Idiopathic Inflammatory Bowel Diseases

19

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Contents

Idiopathic inflammatory bowel disease (IBD) includes ulcerative colitis (UC), Crohn's disease (CD), and gastrointestinal (GI) involvement of Behçet's disease (BD). UC is characterized by continuous and diffuse inflammation of colon which involves rectum in almost all cases. In active UC, blurring of vascularity, erythema, granularity, edema, friability, exudates, mucosal bleeding, and ulcers can be observed. CD can involve any site of GI tract and is characterized by transmural inflammation of the bowel. Aphthous erosions and ulcerations can be observed in the early stage, and linearly arranged aphthous erosions/ulcerations are characteristic of CD. With progress of disease, various ulcers, longitudinal ulcers, and cobblestone appearance develop. CD is more commonly complicated by stricture or perforation of bowel. GI BD is characterized by one or a few large, discrete, and deep ulcers located in the ileocecal area. In this chapter, the clinical and endoscopic features of UC, CD, and GI BD will be presented.

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19.1 Definition

Idiopathic inflammatory bowel diseases (IBD) are defined as chronic inflammatory diseases of gastrointestinal (GI) tract with unknown etiopathogenesis. Generally, idiopathic IBDs encompass two forms: ulcerative colitis (UC) and Crohn's disease (CD). In addition, GI involvement of Behçet's disease (BD) can also be classified as idiopathic IBD. In this chapter, the clinical and endoscopic features of UC, CD, and GI BD will be presented.

19.2 Indication of Colonoscopy in Idiopathic IBD

Colonoscopy makes it possible to directly observe the ileocolonic mucosal change and to get tissues for pathologic diagnosis in cases of suspicious idiopathic IBD. Therefore, colonoscopic evaluation has most important role in correct diagnosis and differential diagnosis of IBD. However, colonoscopy is an invasive procedure and could cause complications such as bowel perforation, bleeding, or toxic megacolon, especially for active IBD. Therefore, clinicians should be fully aware of indications and contraindications of colonoscopy for IBD (Table 19.1) [1].

Table 19.1 Indications and contraindications of colonoscopy for IBD

Indication	Contraindication
Diagnosis of IBD	Patient's refusal
Differential diagnosis of IBD	Poor cooperation
Evaluation of newly developed symptoms	Suspected bowel perforation
Evaluation of disease activity	Fulminant colitis
Evaluation of therapeutic effect	
Assessment of radiologic abnormalities	
Diagnosis of postoperative recurrence in Crohn's disease	
Surveillance for dysplasia and cancer	
Therapy (dilatation of stricture, resection of dysplasia, etc.)	

19.3 Ulcerative Colitis

19.3.1 Clinical Manifestations

Ulcerative colitis (UC) is a chronic inflammatory bowel disease of colon involving mucosal and submucosal layers. The pathogenesis of UC is still unclear and there are no curative medical treatments for UC. After diagnosed usually in young age, disease activity of UC tends to wax and wane. The typical feature of UC is chronic history of bloody diarrhea, mucoid stool, urgency, and tenesmus. For active UC, induction of clinical remission with medical treatment is needed, and long-term maintenance treatment is mandatory for maintaining medically induced remission. Drugs such as 5-aminiosalicylic acids, corticosteroids, thiopurines, cyclosporine and anti-TNF-alpha agents are used for treating UC. Theoretically, UC can be cured by removing the whole colon. The current standard surgical procedure is total proctocolectomy with ileal pouch-anal anastomosis. However, impaired quality of life and development of pouchitis should be considered for decision-making for surgery.

19.3.2 Endoscopic Features

19.3.2.1 Extent of Disease

The typical endoscopic feature of UC is continuous inflammation without skip area and the involvement of rectum in nearly all cases [2]. The extent of UC can be classified as proctitis (inflammation involving up to 15 cm from anal verge), left-sided colitis (inflammation involving up to splenic flexure), and extensive colitis (inflammation involving more proximal area to splenic flexure). When the whole colonic mucosa from cecum to rectum is inflamed, it is called "pancolitis" (Fig. 19.1). The margin between inflamed mucosa and normal mucosa is usually clear (Fig. 19.2), but sometimes it is not

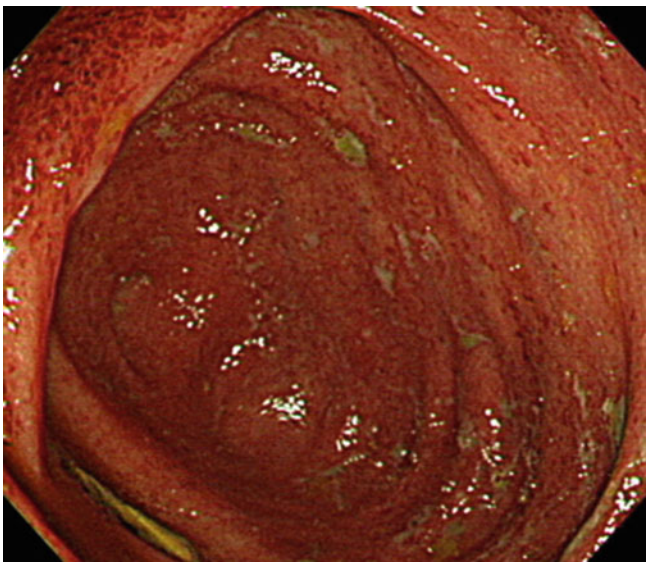


Fig. 19.1 Diffuse inflammation with multiple ulcers in patient with pancolitis

(Fig. 19.3). Rarely, inflammation of terminal ileum in patients with pancolitis can be observed, and it is called "backwash ileitis" (Fig. 19.4). In contrast to traditional concept of continuous inflammation, skip lesions in the proximal colon such as areas around the orifice of appendix were reported [3]. They are now named as "cecal patch" or "appendiceal orifice inflammation (AOI)" (Fig. 19.5). Inflammation of proximal colon and distal area such as rectum with grossly normal areas between proximal and distal areas is also considered as one of the endoscopic features of UC. Sometimes, atypical inflammatory mucosal change characteristic of UC in various colonic segments without continuity can also be observed, and they are also suggested to be one of the features of UC (Fig. 19.6). Therefore, traditional concept of continuous inflammation as a typical feature of UC is recently challenged.

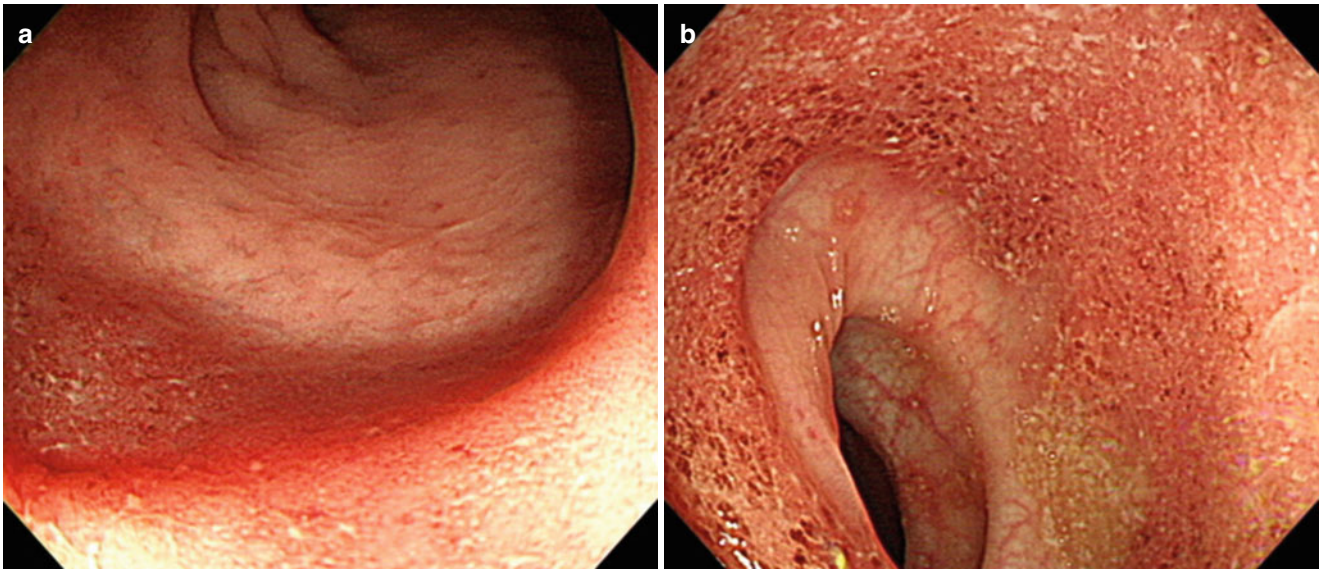


Fig. 19.2 Distinct margin between normal mucosa and inflamed mucosa. (**a**) Rectum. (**b**) Sigmoid colon

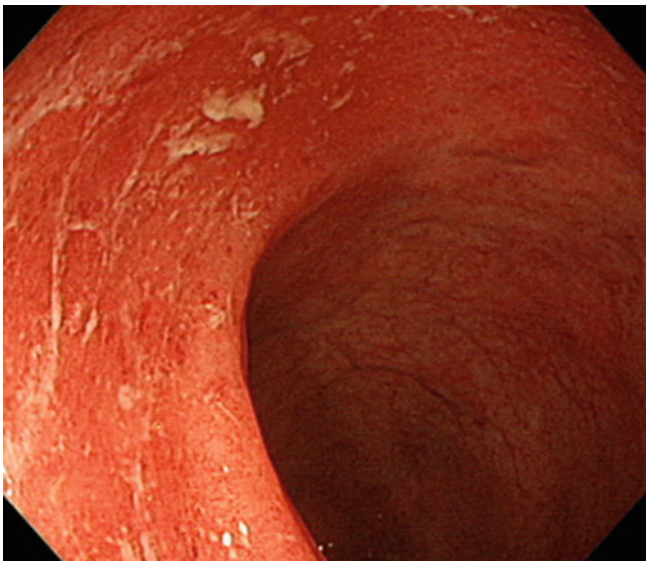


Fig. 19.3 Blurred margin between normal and inflamed mucosa is observed

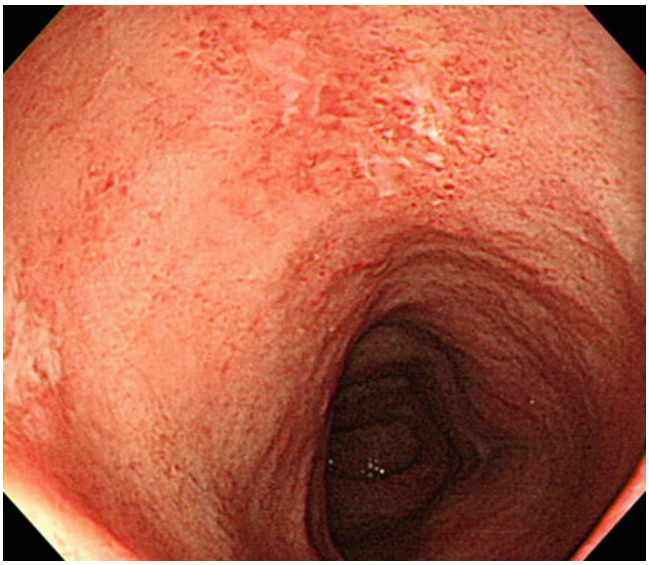


Fig. 19.4 Hyperemic change and shallow ulcers showing backwash ileitis

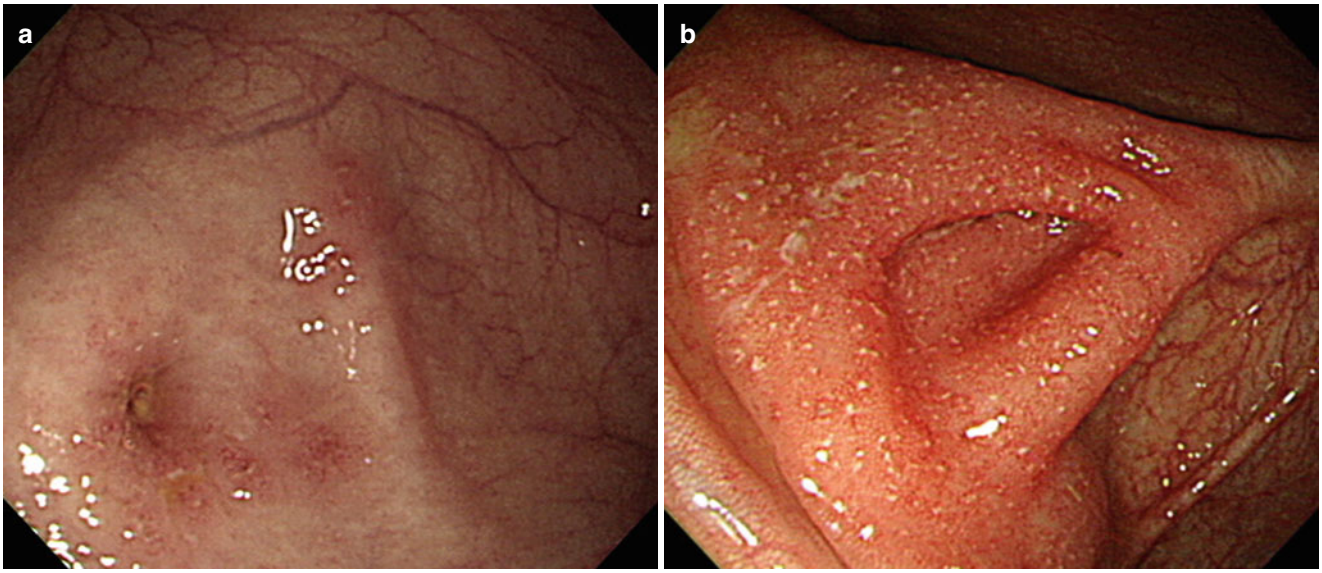


Fig. 19.5 Appendiceal orifice inflammation. (**a**) Mild inflammation in small area in appendiceal orifice. (**b**) More widespread inflammation

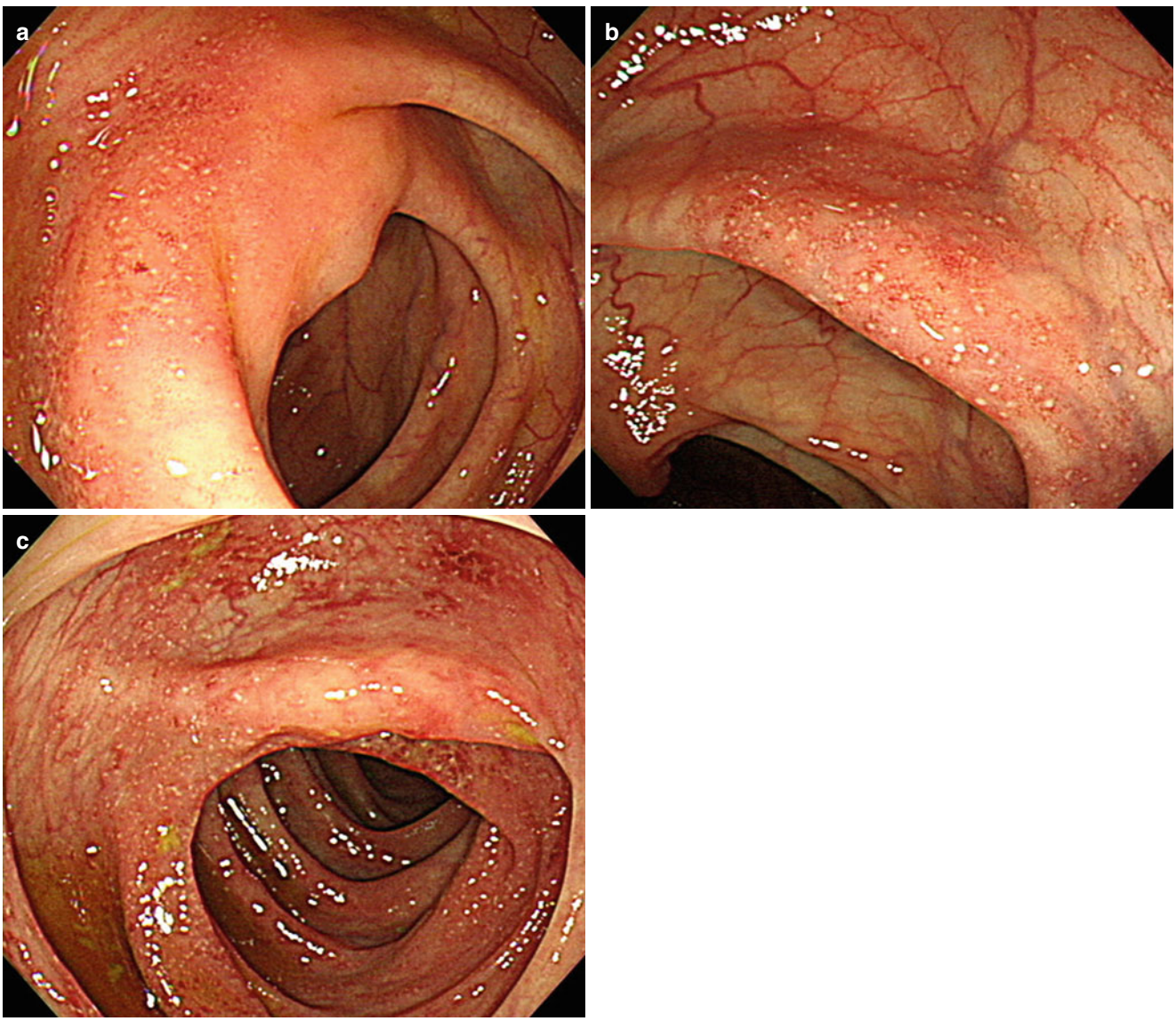


Fig. 19.6 Patchy inflammation in ascending colon without continuity. (**a**) Ascending colon just distal to ileocecal valve. (**b**) Proximal ascending colon. (**c**) Mid-ascending colon

19.3.2.2 Characteristic Endoscopic Features

Active Disease

There are no specific endoscopic features observed only in UC. Therefore, diagnosis of UC should be considered when the following endoscopic features are observed in patients with compatible clinical backgrounds:

- Blurring of vascularity (Fig. 19.7): Compared to normal colonic mucosa, blurring and irregularity of vascularity are observed from the early stage of inflammation. With more severe inflammation, normal vascularity disappears.
- Erythema (Fig. 19.8): Erythematous change of mucosa develops through congestion and dilatation of mucosal vasculature.
- Granularity (Fig. 19.9): Granularity means fine or coarse irregularity of mucosal surface in contrast to the glistening and even mucosa of normal colon.
- Edema (Fig. 19.10): With active inflammation, edematous mucosal change develops and it becomes more severe with the progression of inflammation. With severe edema, lumen of colon shows narrowed appearance. However, it is not true stenotic change.
- Friability: Inflamed mucosa shows easy bleeding to light touch. Spontaneous bleeding after luminal inflation (Fig. 19.11) and even spontaneous bleeding without any stimuli can develop with the progression of inflammation (Fig. 19.12).
- Exudates (Fig. 19.13): Mucopurulent exudates are frequently observed in UC. Exudates are more profuse with more severe inflammation.
- Bleeding (Fig. 19.14): Bleeding in UC is usually oozing bleeding from inflamed mucosa. Therefore, active spurting bleeding or massive bleeding from specific vessels is not common. Bleeding can develop after stimuli such as touch or pressure or spontaneously.
- Ulcers (Fig. 19.15): With light inflammation, the ulcers of UC are small and superficial. However, larger and deeper ulcers can be observed in more severe cases. In that stage, ulcers can show various sizes and shapes. Sometimes, ulcers show longitudinal direction like in CD. Not frequently, cobblestone-like appearance can be observed with deep longitudinal and transverse ulcers. However, the difference between UC and CD is the mucosal change around ulcers. In UC, mucosa around ulcers also shows inflammatory changes like hyperemia, blurred vascularity, granularity, and friability. However, the surrounding mucosa in Crohn's ulcers is usually not inflamed and shows nearly normal appearance. Therefore, ulcers in UC are called "inflammatory ulcers" and ulcers in CD are called "discrete ulcers" [3].
- Grading of severity: There are a lot of grading systems for inflammation in UC. One of the simple and commonly used systems is Mayo endoscopic subscoring system (Table 19.2) [4].

