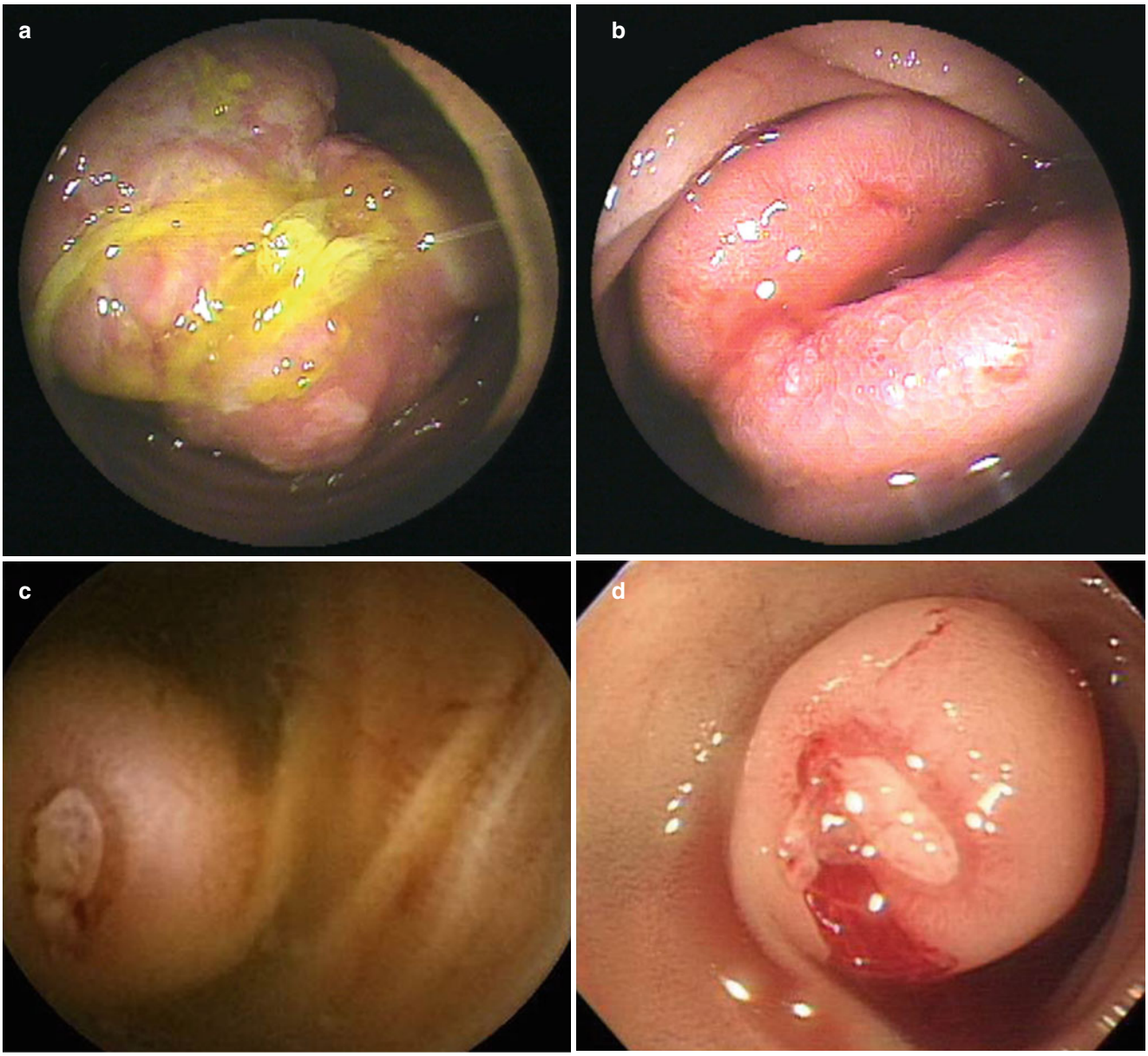


**Fig. 29.6** Various endoscopic features of small bowel GISTs. (**a**) A large GIST with surface erosions and exudation. (**b**) A GIST with a deep central ulceration. (**c**, **d**) Capsule endoscopy and single-balloon enteroscopy findings in a GIST with ulceration in the jejunum

### 29.2.5 Lipoma

Lipomas originate from the submucosal fat tissue. These tumors are usually soft, and yellow fat tissue can show through the mucosal layer (Fig. 29.7). They are typically pliable when stretched with biopsy forceps. "Pillow sign" means that the center of the tumor is easily indented when pushing the center of the tumor with forceps. The lipoma is the most common cause of adult intussusception.