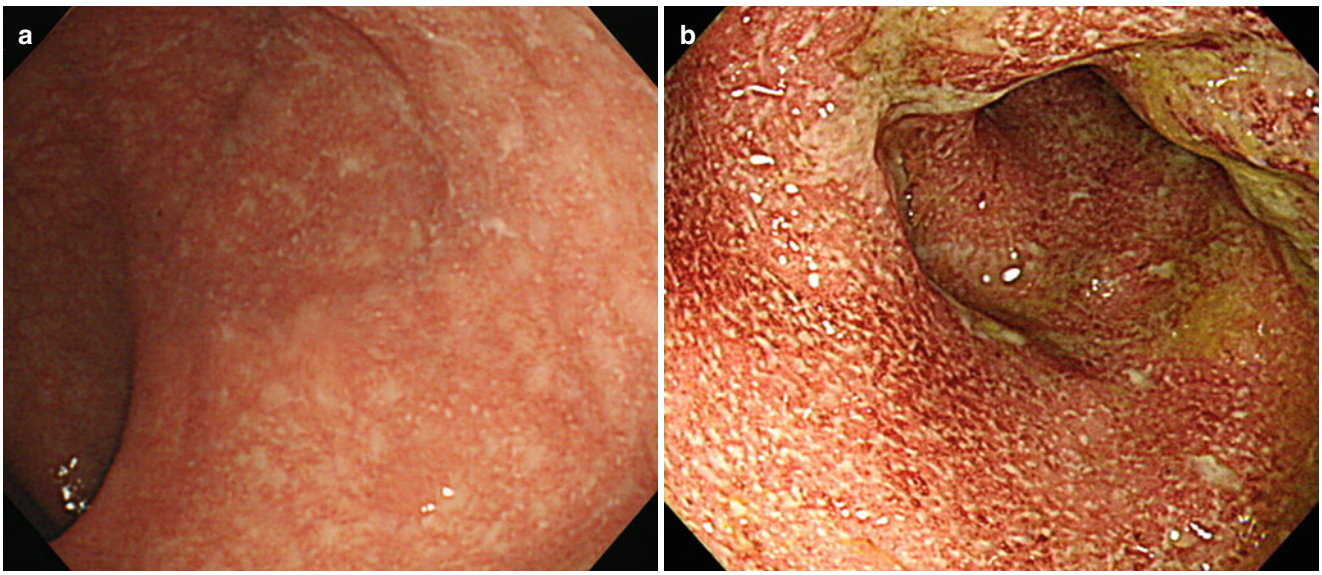


Fig. 19.7 Blurring and loss of vascularity. (**a**) With mild granular change and hyperemia of mucosa, subepithelial vascularity is blurred. (**b**) Vascularity is invisible with more severe inflammation

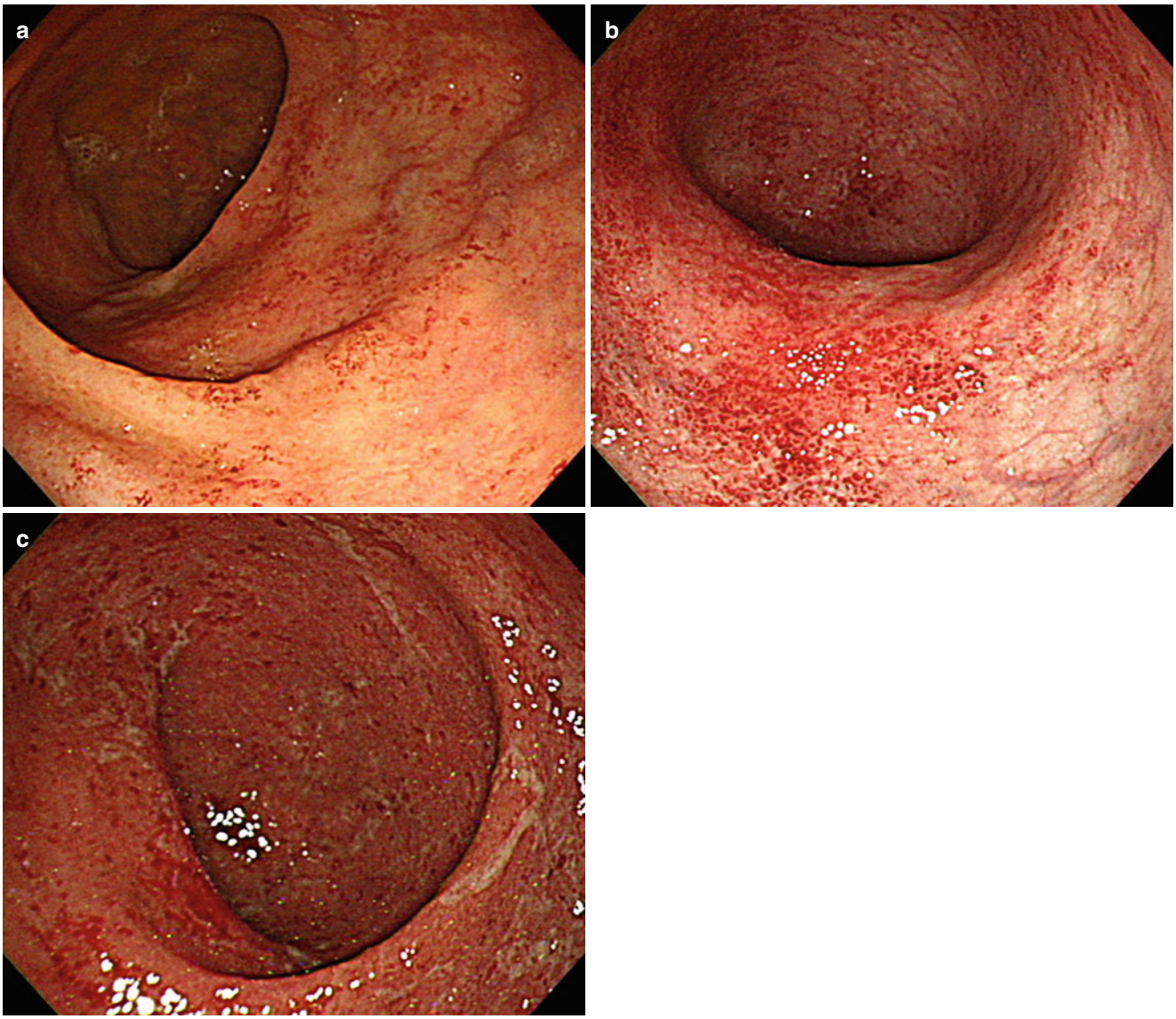


Fig. 19.8 Erythematous mucosal change. (**a**) Mild. (**b**) Moderate. (**c**) Severe

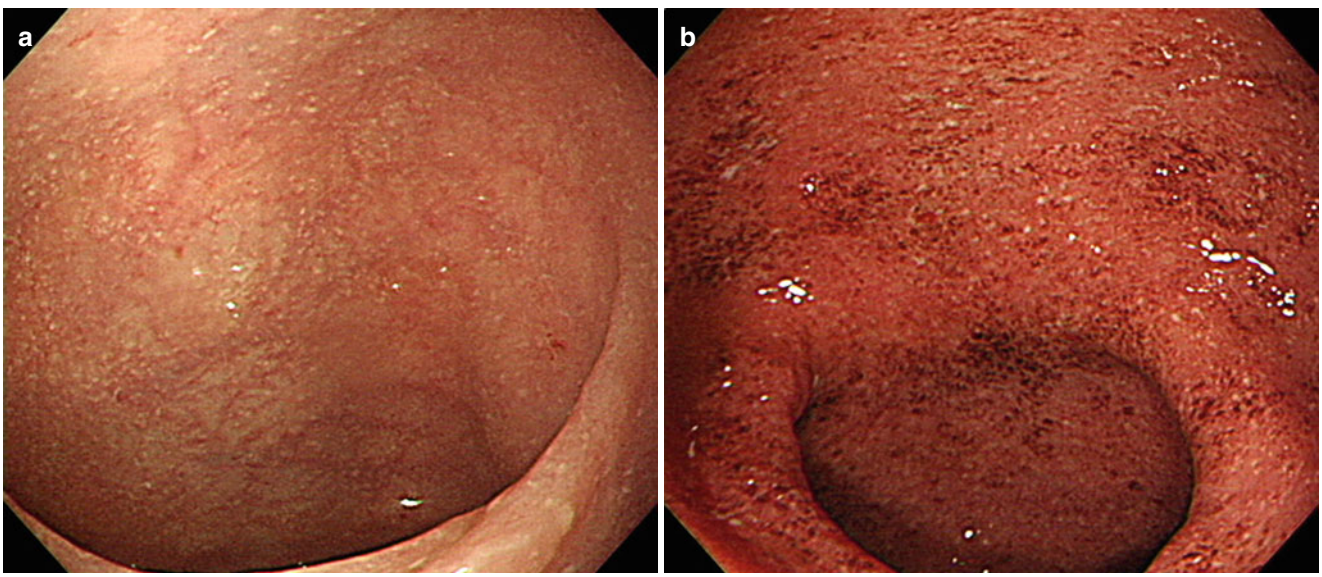


Fig. 19.9 Granular change. (**a**) Mild granularity. (**b**) Coarse granularity

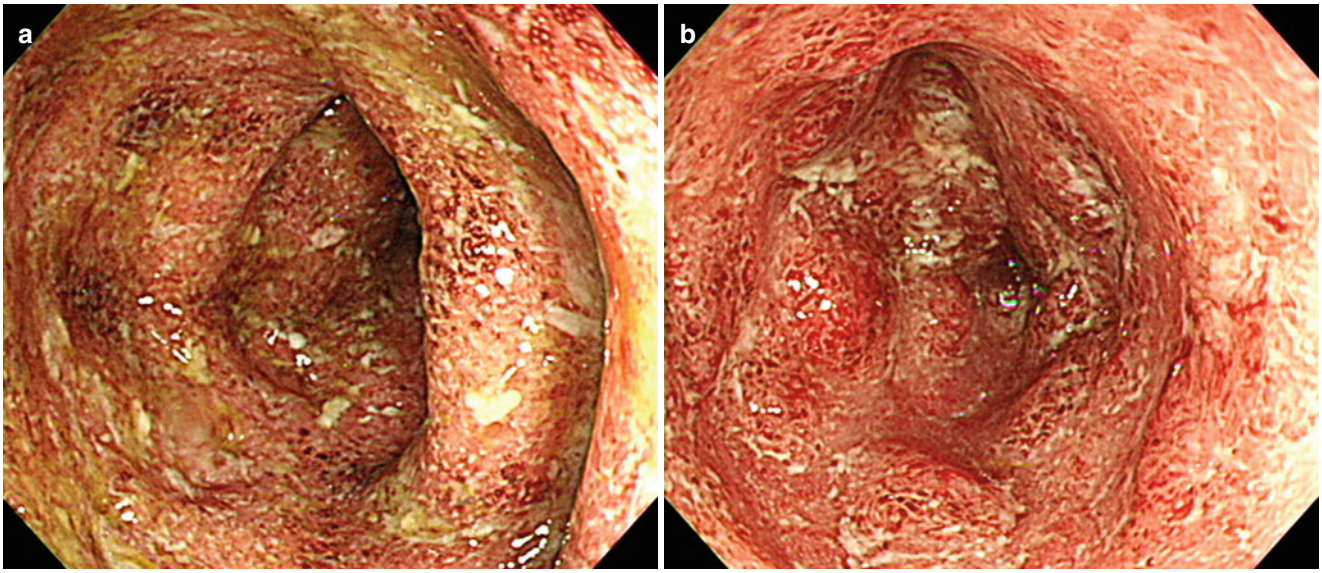


Fig. 19.10 Mucosal edema. (**a**) Severe edema with thickened mucosal folds. (**b**) Colonic lumen appears to be narrowed with mucosal edema

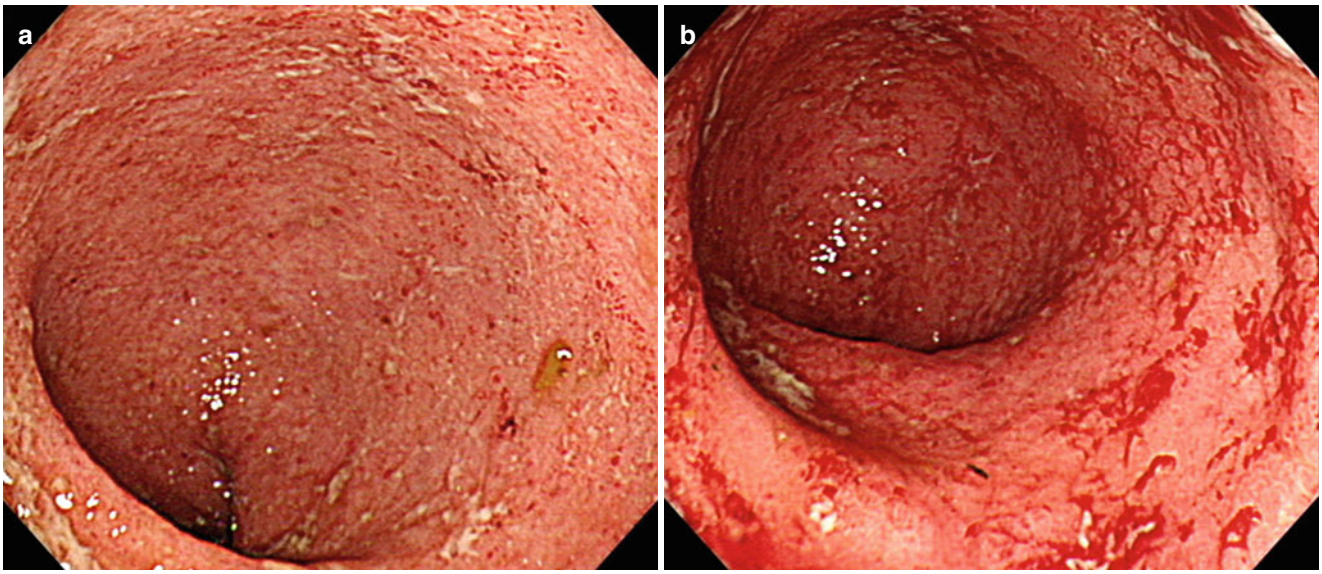


Fig. 19.11 Bleeding after luminal inflation with air. (**a**) Before inflation. (**b**) After inflation, multiple oozing bleeding spots are noted

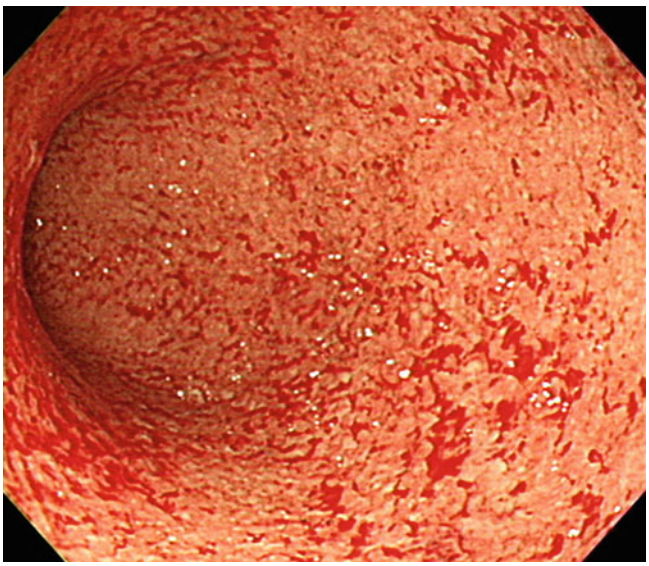


Fig. 19.12 Multifocal bleeding in patients with severe UC

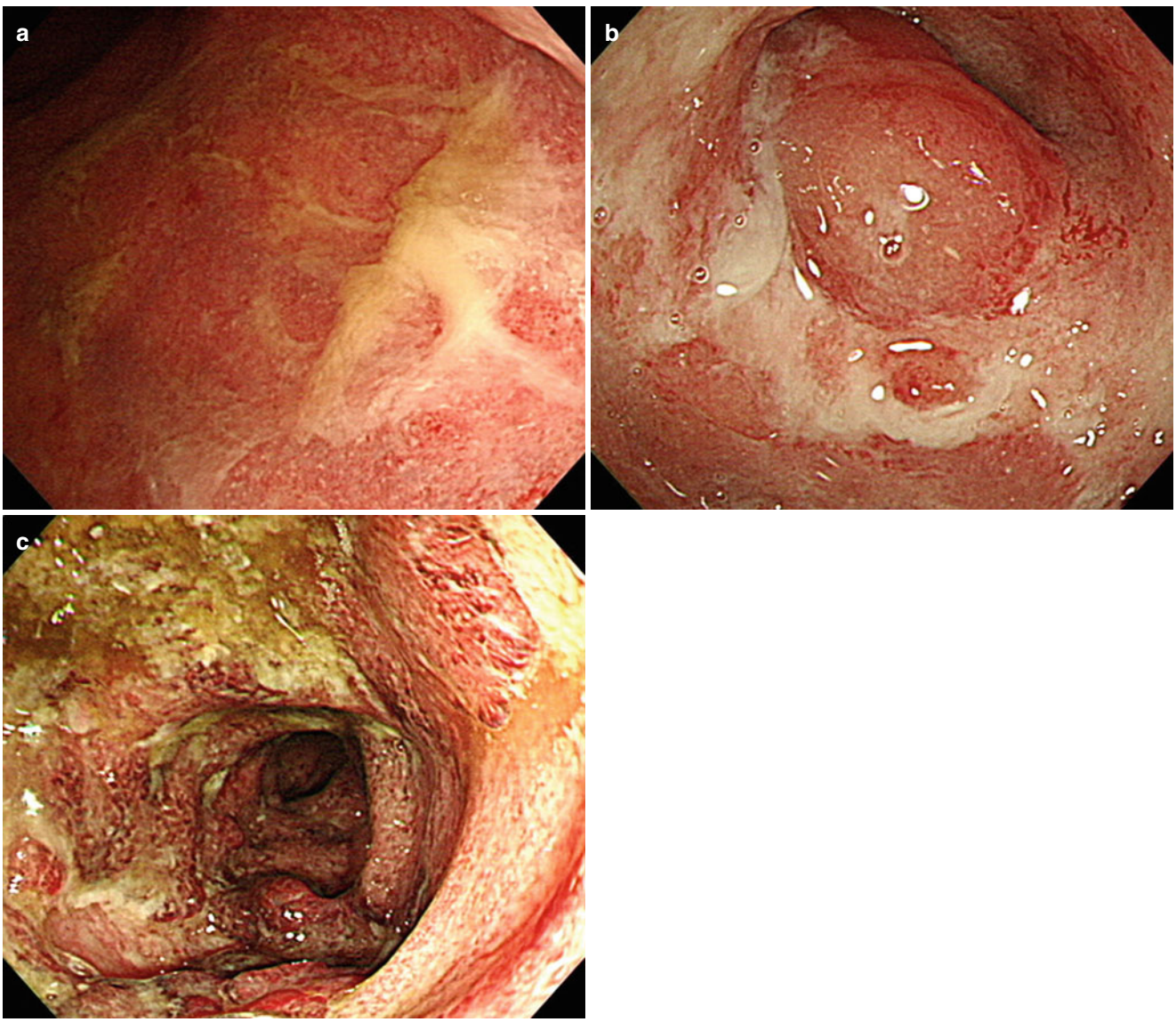


Fig. 19.13 Mucopurulent exudates in UC. (**a**) Mild. (**b**) Thicker exudates with mucosal friability. (**c**) Severe UC with profuse exudates

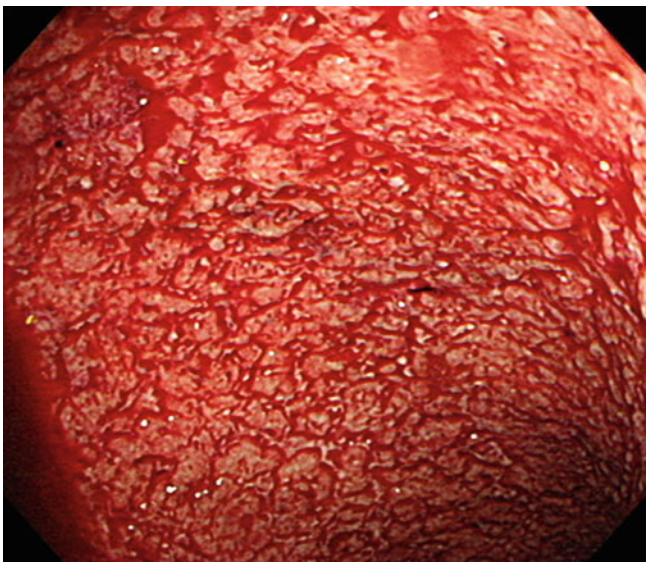


Fig. 19.14 Diffuse oozing bleeding are observed in severe UC

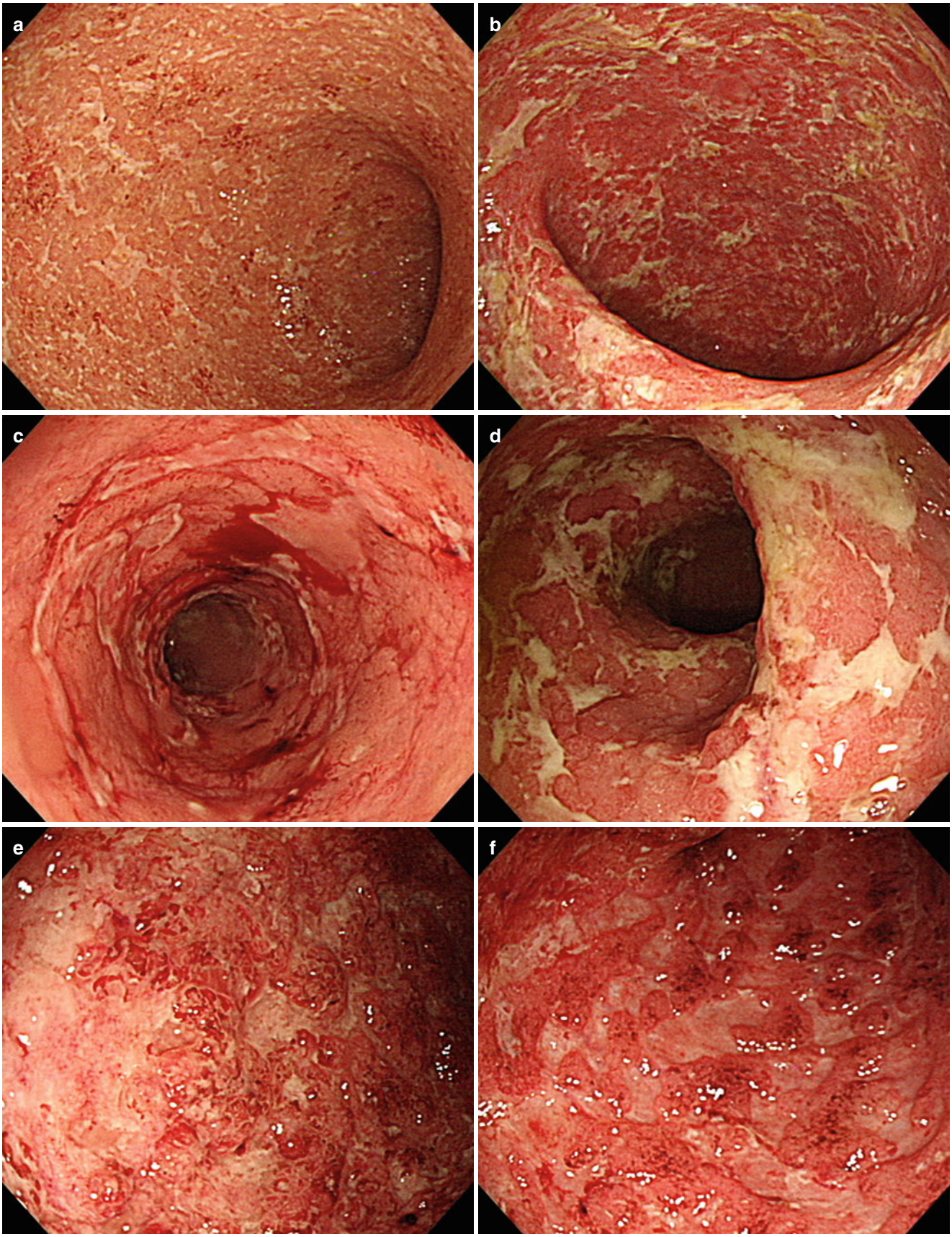


Fig. 19.15 Ulcers in UC. (**a**, **b**) Small superficial ulcers. (**c**, **d**) Larger and deeper ulcers. (**e**) Diffuse deep ulcers with inflamed surrounding mucosa. (**f**, **g**) Deep longitudinal ulcers. (**h**–**j**) Undermining ulcers with visible muscle layer. Remnant mucosa looks like islands in the background of severe ulcers. (**k**, **l**) Cobblestone appearance

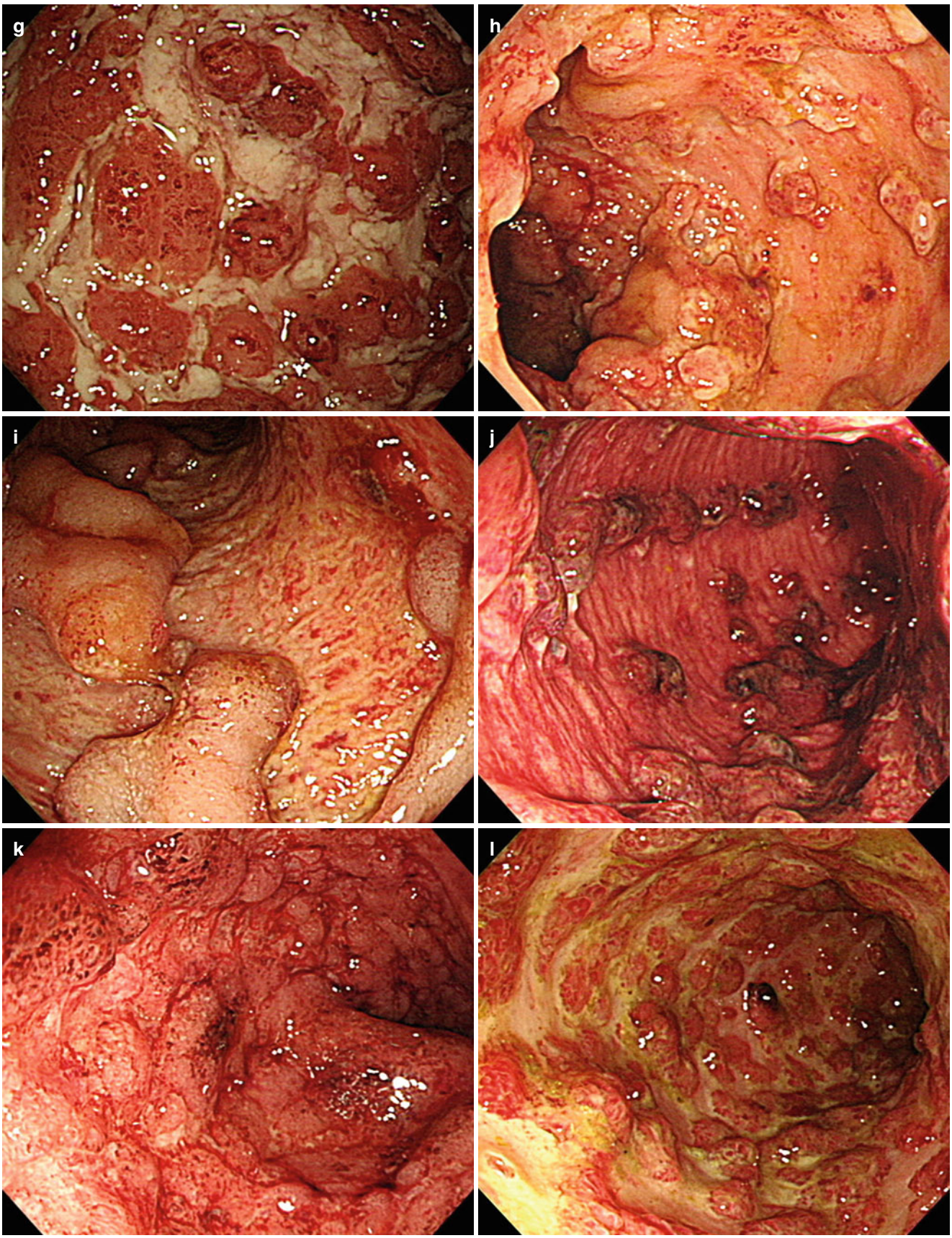


Fig. 19.15 (continued)

Table 19.2 Mayo endoscopic subscoring system

Score	Endoscopic findings
0	Normal or inactive disease
1	Mild disease (erythema, decreased vascular pattern, mild friability)
2	Moderate disease (marked erythema, lack of vascular pattern, friability, erosions)
3	Severe disease (spontaneous bleeding, ulceration)

Inactive Disease

With resolution of mild inflammation, the colonic mucosa shows nearly normal mucosal appearance, and it is even difficult to tell whether there was inflammation or not. However, after recovery from moderate to severe inflammation, the regenerated vascularity shows sparse and irregular appearance with white scar changes. Sometimes, between extensive scars, diverticular structures are formed and they are called "pseudodiverticulums" (Fig. 19.16).

After healing of ulcers, the remained mucosa sometimes can show polypoid appearance. These polypoid structures are called "inflammatory polyps" or "inflammatory pseudopolyps." Inflammatory polyps can show various sizes, shapes, numbers, and surface changes (Fig. 19.17). After recovering from deep undermining ulcers, mucosa can show bridge-like appearance (mucosal bridge).

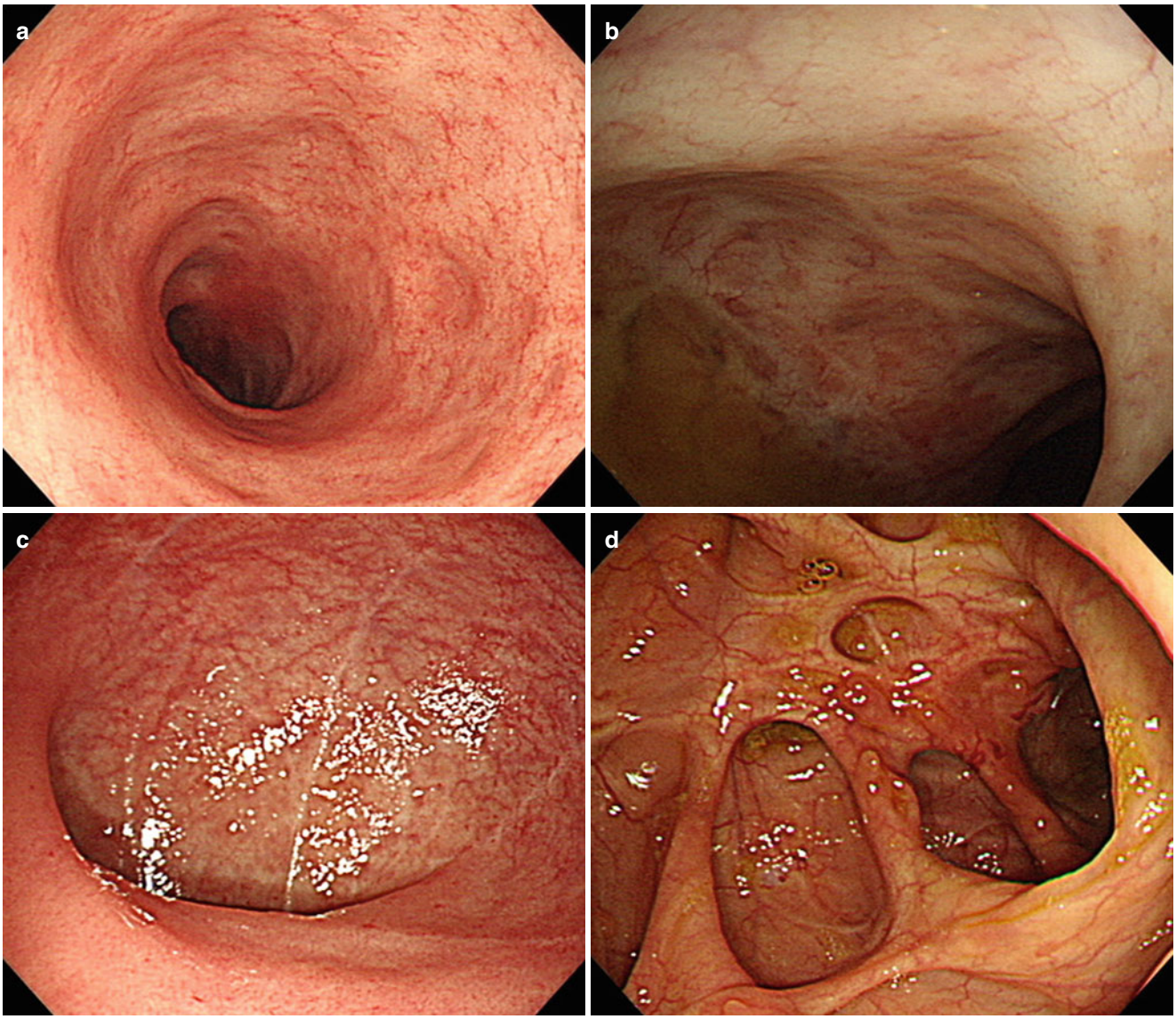


Fig. 19.16 Inactive stage of UC. (**a**) Regenerated vascularity shows sparse and irregular distribution. (**b**) White scar change after healing from active inflammation. (**c**) Linear white scars. (**d**) Extensive scars with pseudodiverticulums

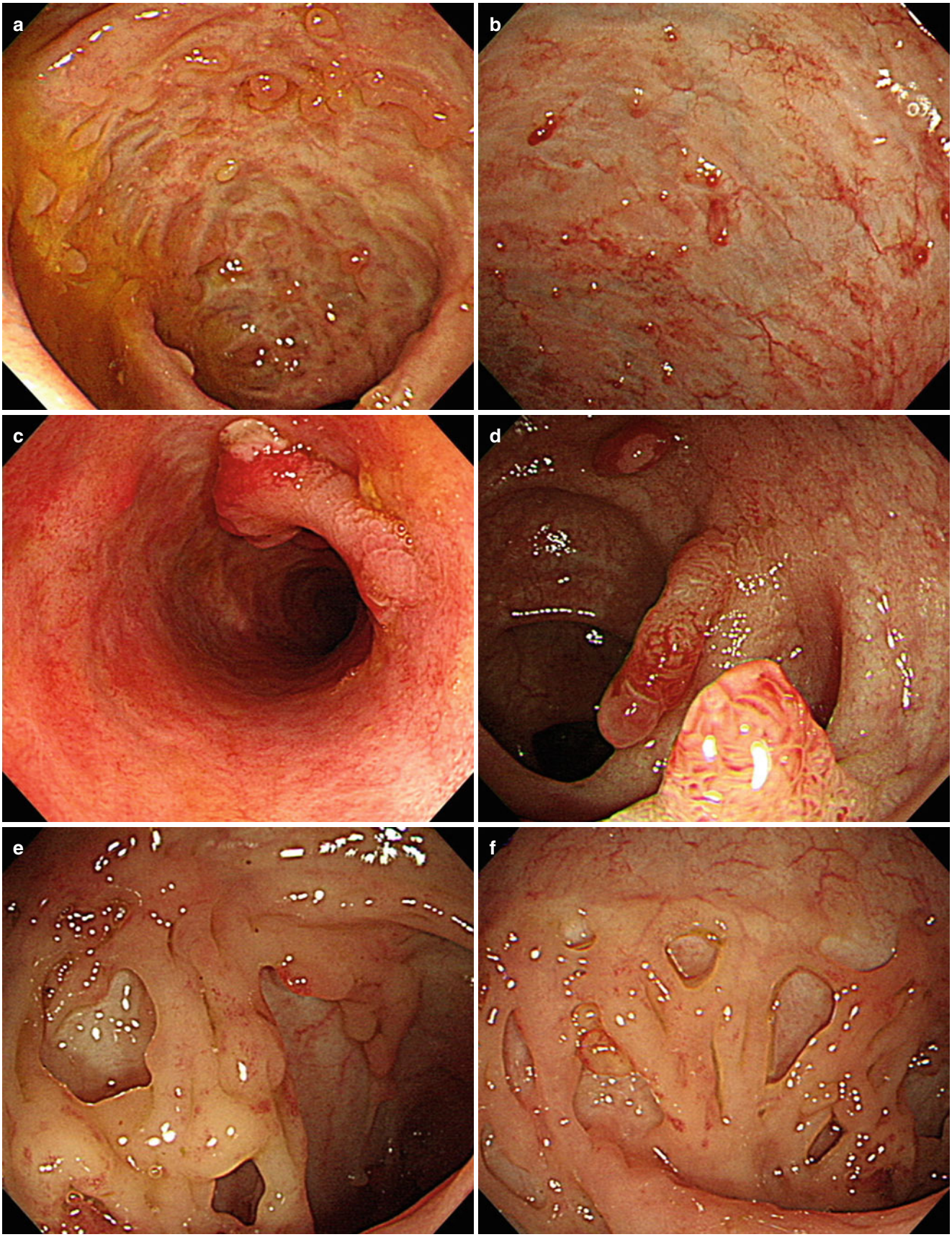


Fig. 19.17 Inflammatory polyps. (**a**) Multiple small pseudopolyps with white scars. Regenerated vascularity shows sparse and irregular distribution. (**b**) White scar change after healing from active inflammation. (**c**) Long pseudopolyp. (**d**) Pseudopolyps with various sizes and shapes. (**e**, **f**) Mucosal bridges

In active stage, inflammatory polyps can also be observed (Fig. 19.18).

After long-standing severe inflammation, the lumen of colon can become narrowed and the haustration can decrease. The change gives the colon its "lead-pipe appearance" (Fig. 19.19). The shortening of colon length can also develop.

After severe inflammation, stricture of lumen can develop. The luminal stricture in UC is usually shorter compared with luminal stricture in CD (Fig. 19.20). The important point is the differentiation between benign and malignant strictures. In patients with long history of UC, malignant strictures should be suspected in every case of colonic stricture, and biopsy of strictured segment should be performed. Because the yield of biopsy is not satisfactorily high, surgical treatment should be considered for stricture in patient with long history of UC.