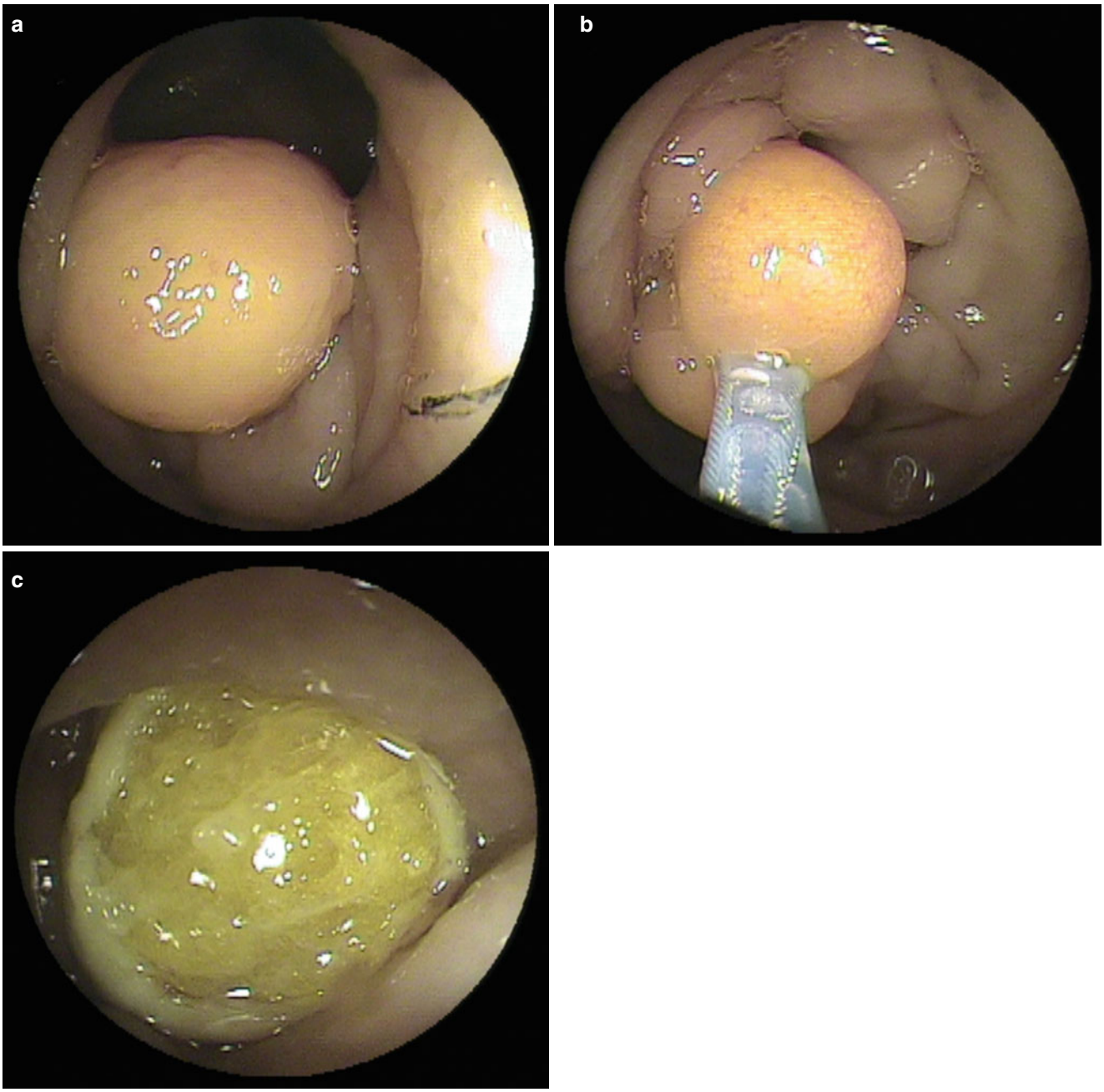


**Fig. 29.7** A jejunal lipoma. (**a**) The surface of the tumor is covered with normal mucosa, and yellow fat tissue is seen through the overlying mucosal layer. (**b**, **c**) Unroofing: snaring the top or middle of the tumor usually induces the exposure of the internal adipose tissue and results in the resolution of the tumor

### 29.2.6 Lymphoma

The gastrointestinal tract is the most common extranodal site involved by lymphoma, and the small bowel is the second most frequent site following the stomach. Lymphoma of the small intestine tends to occur at the distal jejunum or ileum. Most gastrointestinal lymphomas have a B cell origin, including the mucosa-associated lymphoid tissue (MALT) type (marginal zone B cell lymphoma, Fig. 29.8), diffuse large B cell lymphoma, mantle cell lymphoma, follicular lymphoma, Burkitt lymphoma, and immunoproliferative lymphoma. T cell lymphomas are rare and often associated with celiac disease.