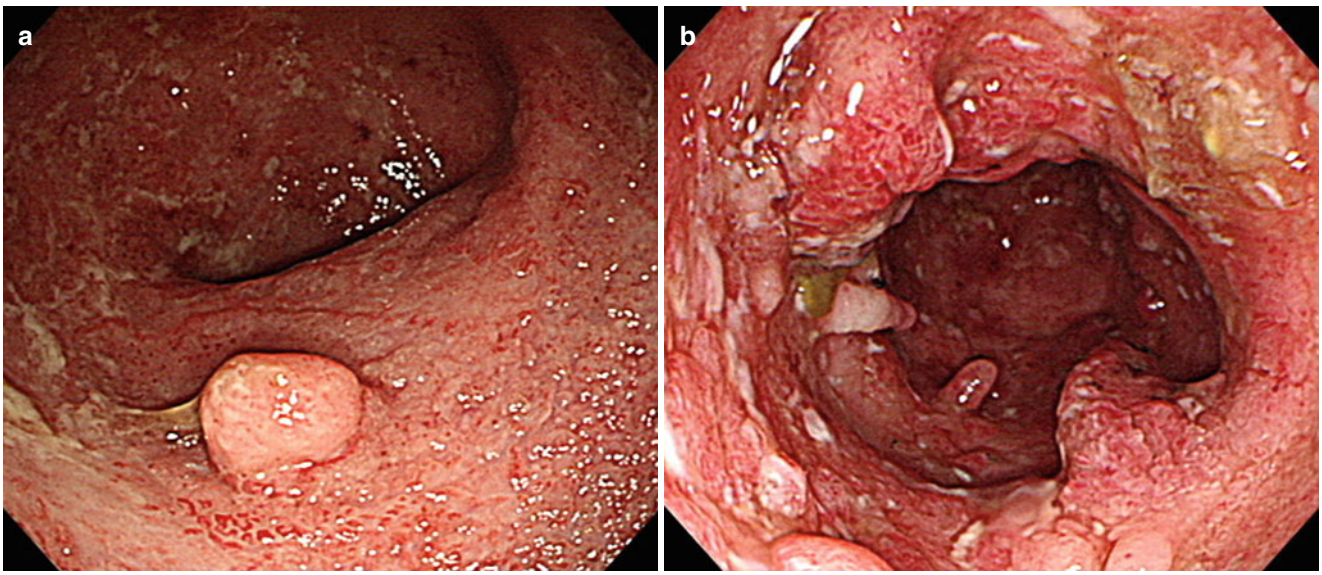


Fig. 19.18 Inflammatory polyps in active UC. (**a**) Solitary inflammatory polyp. (**b**) Multiple inflammatory polyps

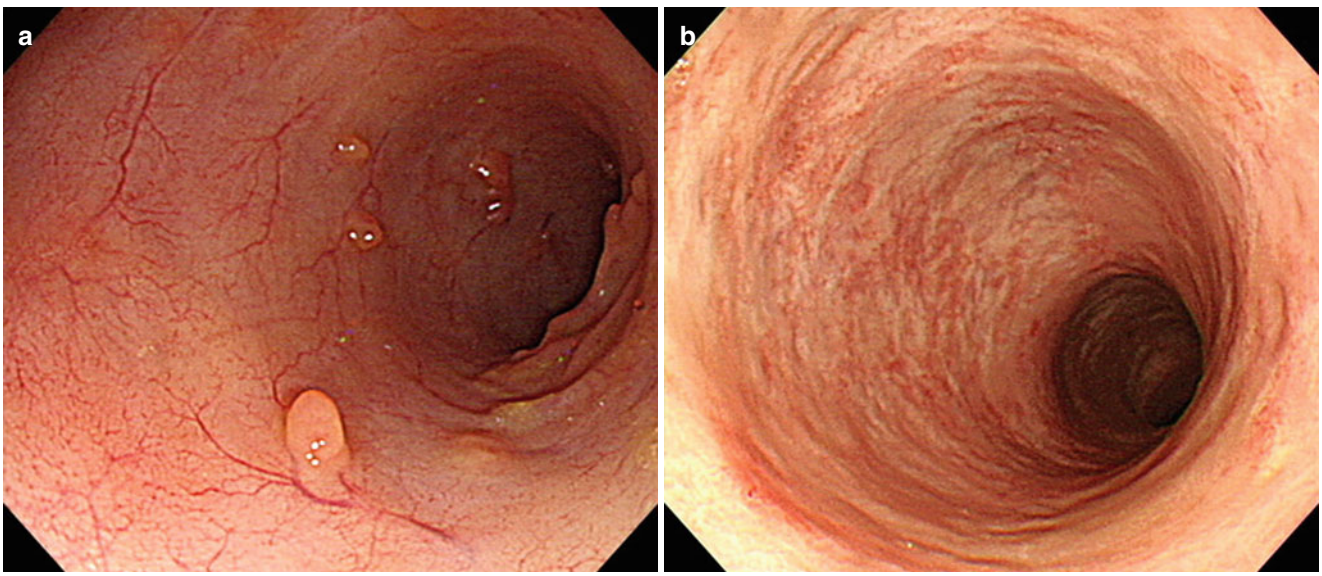


Fig. 19.19 Change of lumen after long-standing inflammation. (**a**) Decreased haustration with multiple pseudopolyps. (**b**) Loss of folds with extensive scar formation

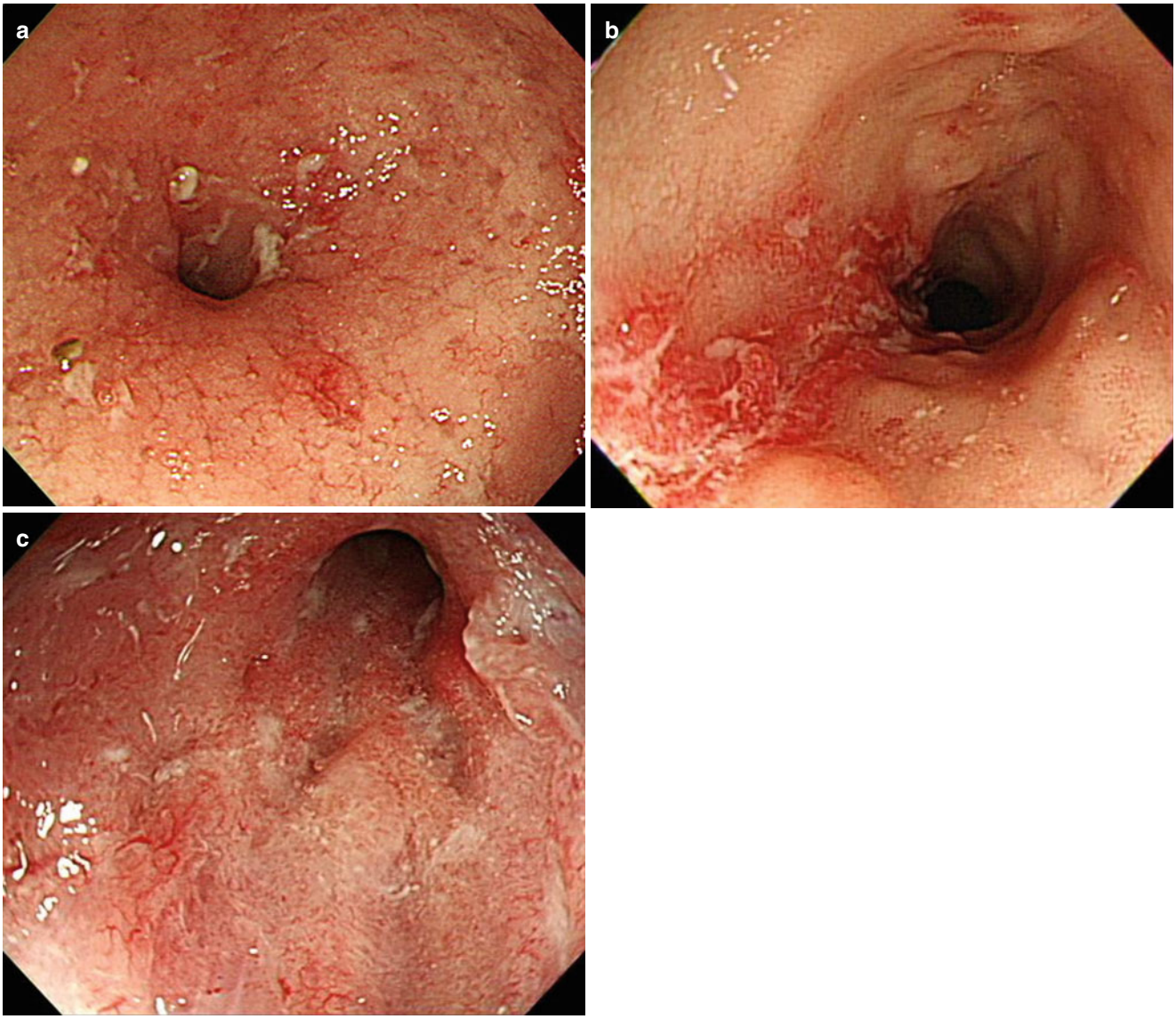


Fig. 19.20 Benign colonic strictures. (**a**) Colonoscope could not pass the stricture. (**b**) Active inflammation is noted around stricture. (**c**) Rectal stricture

UC Associated with Primary Sclerosing Cholangitis (Fig. 19.21)

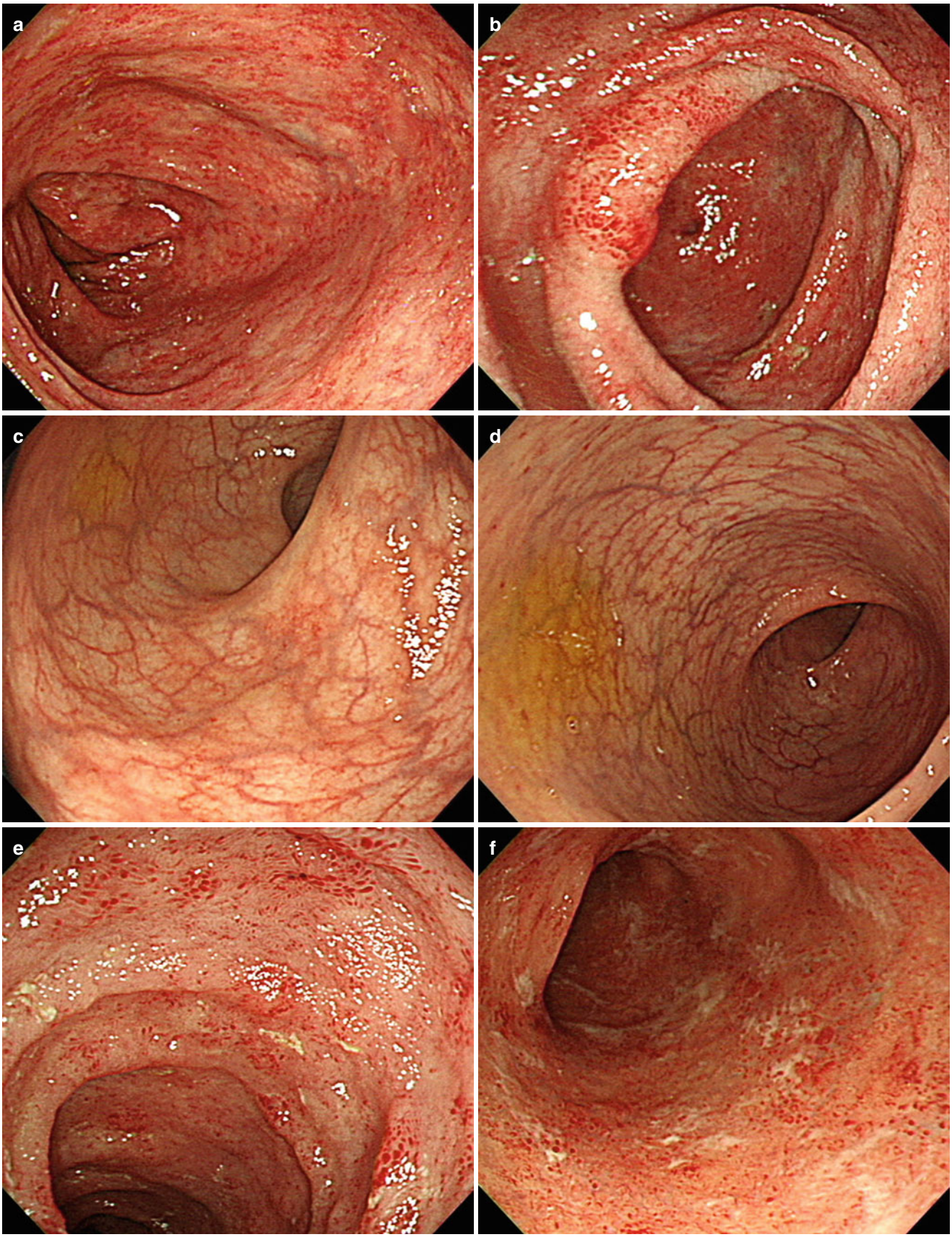


Fig. 19.21 Characteristic endoscopic features of UC associated with primary sclerosing cholangitis. (**a**, **b**) Mild inflammation. (**c**, **d**) Rectal sparing. (**e**, **f**) Backwash ileitis

Primary sclerosing cholangitis (PSC) is one of the extraintestinal manifestations of UC. UC associated with PSC shows more characteristic features compared with UC not associated with PSC. First, colonic inflammation is not usually severe. Second, rectal mucosa is relatively spared. Third, the frequency of pancolitis is high. Fourth, backwash ileitis is more common [3].

19.3.2.3 Dysplasia and Cancer

With long-standing inflammation in UC, the risk of dysplasia and cancer becomes high. When early dysplastic change develops in the background of inflammation, it is difficult to detect dysplastic change. When dysplastic changes can be grossly observed, they are called "dysplasia-associated lesion or masses" (DALM). It is not easy to differentiate between DALM and sporadic adenomatous polyps. When dysplastic change is observed from biopsy of mucosa around suspicious DALM, lesion can be diagnosed as DALM, not sporadic adenoma (Fig. 19.22).

Cancers associated with long-standing UC show various features. Usually, they show infiltrative features and the margin of cancer is not distinct. Other forms of cancer such as ulcerofungating mass, polypoid mass, or luminal stricture can be also observed (Fig. 19.23). The risk of dysplasia and cancer is considered to be high in patients with UC associated with PSC.

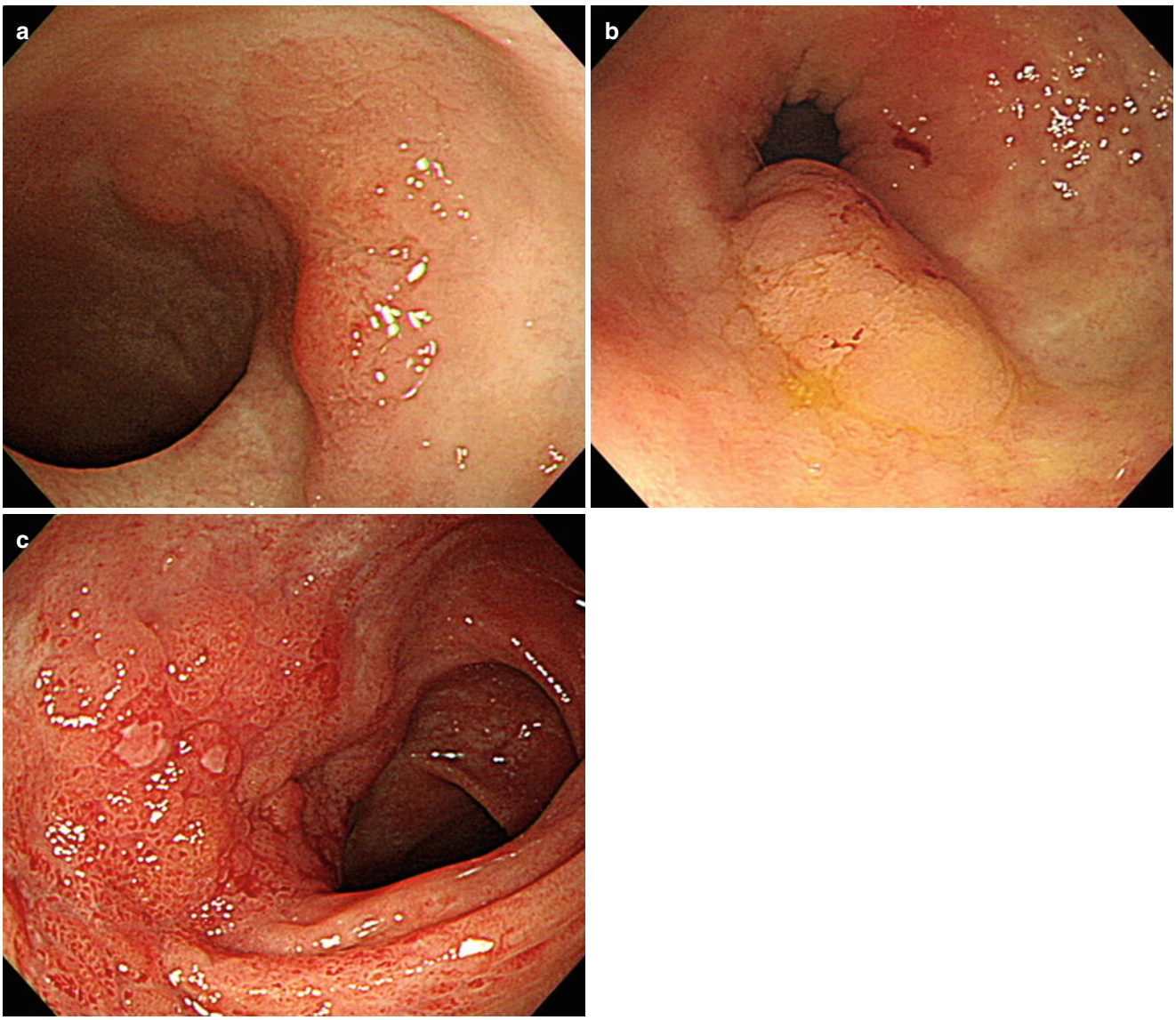


Fig. 19.22 Various features of DALM. (**a**) DALM in a female patient with a 13-year history of UC. (**b**) DALM in a female patient with an 18-year history of UC. (**c**) DALM in a patient with UC associated with primary sclerosing cholangitis

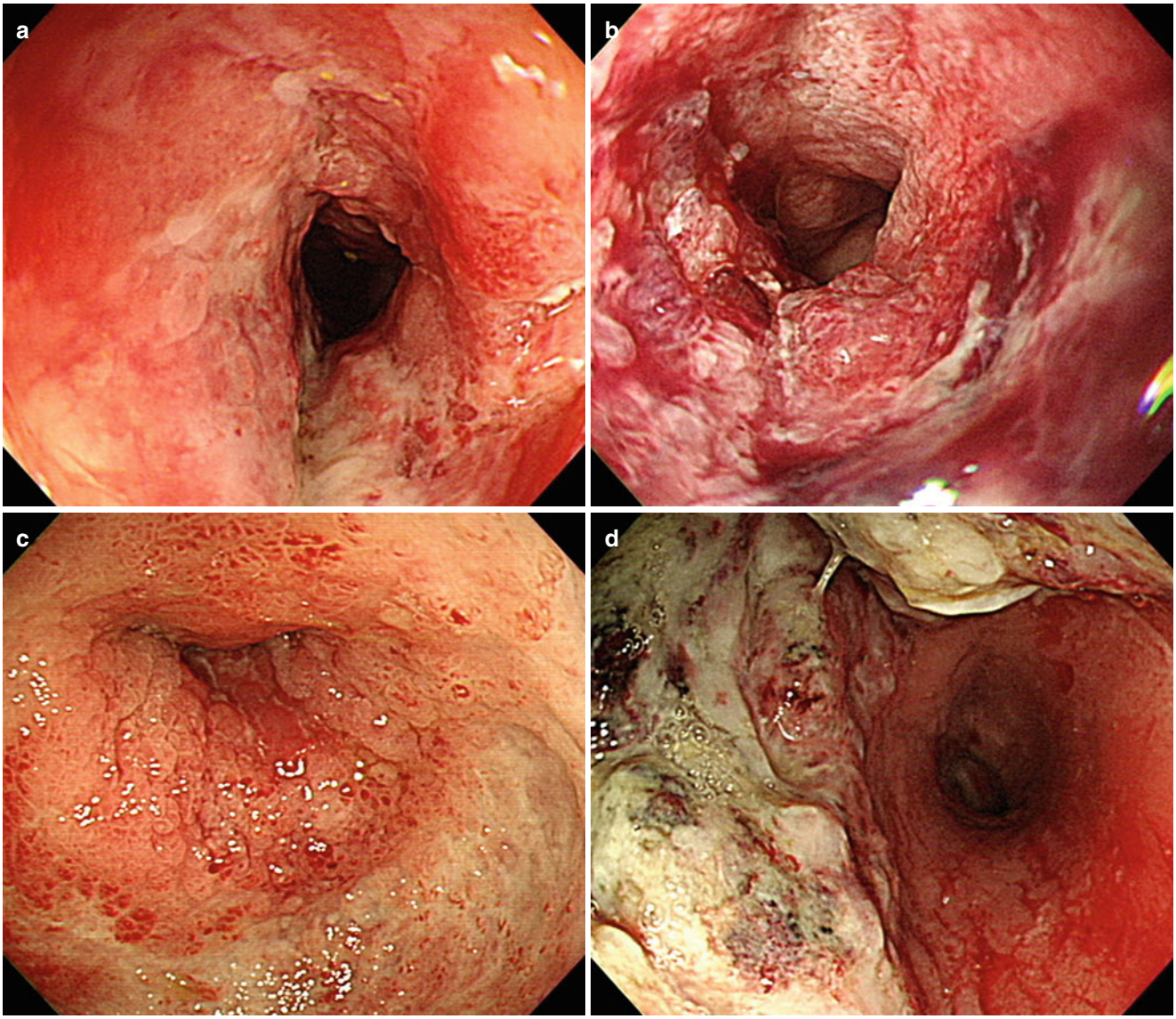


Fig. 19.23 Colitic cancers in patients with UC. (**a**–**c**) Diffuse infiltrative rectal adenocarcinoma with luminal stricture. (**d**) Ulcerofungating rectal adenocarcinoma. (**e**) Ulcerofungating adenocarcinoma of descending colon. (**f**, **g**) Polypoid adenocarcinoma of sigmoid colon