Capsule endoscopy is useful for the detection of small bowel lymphoma, and device-assisted enteroscopy enables the avoidance of surgical tissue procurement.

Only a few studies have reported on the gross types of small bowel lymphoma: the tumorous type is most common, followed by the infiltrative type, polypoid type, and ulcerative type. However, the gross appearance of small bowel lymphoma is varied, and mass formation, ulceration, infiltration, and exudation can be observed in a tumor (Fig. 29.9). The elevated margin of the tumor without ulceration may be covered with normal mucosa. Lymphomas are usually less firm and less fragile than adenocarcinomas. Associated complications are stricture, bleeding, perforation, and fistula (Fig. 29.10).

Enteropathy-type T cell lymphoma is a rare and aggressive malignant tumor that originates from intraepithelial T cells. It commonly arises in the proximal jejunum. Typical endoscopic findings of enteropathy-type T cell lymphoma are multiple shallow ulcerations and diffuse thickening of mucosa with coarse and fine granular elevations like a mosaic (Fig. 29.11).

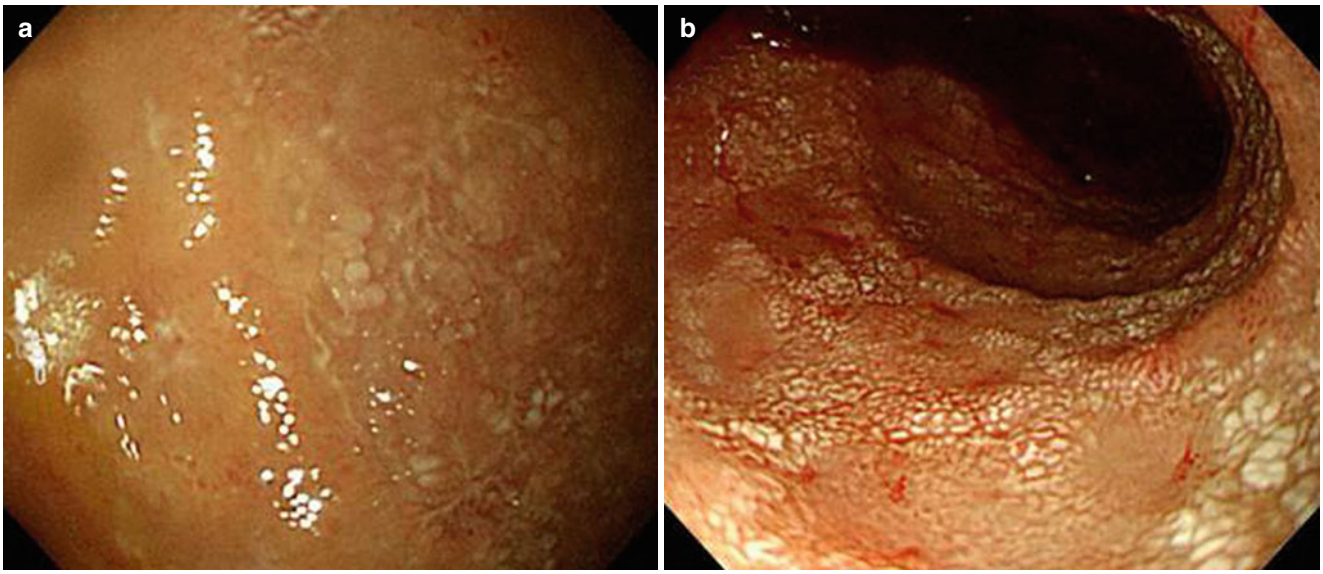


**Fig. 29.8** MALT lymphoma of the jejunum. (**a**, **b**) Flattening of villi, whitish spots, edema, friability, and shallow ulcers are noted