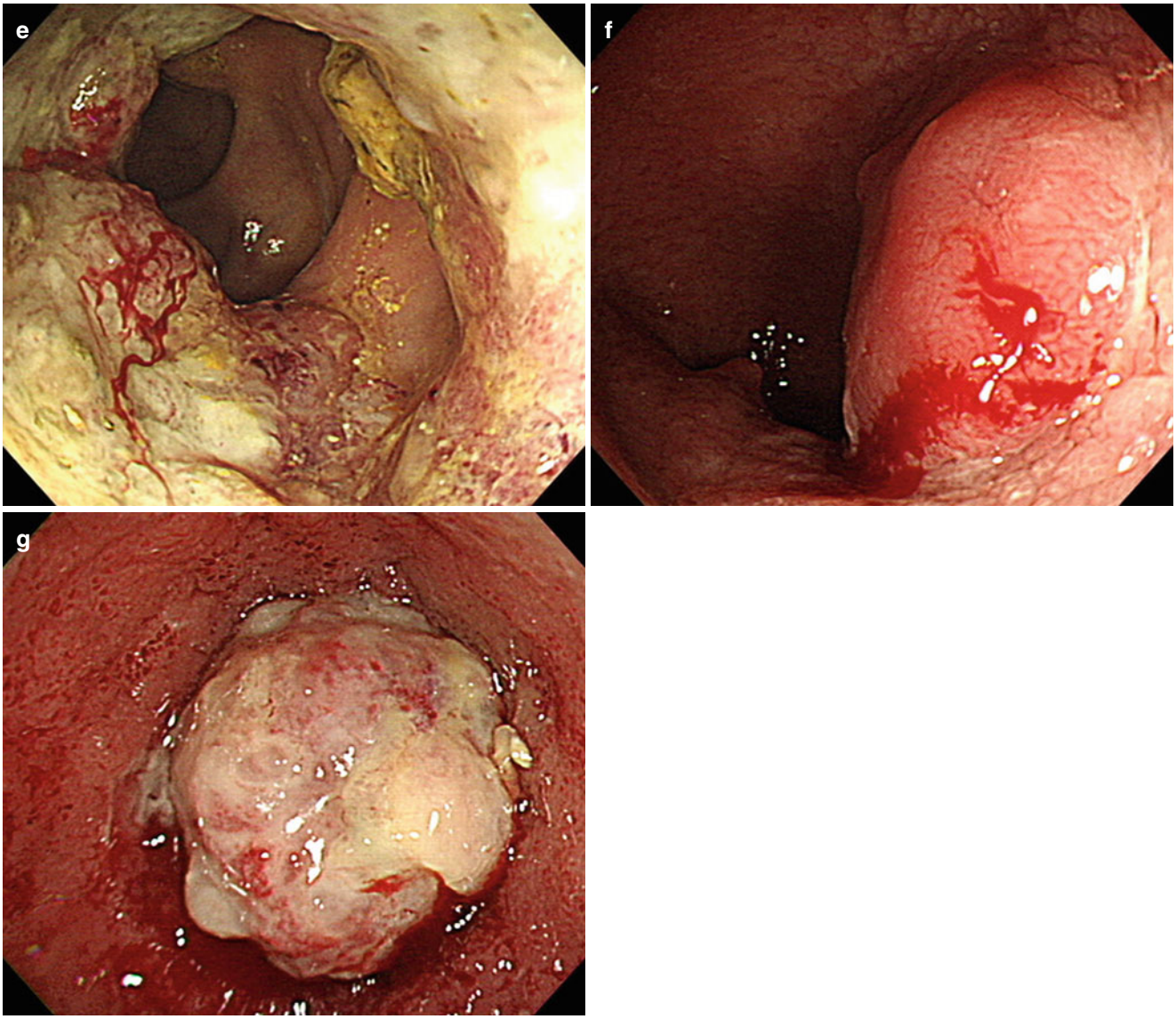


Fig. 19.23 (continued)

19.3.2.4 Pouchitis

After total proctocolectomy with ileal pouch-anal anastomosis for UC, inflammation of pouch can develop and it is called "pouchitis." The ileal mucosa shows hyperemia, edema, friability, and ulcers and it looks like mucosal change in active UC. The extent and degree of inflammation are various (Fig. 19.24). In patients with remnant rectal cuff, inflammation of cuff (cuffitis) can develop (Fig. 19.25).

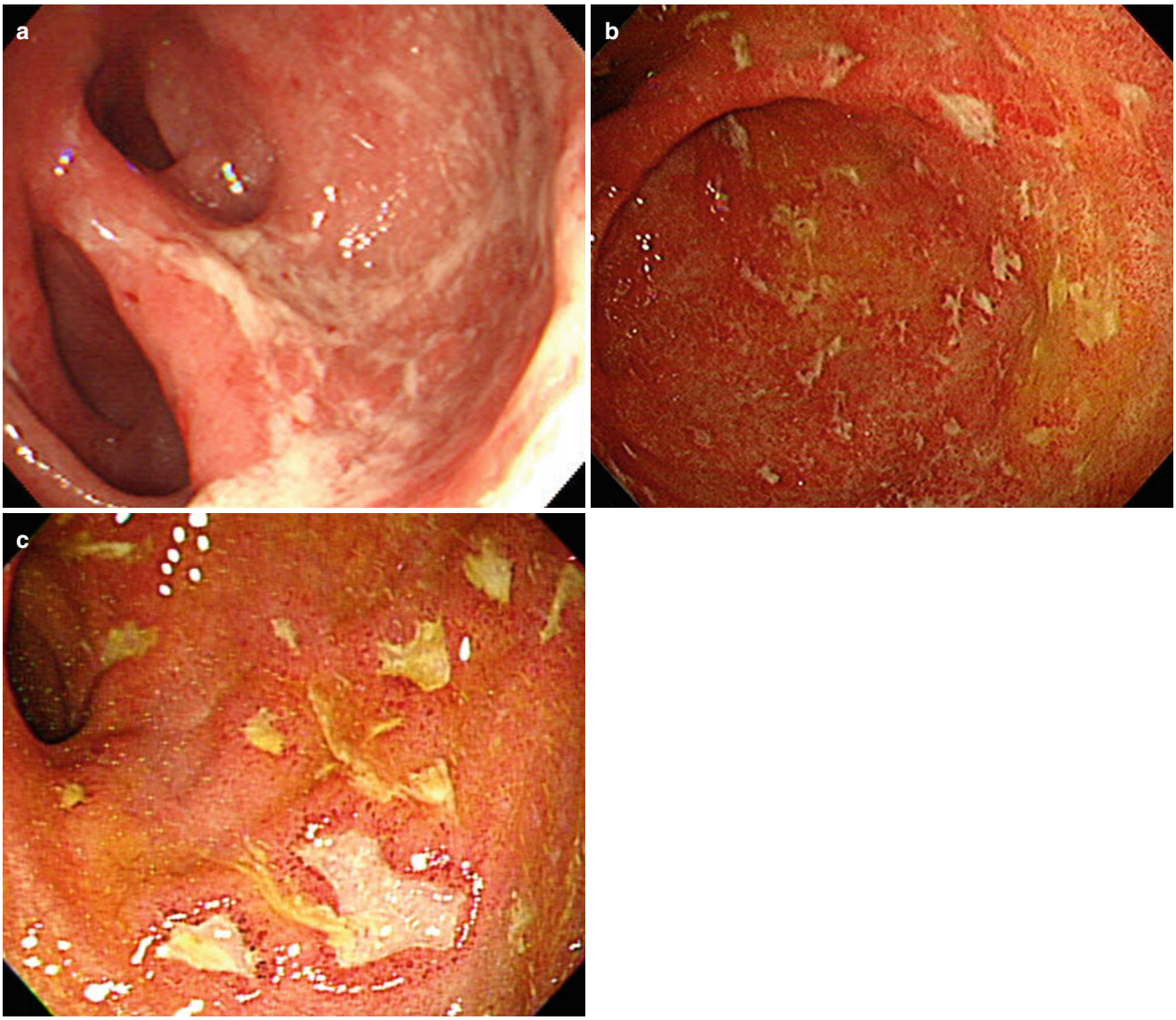


Fig. 19.24 Pouchitis. (**a**) Mild to moderate pouchitis with exudates. (**b**, **c**) Severe pouchitis with multiple ulcers

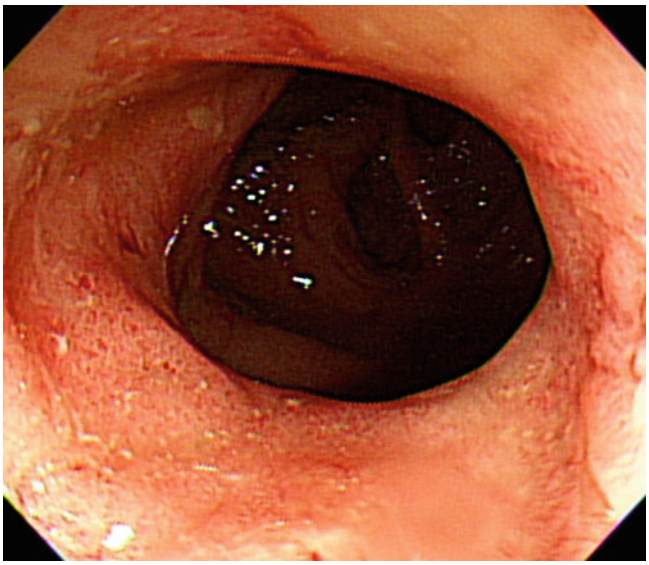


Fig. 19.25 Mild cuffitis

19.3.2.5 Gastroduodenal Lesions in Ulcerative Colitis

Traditionally, UC was regarded as the inflammation limited to colon except backwash ileitis. However, chronic gastroduodenal lesions have been reported in patients with UC, recently (Fig. 19.26). The clinical significance of gastroduodenal lesions is unclear yet, and the consensus criteria for diagnosing gastroduodenal lesions associated with UC were not established.

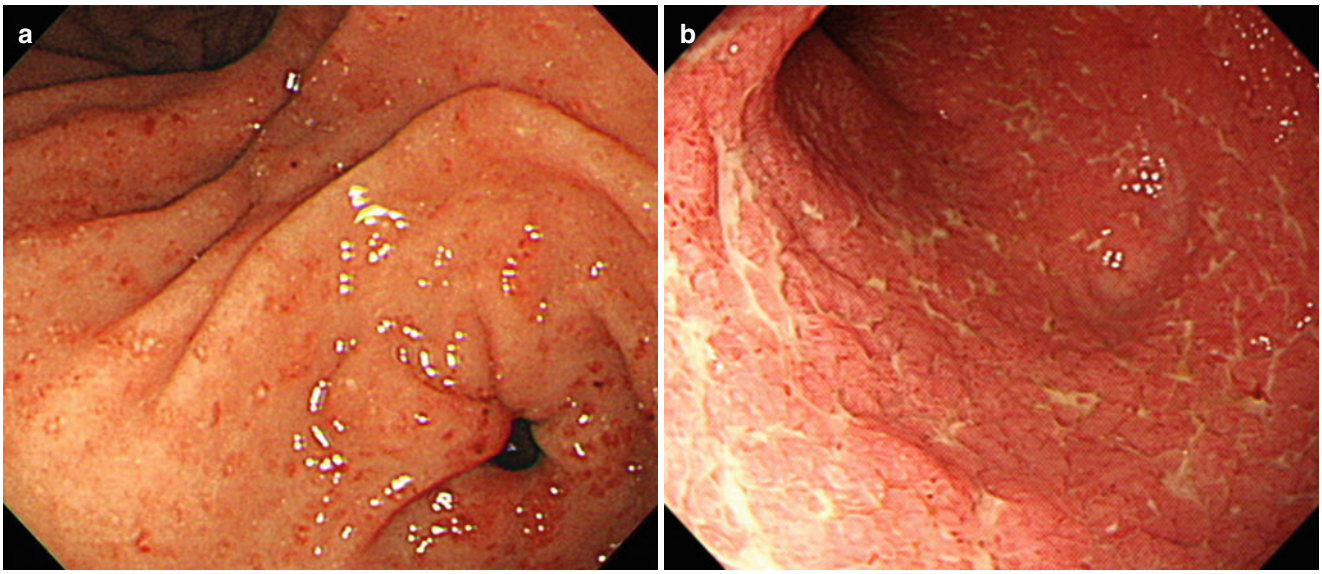


Fig. 19.26 Gastroduodenal inflammation in patients with UC. (**a**) Multiple aphthous lesions and erythematous spots are noted at gastric antrum. (**b**) Diffuse inflammation and superficial ulcers mimicking colonic inflammation are noted at duodenal bulb

19.4 Crohn's Disease

19.4.1 Clinical Manifestations

Crohn's disease (CD) is a chronic inflammatory disease of gastrointestinal tract which can involve whole gut from mouth to anus. Similar with UC, the etiopathogenesis of CD is still unclear and there are no curative medical treatments for CD. Its usual clinical manifestation is chronic abdominal pain, weight loss, and diarrhea developing in young adolescents. Interestingly, perianal abscess and fistula can develop before, at, or after the diagnosis of CD. The disease activity of CD tends to change over time, but bowel damage accumulates. In contrast to UC, transmural inflammation is characteristic of CD, and complications such as stricture or penetration of bowel walls commonly develop with long duration of illness. Penetration encompasses bowel perforation, inter-bowel fistula, perianal fistula, and fistula between bowel and adjacent organs such as bladder or vagina. Therefore, patients frequently need repeated surgery during the course of illness. Like UC, induction of clinical remission with medical treatment is needed for active CD, and long-term maintenance therapy is necessary for maintaining medically induced remission. Drugs such as 5-aminiosalicylic acids, antibiotics, corticosteroids, thiopurines, methotrexate, and anti-TNF-alpha agents are used for treating CD. In addition to medications, optimized surgical treatments are needed depending on the status of patients. Recently, various biologic agents based on the pathophysiologic mechanism of inflammation are being tried for CD.

19.4.2 Endoscopic Features

In contrast to UC, lesions in CD are distributed in skipped pattern with normal intervening mucosa [1]. Usually, the lesions are eccentric rather than concentric. The most commonly involved area is ileocecal areas, but inflammation can be observed throughout the colon.

The early lesions of CD are tiny punctuate hyperemia with edema (Fig. 19.27). A number of lesions are various. They are considered to be the preceding lesions of aphthous erosions/ulcerations.

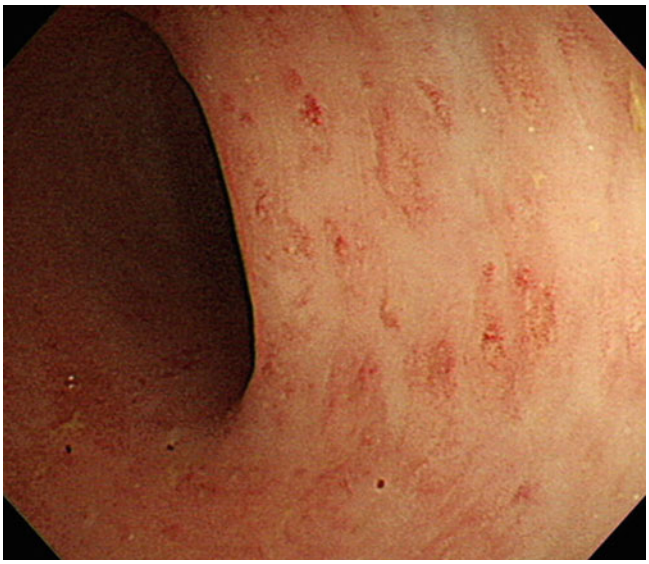


Fig. 19.27 Multiple hyperemic lesions of CD at rectum. They are regarded as early lesions of CD

Aphthous erosions/ulcerations are flat or slightly depressed shallow erosions/ulcerations smaller than 5 mm (Fig. 19.28). Lesions are usually surrounded by hyperemic rims. The surrounding mucosa of aphthous erosions/ulcerations shows normal appearance or blurred vasculatures. Although aphthous erosions/ulcerations, especially longitudinally arranged aphthous erosions/ulcerations, are typical early lesions of CD (Fig. 19.29), they can be observed in other inflammatory diseases.

With disease progression, larger ulcerations with various features can develop (Fig. 19.30). Typically, longitudinal ulcerations show serpiginous features or tram track-like appearance (Fig. 19.31).

In addition to longitudinal ulcers, geographic ulcers and transverse ulcers can also be observed (Fig. 19.32).

If deep longitudinal ulcers and transverse ulcers are connected with each other, the intervening protruded mucosa appears like a cobblestone. Although this so-called cobblestone appearance is not pathognomonic of CD, it is useful for endoscopic diagnosis of CD (Fig. 19.33).

After healing of chronic deep ulcers and fissures, stricture of bowel lumen can develop. The stricture of CD is usually longer than that of UC and is commonly accompanied by ulcerations and cobblestone appearance (Fig. 19.34).

Fistula formation is another feature of CD and represents penetrating complication of CD. Sometimes, the opening of fistula can be evidently seen. However, in many cases, the opening is not evident and the fistula could be suspected by observing mucosal edema, hyperemia, or pseudopolyps around small opening of fistula (Fig. 19.35).

After healing of inflammation, scars and inflammatory pseudopolyps can be observed like UC (Fig. 19.36).

Terminal ileum is commonly involved in CD and it is important to intubate terminal ileum and to observe for differential diagnosis between CD and UC if it is difficult to differentiate between two diseases with colonic features. In terminal ileum, all kinds of various mucosal changes developing in colon can also be observed. After bowel resection and ileocolic anastomosis, endoscopic recurrence in the neoterminal ileum can develop in the early postoperative period, and it usually precedes clinical recurrence. The early endoscopic recurrence is small aphthous ulcers, and with severe recurrence, the number and size of ulcers increase. In cases of advanced endoscopic recurrence, huge ulcers with luminal stenosis can be observed (Fig. 19.37).

In patients with long-standing inflammation, inflammation-associated cancer can develop (Fig. 19.38).

Because CD can involve any site of gastrointestinal tract, upper GI tract can also be involved. Any features observed in ileocolon can be detected during upper GI endoscopy (Fig. 19.39).