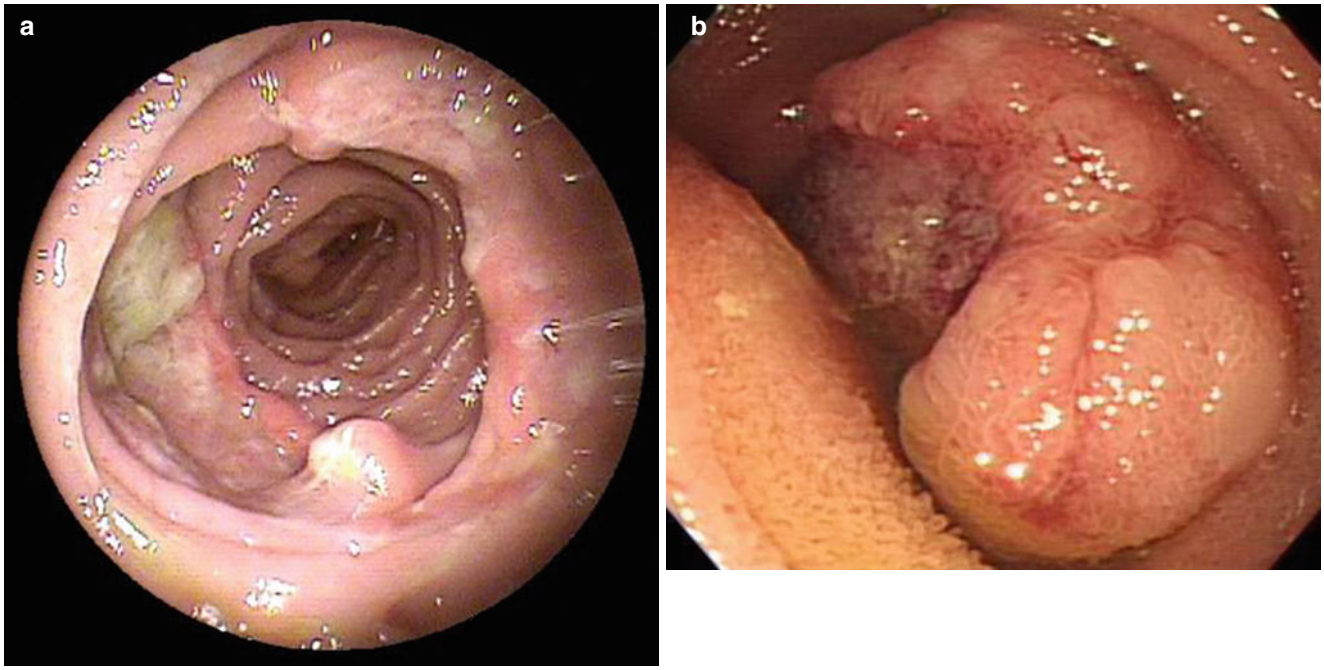


**Fig. 29.9** Diffuse large B cell lymphomas. (**a**) Nodular masses with ulcerations, white exudations, and thick everted margins at the distal jejunum. (**b**) A nodular mass with central ulceration in the ileum

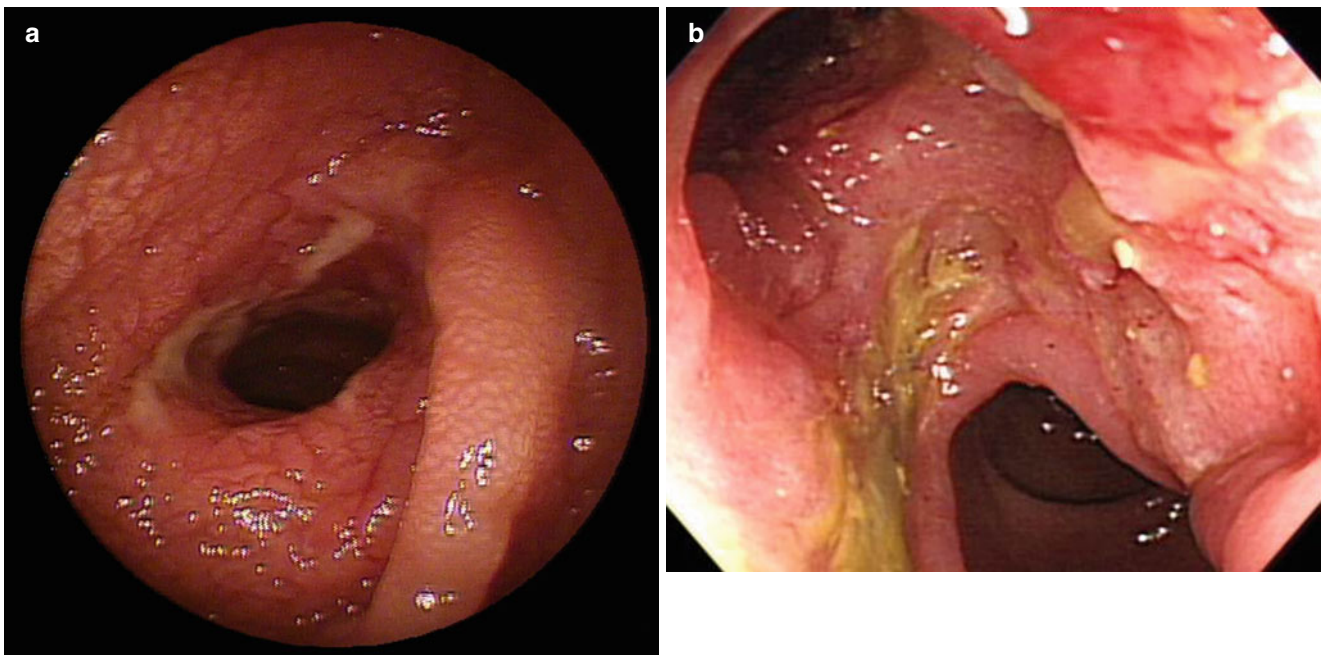


**Fig. 29.10** Complicated lymphomas. (**a**) Luminal stricture by a lymphoma infiltration, (**b**) Enterocolic fistula by a lymphoma: a view from the descending colon

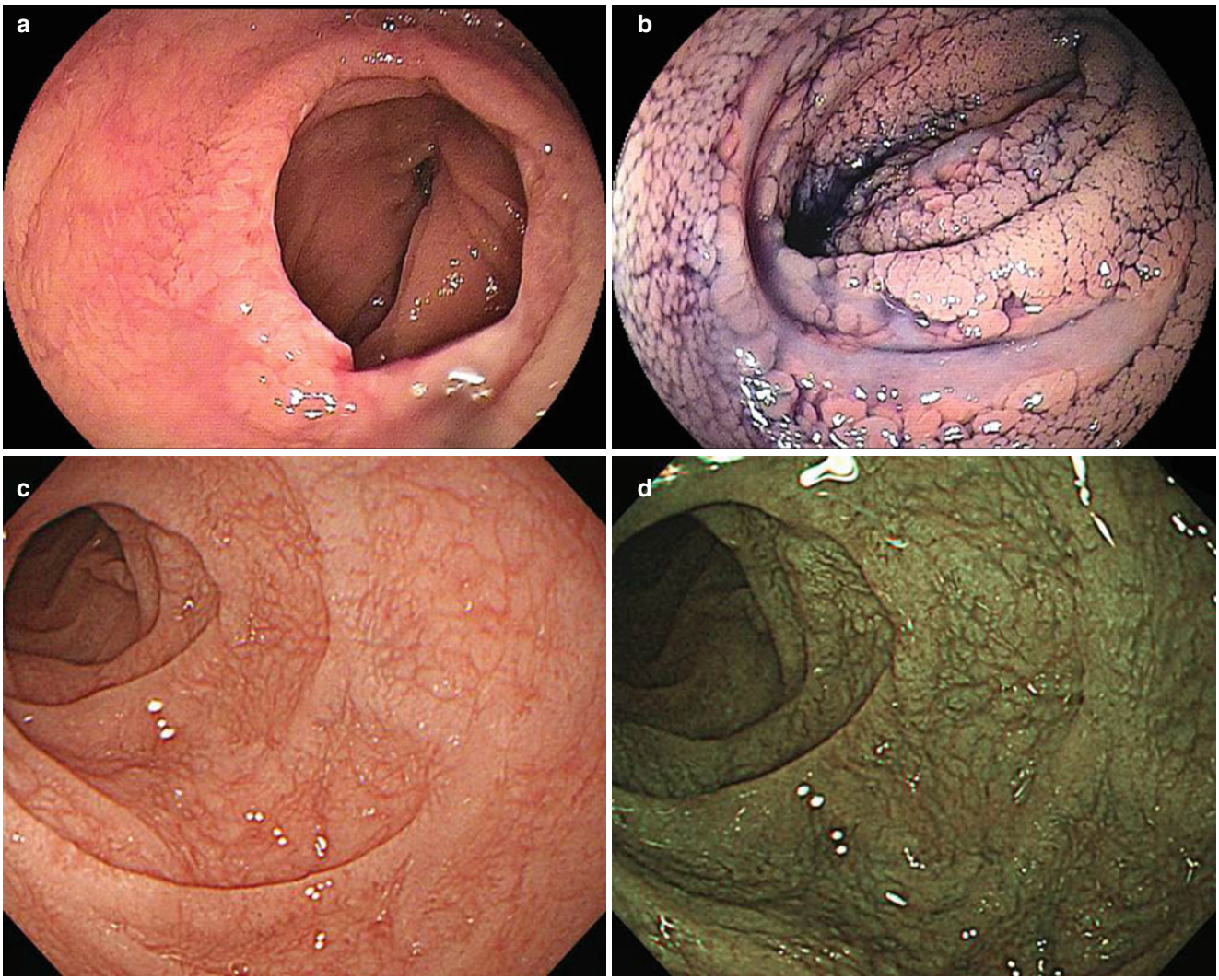


**Fig. 29.11** An enteropathy-type T cell lymphoma of the proximal jejunum. (**a**) Double-balloon enteroscopy shows multiple tiny, shallow ulcers with edema and fusion of the villi. (**b**) Double-balloon enteroscopy with indigo carmine spray. (**c**) Push enteroscopy also shows thickened and mosaic-like mucosa. (**d**) Push enteroscopy with NBI enhances mucosal granularity