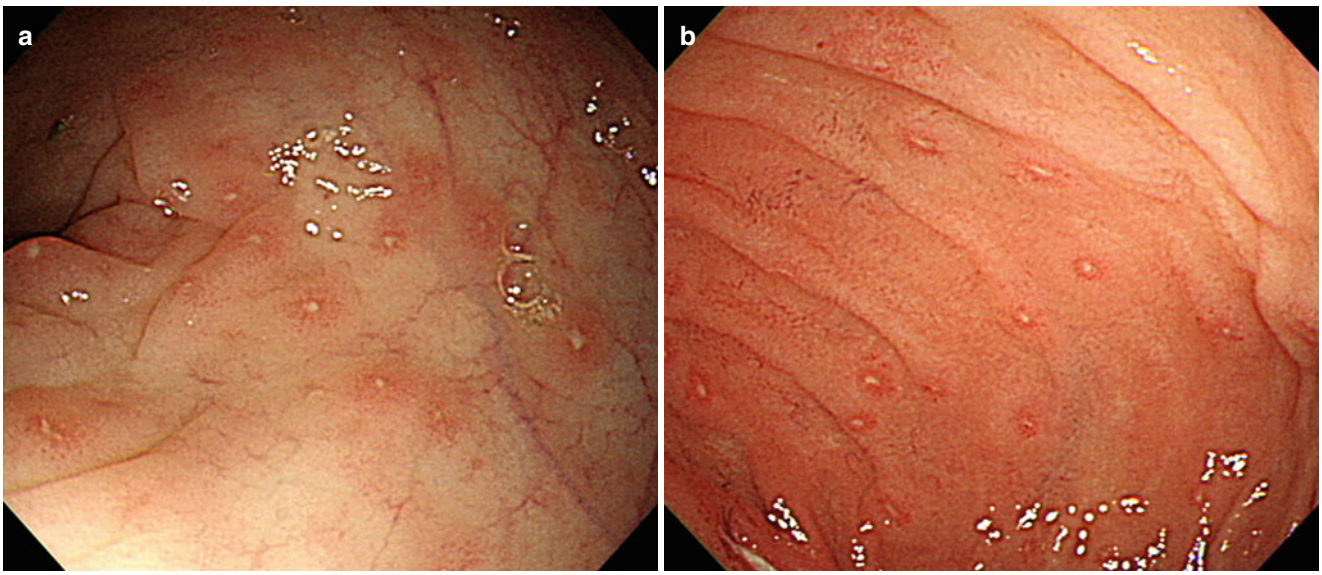


Fig. 19.28 Aphthous erosions. (**a**) Rectum (**b**) Terminal ileum

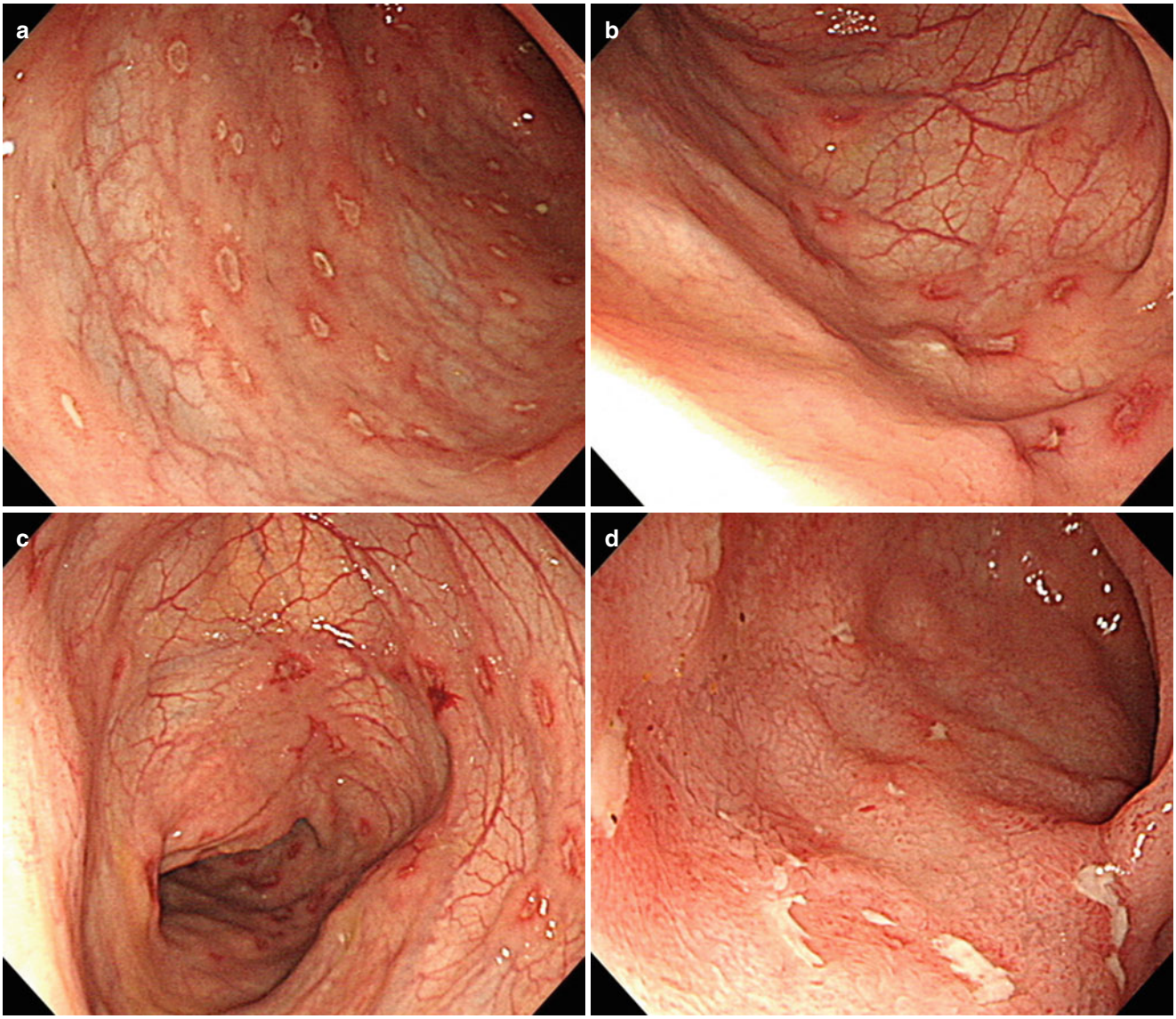


Fig. 19.29 Aphthous ulcerations. (**a**) Multiple aphthous ulcerations in colon. (**b**, **c**) Aphthous ulcerations of colon arranged in longitudinal fashion. (**d**) Longitudinally arranged aphthous ulcerations in terminal ileum

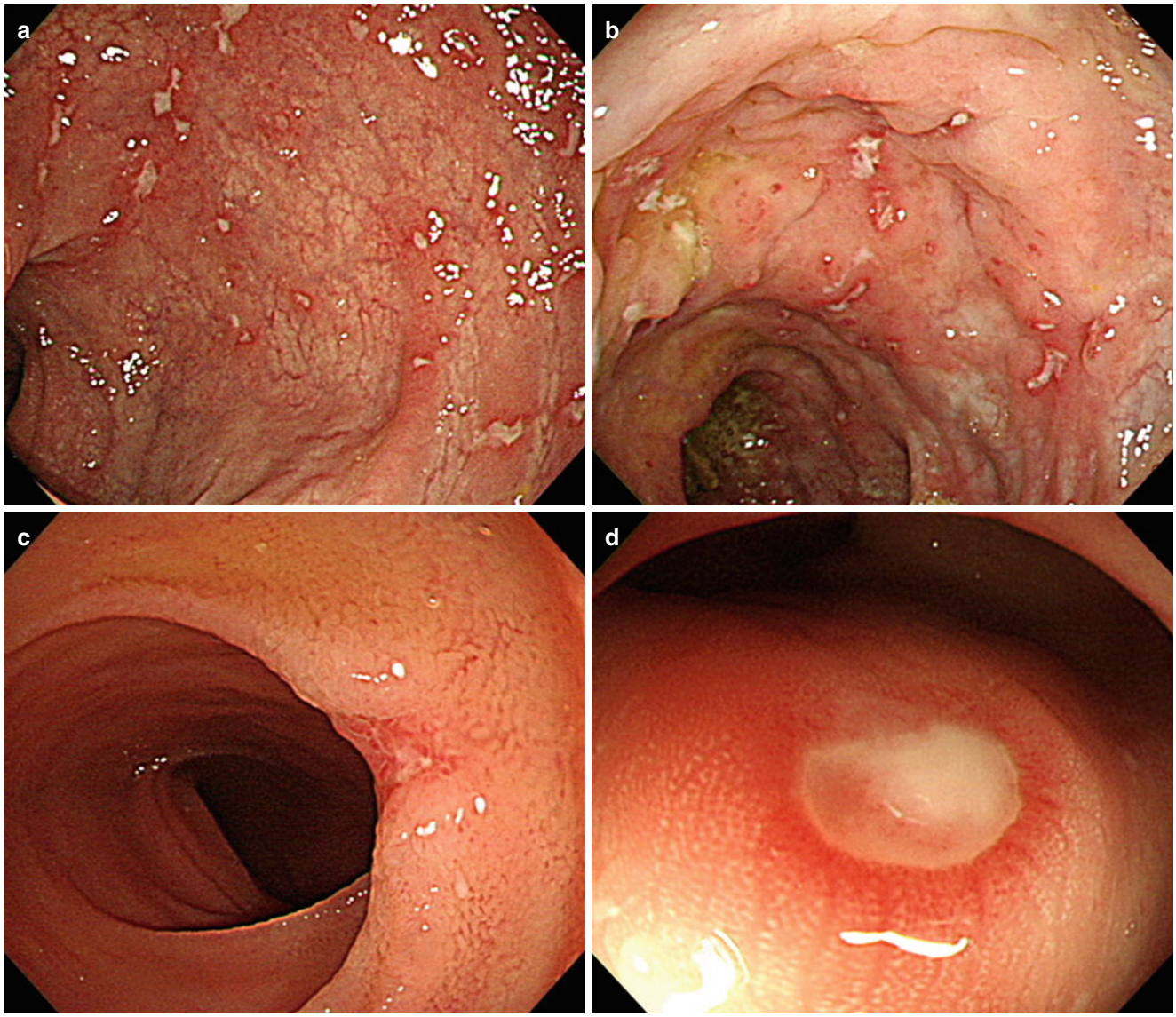


Fig. 19.30 Various shapes of ulcers. (**a**, **b**) Superficial ulcers are arranged in longitudinal fashion. (**c**) Triangular-shaped ulcer in terminal ileum. (**d**) Ovoid colonic ulcer

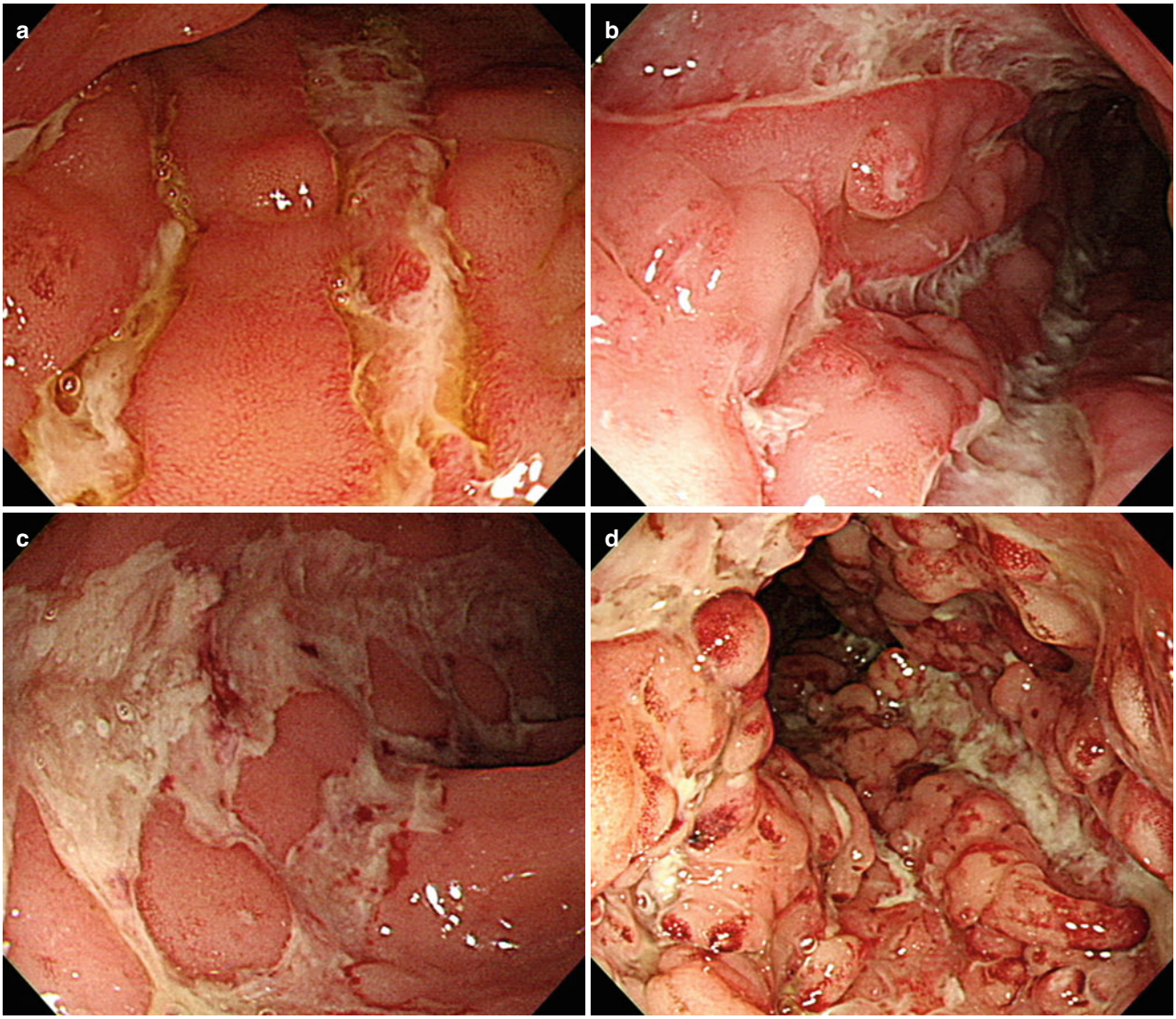
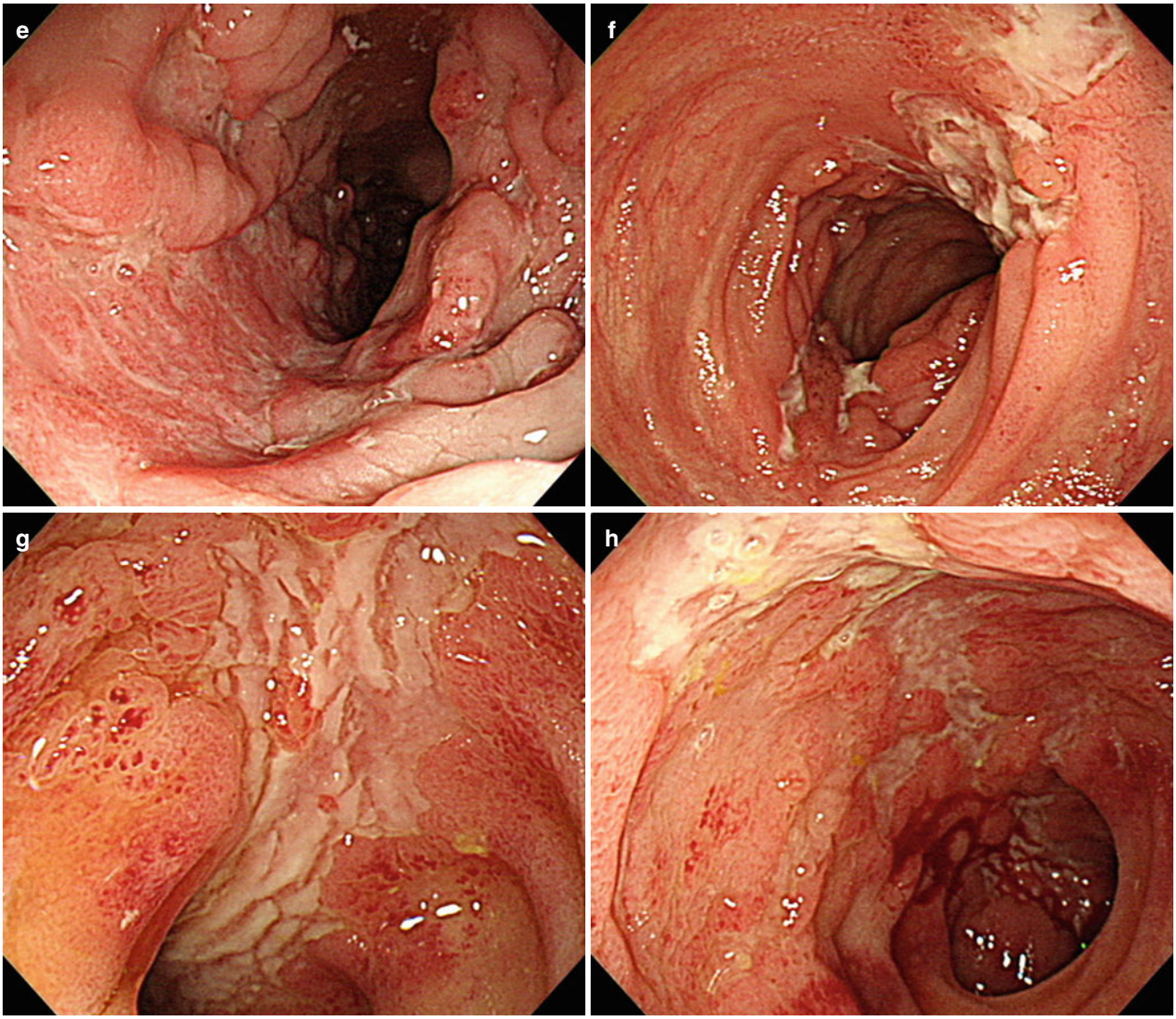


Fig. 19.31 Longitudinal ulcers. (**a**, **b**) Longitudinal ulcers of colon showing tram track-like appearance. (**c**) Longitudinal ulcers of colon with discrete margin. (**d**, **e**) Deep longitudinal ulcer. Mucosa around ulcers shows nodular appearance with hyperemia. (**f**–**h**) Longitudinal ulcers of terminal ileum



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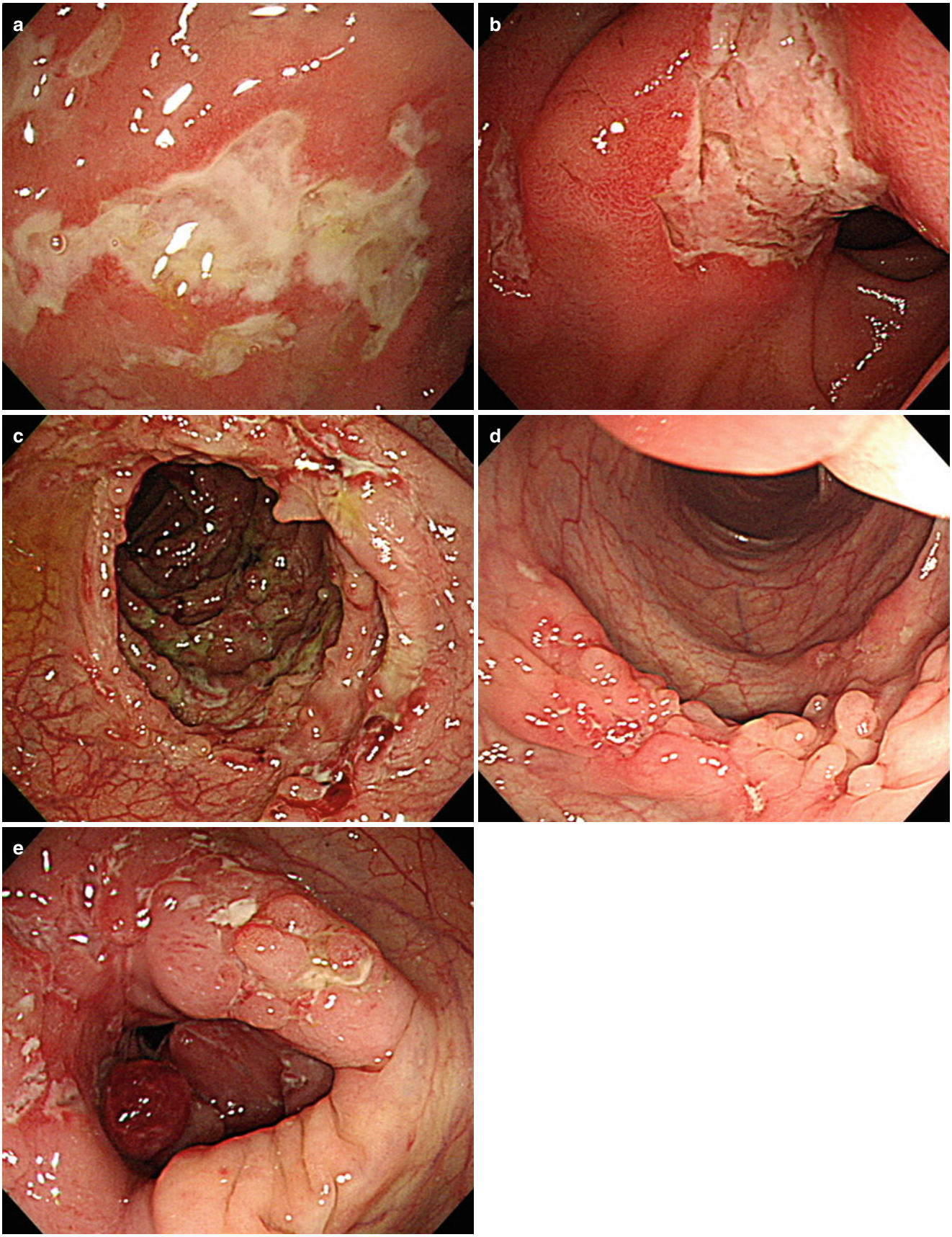


Fig. 19.32 Geographic ulcers and transverse ulcers. (**a**) Geographic ulcers of colon with discrete margin and clean base. (**b**) Geographic ulcer at the ileosigmoid anastomosis site. (**c**, **d**) Transverse ulcers encircling colonic lumen. Longitudinally directed ulcers are also noted. (**e**) Transversely arranged ulcers

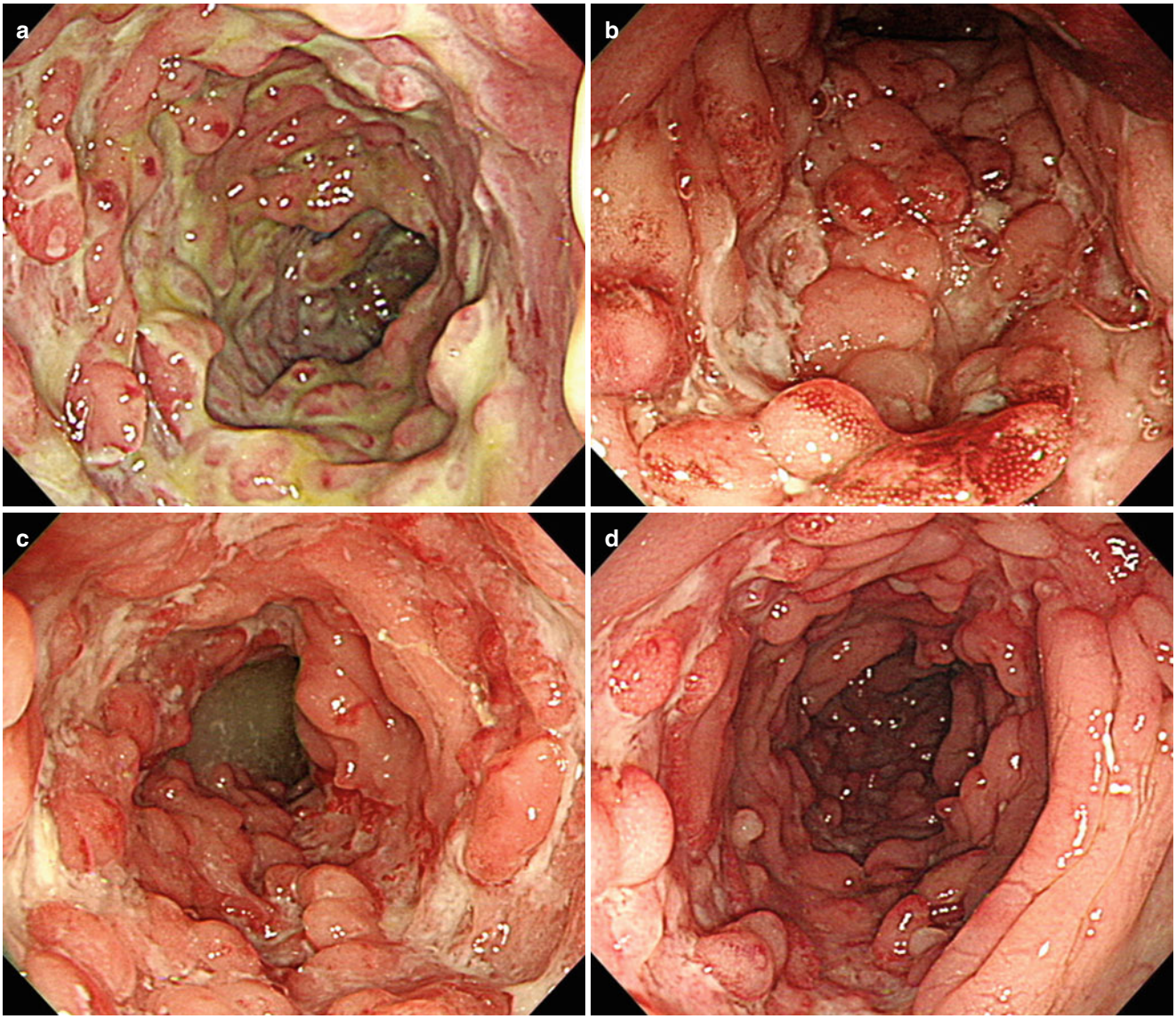


Fig. 19.33 Cobblestone appearance. (**a**, **b**) Longitudinal and transverse ulcers make cobblestone-like mucosa. (**c**, **d**) Cobblestone appearance showing normal or only hyperemic nodular intervening mucosa

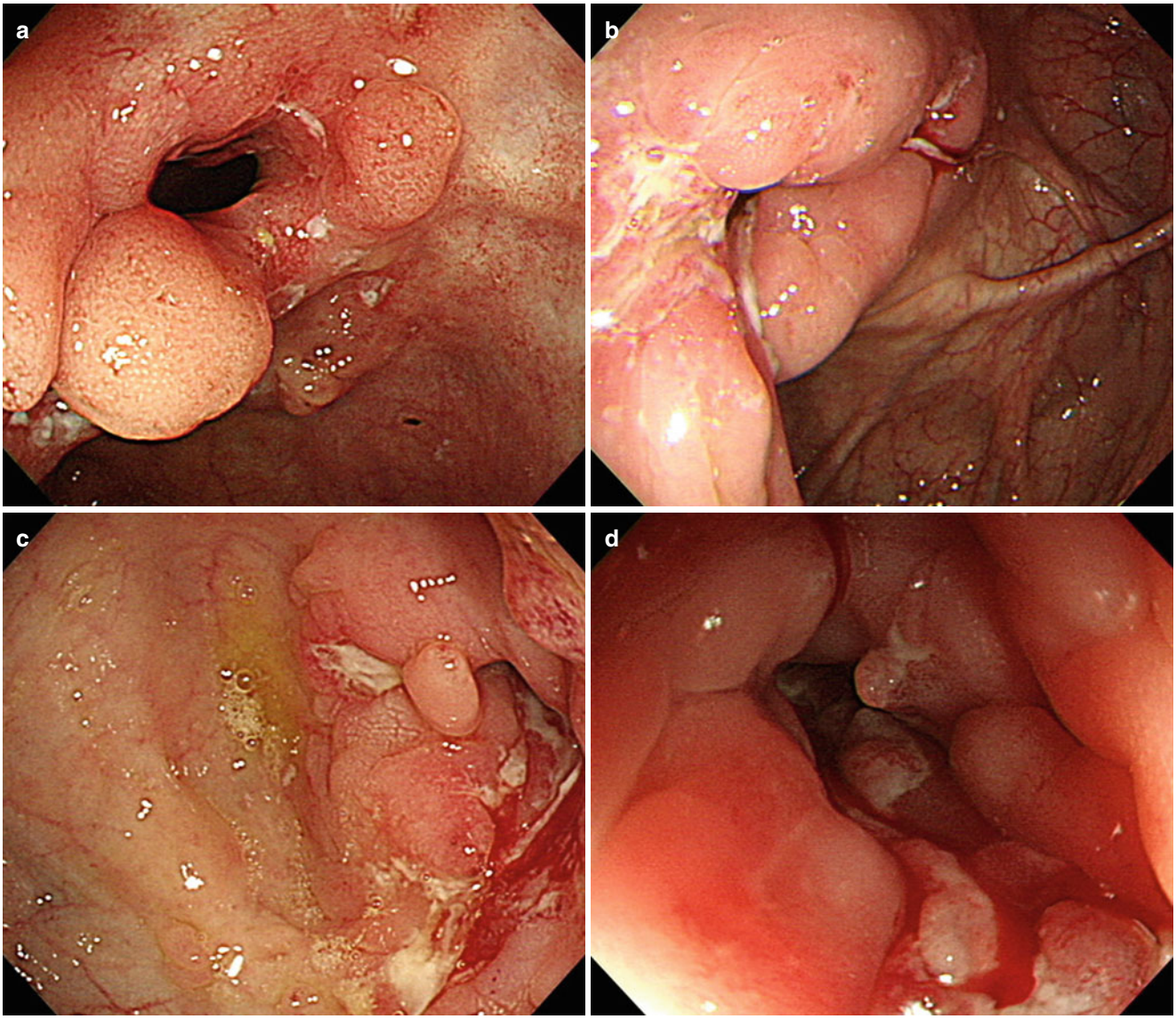
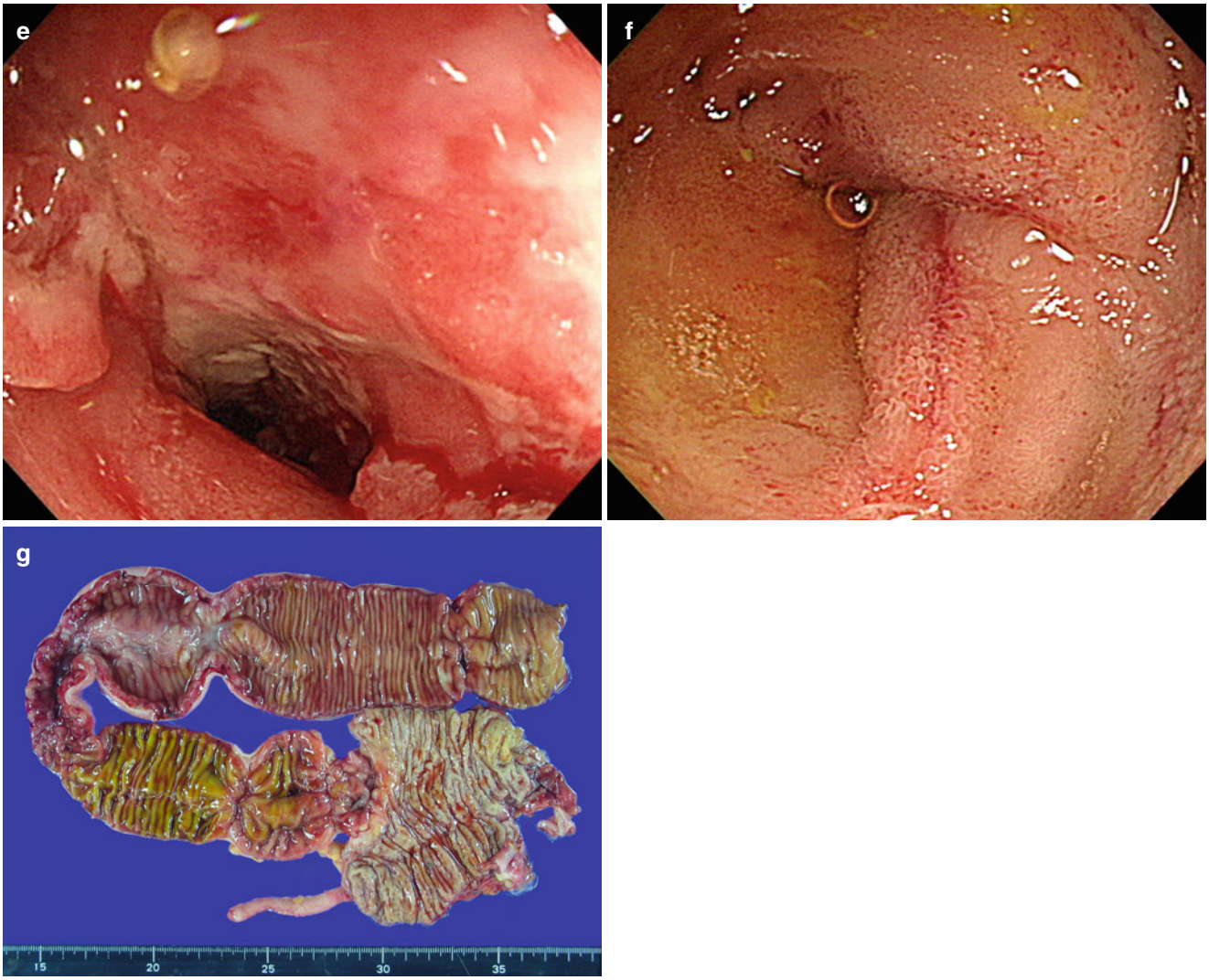


Fig. 19.34 Stricture. (**a**, **b**) Stricture of ileocecal valve. (**c**–**e**) Stricture of colon. Ulcers and/or pseudopolyps are observed in the orifice of stricture. (**f**, **g**) Stricture of terminal ileum and resected specimen showing multifocal strictures of terminal ileum



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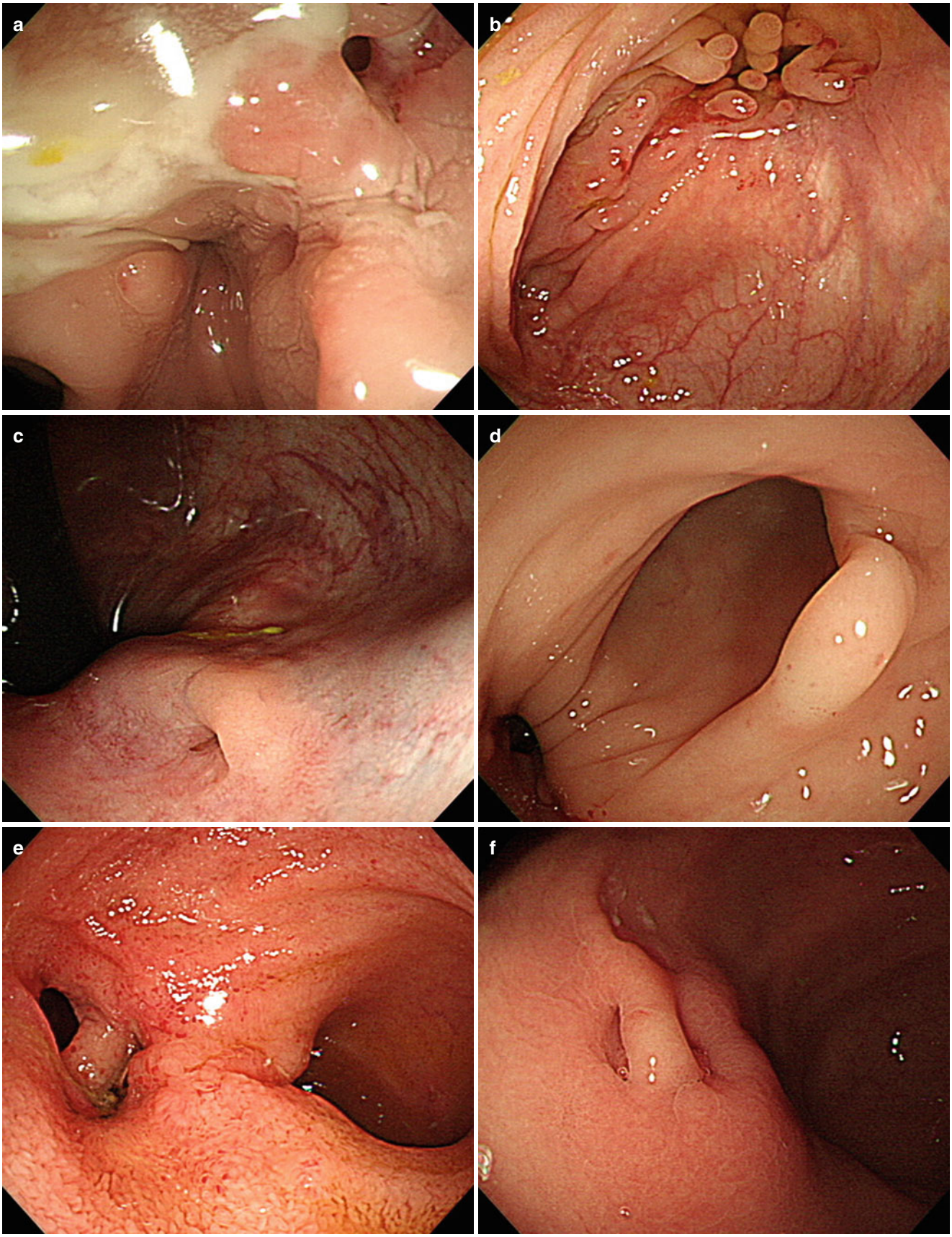

Fig. 19.35 Fistula of CD. (**a**) In the *upper right side* of the figure, fistula opening with converging folds is observed. (**b**) Opening of fistula is not evident, but multiple pseudopolyps with converging folds suggest the opening of fistula. (**c**) On the retroflexion of colonoscope, internal opening of perianal fistula is suspected. (**d**) On the *left lower side* of the figure, fistula opening is observed. Deformity of colonic lumen is accompanied. (**e**) Fistula opening in the terminal ileum (*left side* of the figure). (**f**) In stomach, opening of gastroduodenal fistula is observed

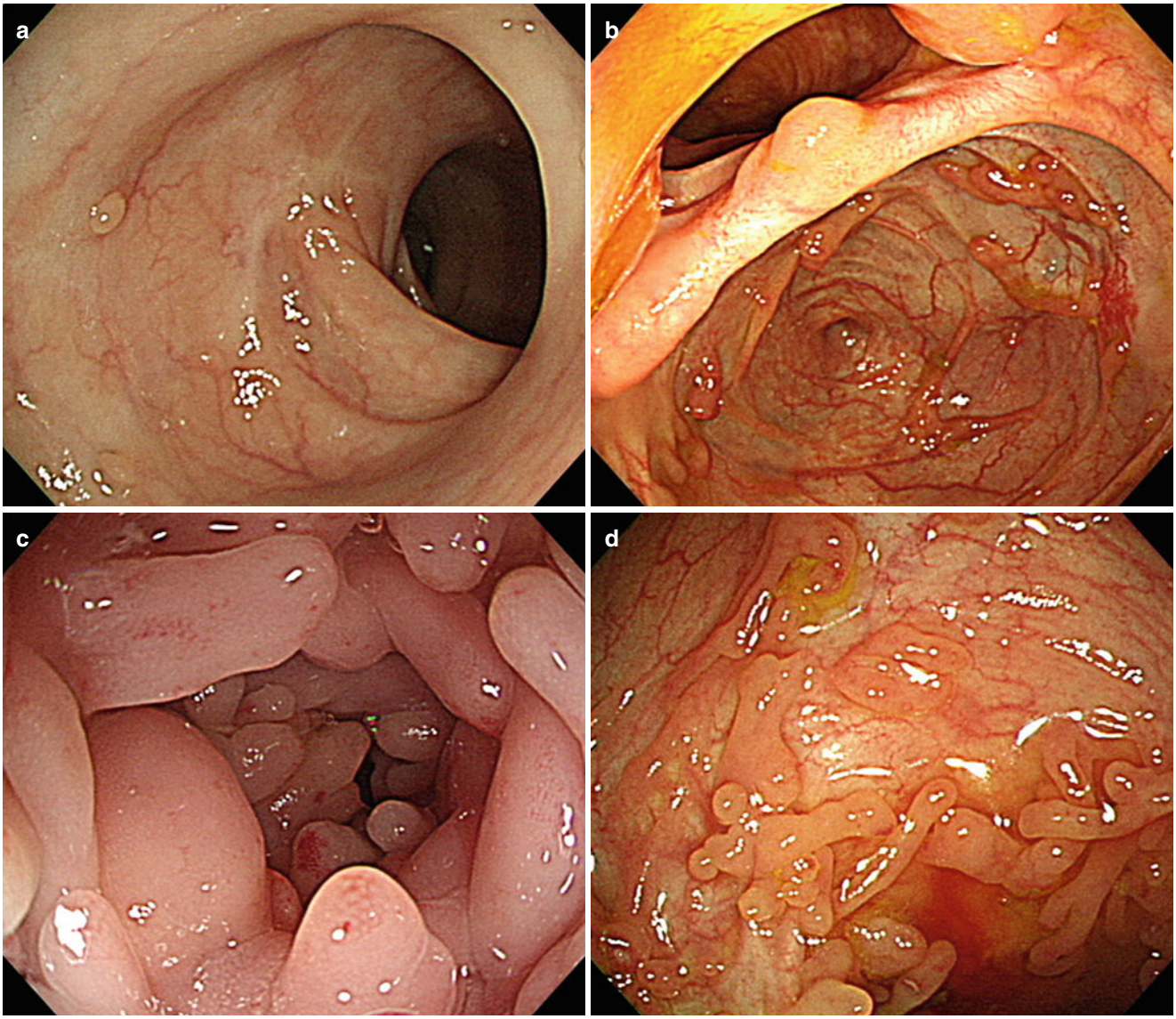

Fig. 19.36 Scars and pseudopolyps. (**a**) White scars with pseudopolyps in colon. (**b**) Patulous ileocecal valve due to scar formation and multiple pseudopolyps. (**c**) With multiple pseudopolyps, colonic lumen appears to be narrowed. (**d**) After healing of inflammation, multiple pseudopolyps with scar are observed. (**e**, **f**) After healing of undermining ulcers, mucosal bridges were formed. (**g**) Active geographic ulcer is observed in the ascending colon. (**h**) After treatment with infliximab, only scar with pseudopolyp is observed