### 29.2.7 Vascular Tumors

These will be discussed in Chap. 24.

### 29.2.8 Lymphangioma

Lymphangioma is a benign tumor of the lymphatic system, and it rarely occurs in the small bowel. Small bowel lymphangiomas are usually asymptomatic but have occasionally been reported as a cause of GI bleeding. Lymphangioma is usually observed as a whitish-yellow, cystic, submucosal mass (Fig. 29.12).

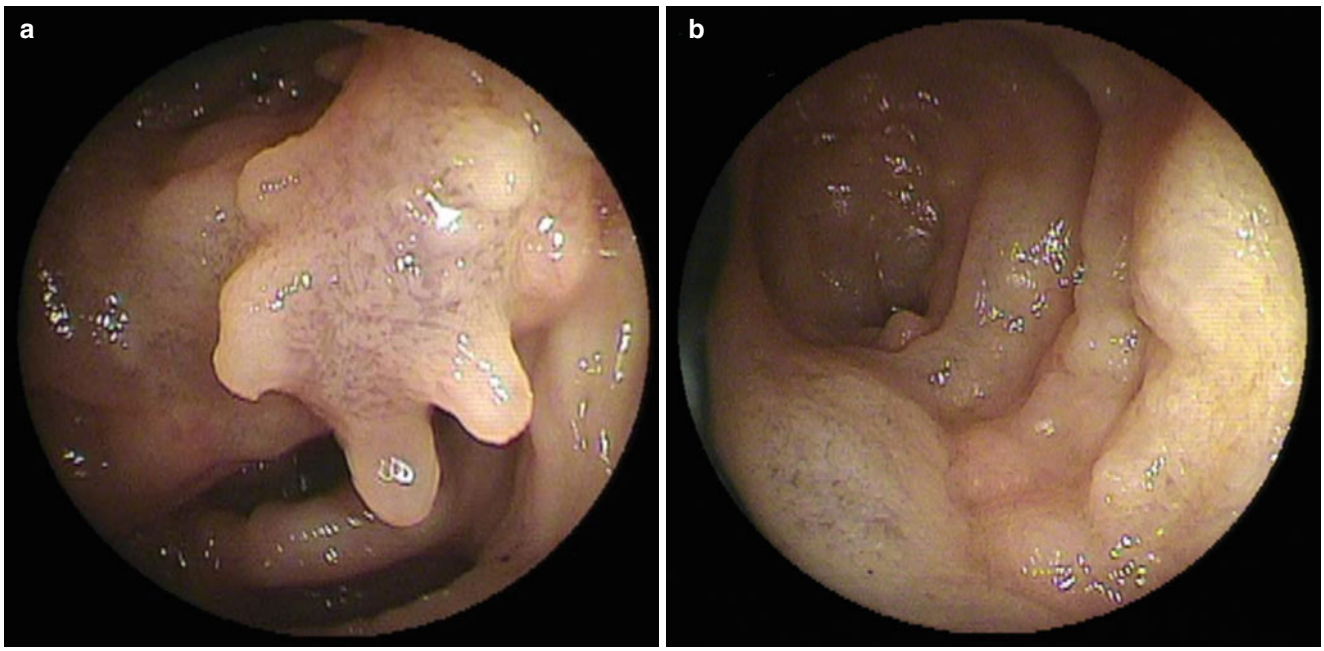


**Fig. 29.12** (**a**, **b**) Ileal lymphangiomas in a patient with Gorham's disease, which is characterized by nonmalignant proliferation of vascular or lymphatic structures that results in progressive bone destruction and often extends into the surrounding soft tissues. Gastrointestinal involvement in Gorham's disease is rare

### 29.2.9 Inflammatory Fibroid Polyp

The inflammatory fibroid polyp is not a true neoplastic tumor. It is a nonneoplastic proliferating lesion of the gastrointestinal tract. Its pathogenesis is unknown, but an abnormal inflammatory response may be associated. The lesions are most commonly encountered in the distal stomach and the terminal ileum. They are mostly submucosal and may be sessile or pedunculated (Fig. 29.13). Histologically, the polyps consist of fibrous tissues including vessels, spindle cells, and inflammatory cells, particularly eosinophils. Patients with inflammatory polyps are usually asymptomatic until complications such as a small bowel obstruction or bleeding occur.

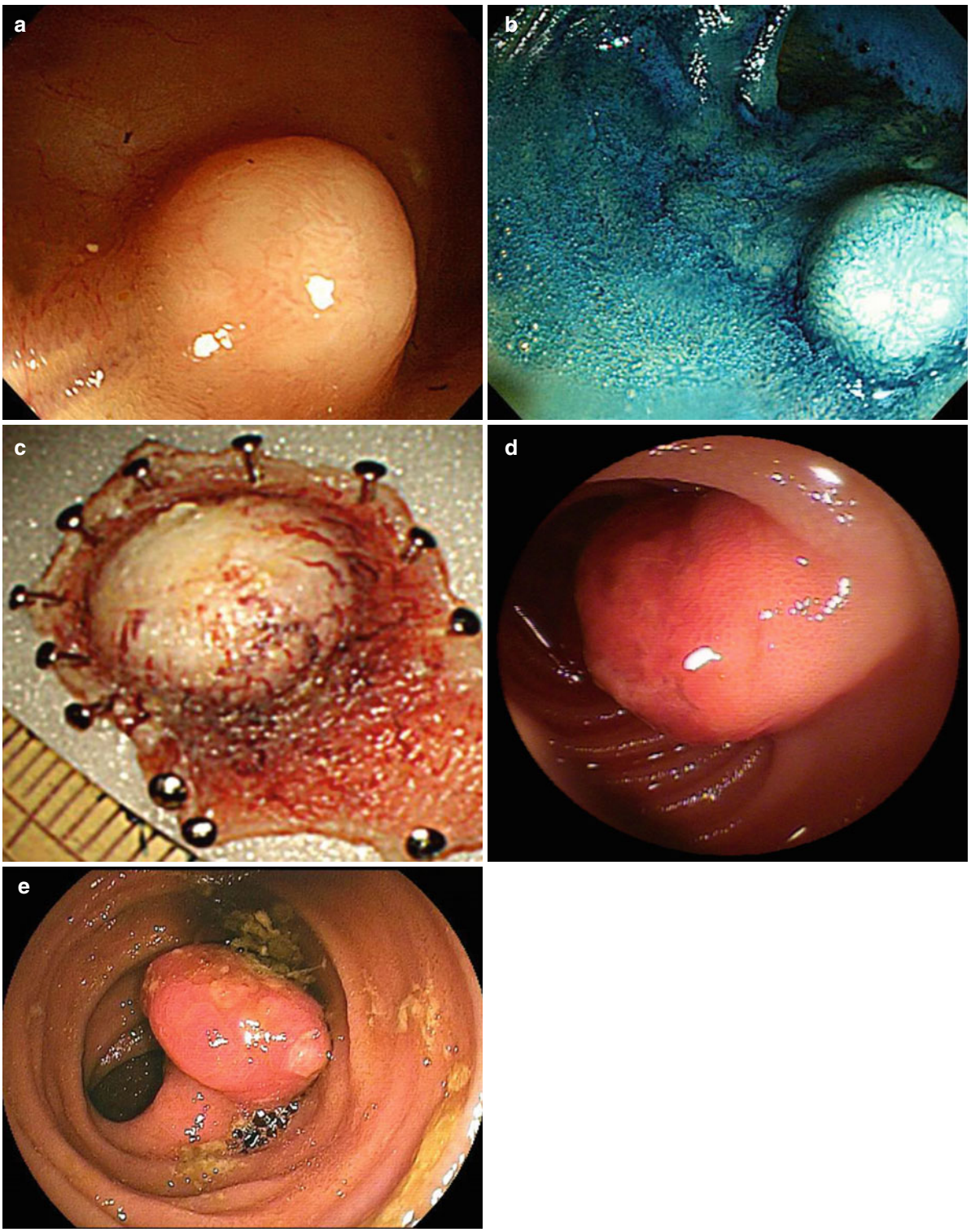


**Fig. 29.13** Inflammatory fibroid polyps. (**a**) The lesions usually feature as submucosal lesion. (**b**) After indigo carmine spray. (**c**) The polyp was removed by EMR. The specimen was fixed with pins to make the submucosal layer face the top. (**d**) Another inflammatory fibroid polyp with surface hemorrhage. The mucosal surface is eroded. (**e**) An inflammatory fibroid polyp with a stalk

### 29.2.10 Metastatic Tumors

Metastatic tumors of the small bowel occur in two ways: by direct invasion from adjacent organs or by distant metastasis. Direct invasion can occur from the stomach, pancreas, liver, kidneys, adrenals, uterine cervix, and ovaries. Distant metastasis can occur from melanomas, breast cancers, lung cancers, renal cancers, and testicular cancers (Fig. 29.14). Differential diagnosis from primary small bowel cancer may not be easy.

Melanoma is the most common cancer causing distant metastasis to the small bowel. Metastatic melanoma is often pigmented, but this is not a necessary feature (Fig. 29.15).

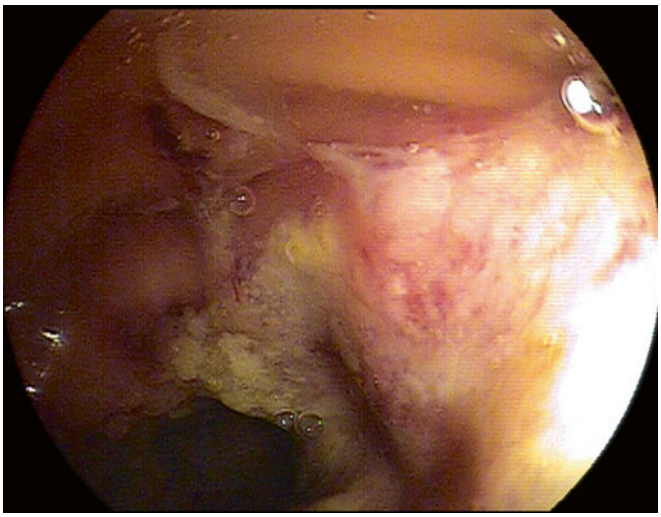

**Fig. 29.14** Small bowel metastasis from lung cancer. Peroral double-balloon enteroscopy, which was performed due to the patient's obstructive symptoms, showed an ulceroinfiltrative lesion of the mid-ileum, and biopsy confirmed metastatic squamous cell cancer. Endoscopic differential diagnosis from primary small bowel cancer is occasionally challenging

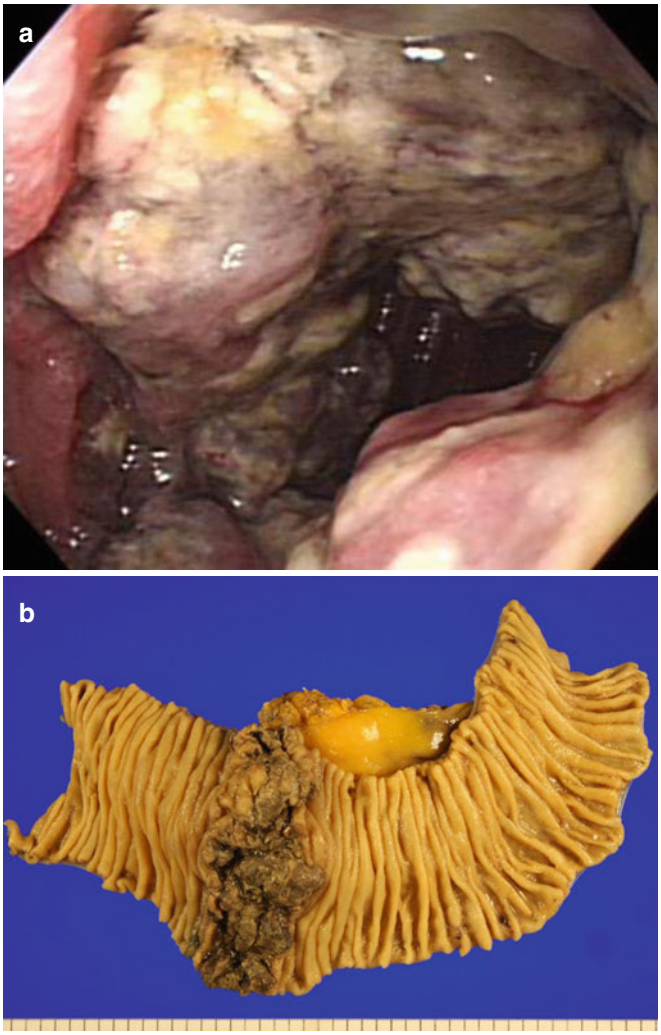


**Fig. 29.15** Metastatic melanoma of the jejunum. The patient was diagnosed with a malignant melanoma of the right foot 5 years prior. Scattered pigmentation on the tumor surface is noted. (**a**) Single-balloon enteroscopy. (**b**) The surgical specimen

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