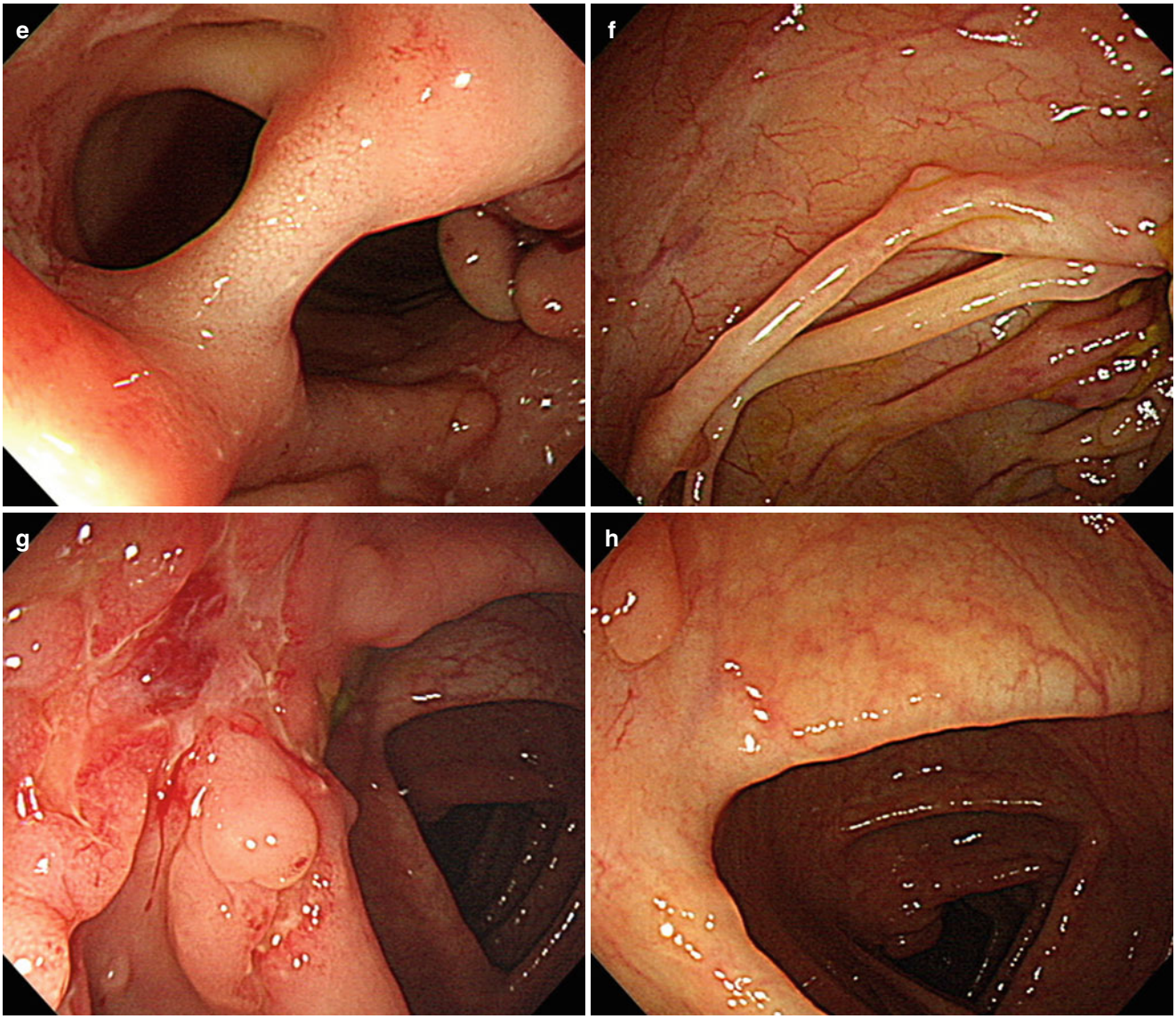

Fig. 19.36 (continued)

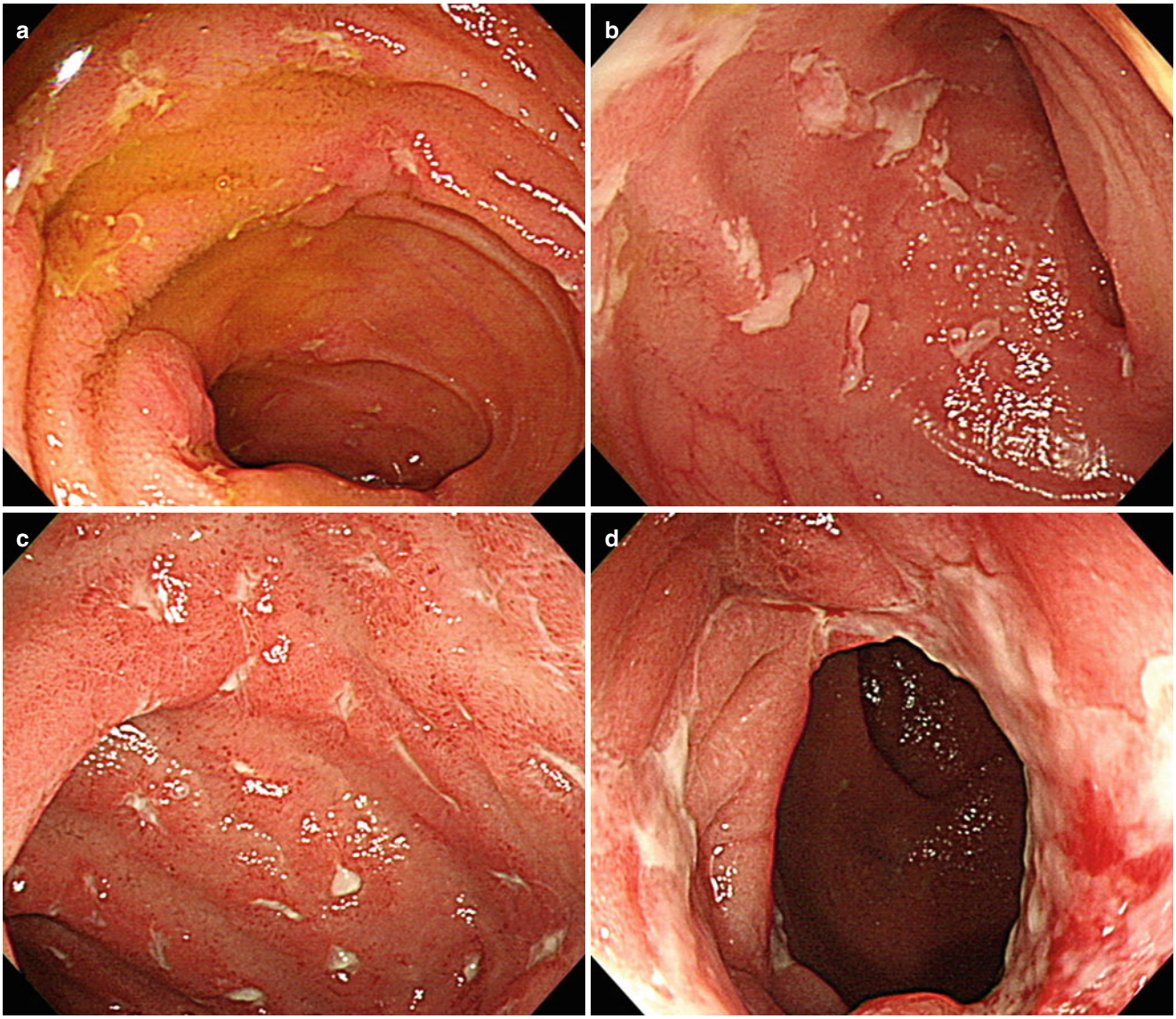

Fig. 19.37 Postoperative endoscopic recurrence. (**a**, **b**) Several small ulcers and geographic ulcers are observed in neoterminal ileum. (**c**) Multiple ulcers of neoterminal ileum. (**d**) Large ulceration with luminal narrowing in neoterminal ileum

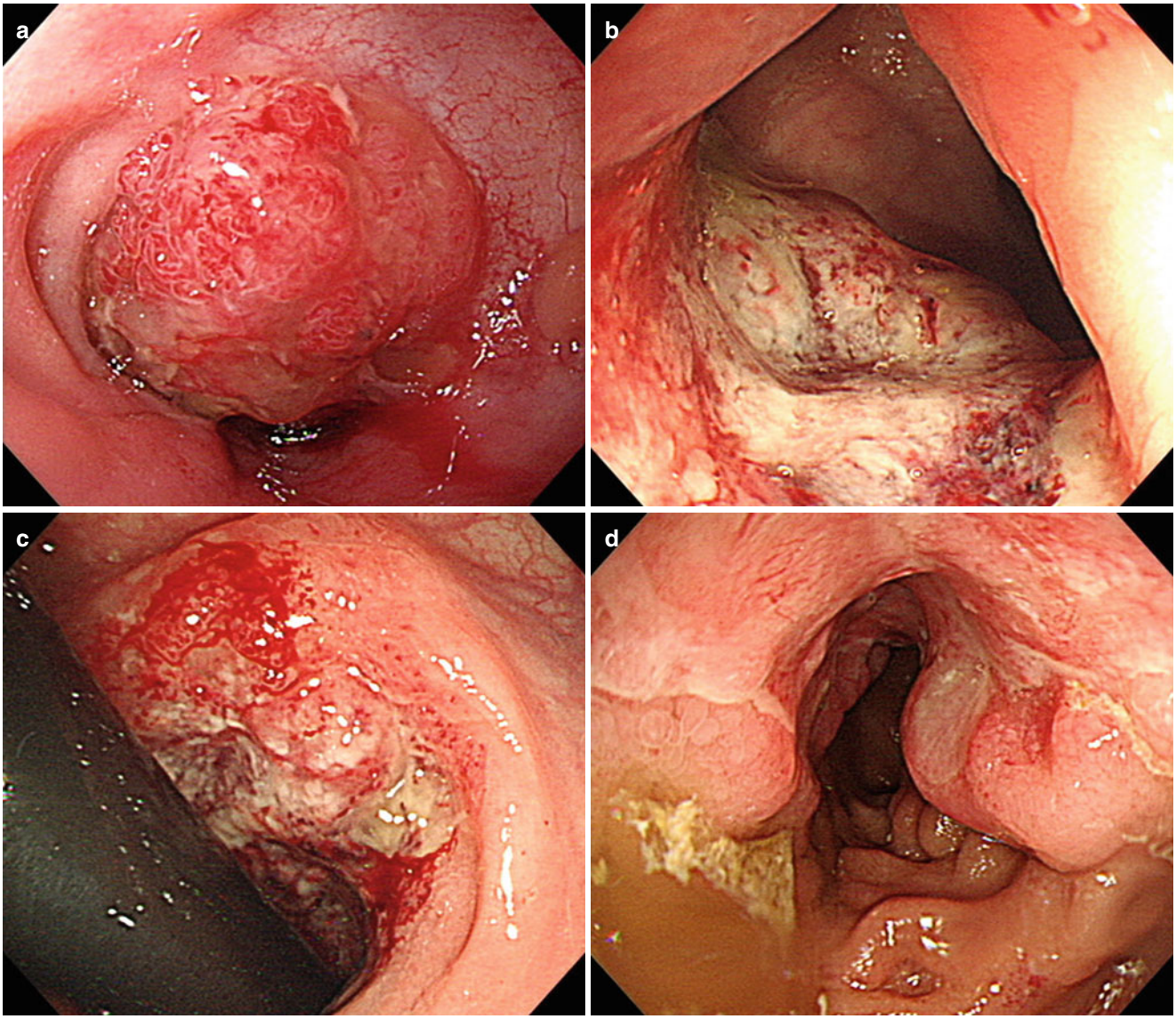

Fig. 19.38 Intestinal cancers developed in patients with long-standing CD. (**a**) Adenocarcinoma of ascending colon. (**b**, **c**) Rectal adenocarcinoma. (**d**) Ileal adenocarcinoma

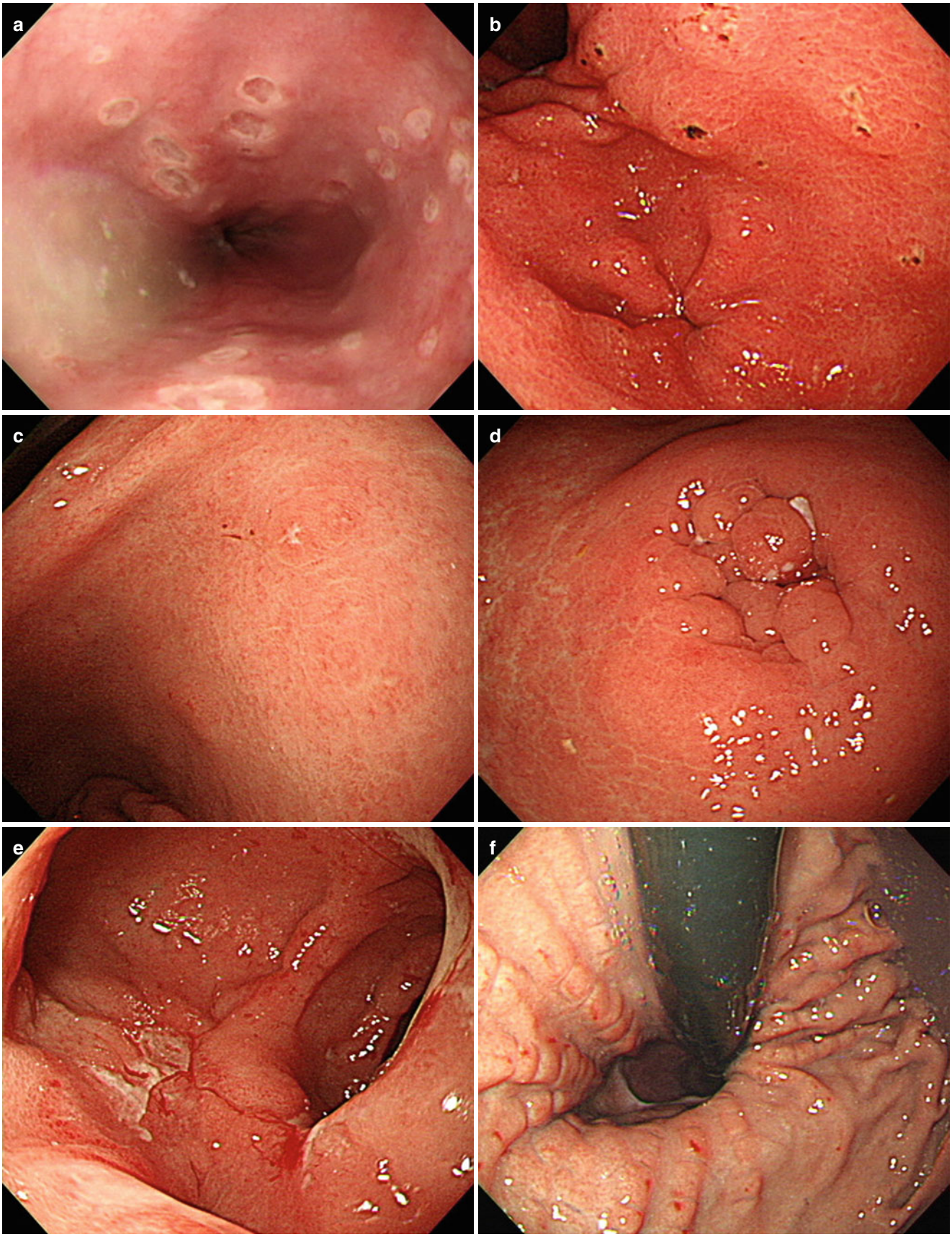

Fig. 19.39 Upper GI tract lesions in patients with CD. (**a**) Multiple esophageal ulcers. (**b**) Multiple erosions in gastric antrum. Noncaseating granuloma was observed in pathologic examination. (**c**) Biopsy of tiny aphthous erosion in the posterior wall of antrum showed noncaseating granuloma. (**d**) Pyloric stricture. (**e**) In duodenal bulb, multiple ulcers with luminal deformity are observed. Pathologic examination showed noncaseating granuloma. (**f**) At cardia of stomach, multiple short transverse grooves are observed. This "bamboo-joint-like appearance" is known to be more frequently observed in patients with CD. Clinical significance of these lesions is unclear

19.5 Intestinal Behçet's Disease

19.5.1 Clinical Manifestations

Behçet's disease (BD) is a chronic inflammatory disease that can involve all organs of body. The etiology of BD is uncertain, but the main pathologic feature is considered to be vasculitis [5]. The main clinical features of BD are recurrent oral ulcers, recurrent genital ulcers, eye involvement, and skin lesions. The diagnostic criteria for BD developed by International Study Group for BD is shown in Table 19.3. Gastrointestinal tract can also be involved in BD and it is regarded as additional features of systemic BD. In contrast to simple criteria by International Study Group for BD (Table 19.3), GI lesions are also adopted as minor feature for diagnosis according to the criteria suggested by BD Research Committee of Japan (Table 19.4). The usual symptoms of GI BD are abdominal pain, diarrhea, and hematochezia. As complications, bleeding, luminal stricture, and perforation can develop. The treatment strategy for gastrointestinal BD is not established yet. Usually, drugs used for managing CD are also used for GI BD. Patients with GI BD sometimes need intestinal resection due to medical intractability, perforation, and stricture. Young age at diagnosis, male gender, eye lesions, central nervous system involvement, and vascular involvement are considered to be poor prognostic factors for BD [5].

Table 19.3 Diagnostic criteria for BD (International Study Group for BD) [6]

Recurrent oral ulceration
Plus 2 of the followings:
Recurrent genital ulceration
Eye lesions
Skin lesion
Positive pathergy test

Table 19.4 Diagnostic criteria for BD (BD Research Committee of Japan) [7]

I. *Major*
1. Recurrent aphthous ulceration of the oral mucous membrane
2. Skin lesions:
Erythema nodosum
Subcutaneous thrombophlebitis
Folliculitis, acne-like lesions
Cutaneous hyperirritability
3. Eye lesions:
Iridocyclitis
Chorioretinitis, retinouveitis
Definite history of chorioretinitis or retinouveitis
4. Genital ulcers
II. *Minor*
1. Arthritis without deformity and ankylosis
2. Gastrointestinal lesions characterized by ileocecal ulcers
3. Epididymitis
4. Vascular lesions
5. Central nervous system symptoms
III. *Diagnosis*
1. Complete type: 4 major features
2. Incomplete type:
(a) Three major features
(b) 2 major + 2 minor features
(c) Typical ocular symptom + 1 major or 2 minor features
3. Suspected type:
(a) Two major features
(b) 1 major + 2 minor features

19.5.2 Endoscopic Features

GI involvement of BD is usually manifested as mucosal ulcers and ulcers are most frequently observed in the ileocecal area [5]. Typical features of BD are single ulcer or a few ulcers of the ileocecal area. However, multiple ulcers can also be observed. When the size of ulcer is small, it shows the aphthoid feature, round shape, or ovoid shape (Fig. 19.40).

Large ulcer also shows round or ovoid shape. Typically, the margin of ulcer is discrete and clear. The base of ulcer is usually even and is covered with white or yellow and thick mucus. The surrounding mucosa shows normal appearance (Fig. 19.41).

Sometimes, ulcer is so huge that the whole feature of lesions is hard to be characterized. When ulcer is large, the surrounding mucosa commonly shows nodular elevation and sometimes hyperemia. Therefore, the lesion is sometimes misdiagnosed as malignant lymphoma or adenocarcinoma. However, it can be differentiated from malignant lesions because friability is not observed in the surrounding mucosa of ulcer in BD. Because the ulcers of BD are usually large and deep, scar and deformity are usually observed after the healing of ulcers (Fig. 19.42).

As complications, stricture of lumen or fistula can develop. Fistula due to deep penetrating ulcer is more common than stricture (Fig. 19.43).

After resection of bowel, endoscopic recurrence is commonly observed. Ulcers are commonly observed along the anastomotic site (Fig. 19.44).

Ulcers of GI BD can develop in any site of GI tract. When ulcers with typical features in patients with compatible clinical backgrounds are observed, ulcer related with GI BD should be suspected (Fig. 19.45).

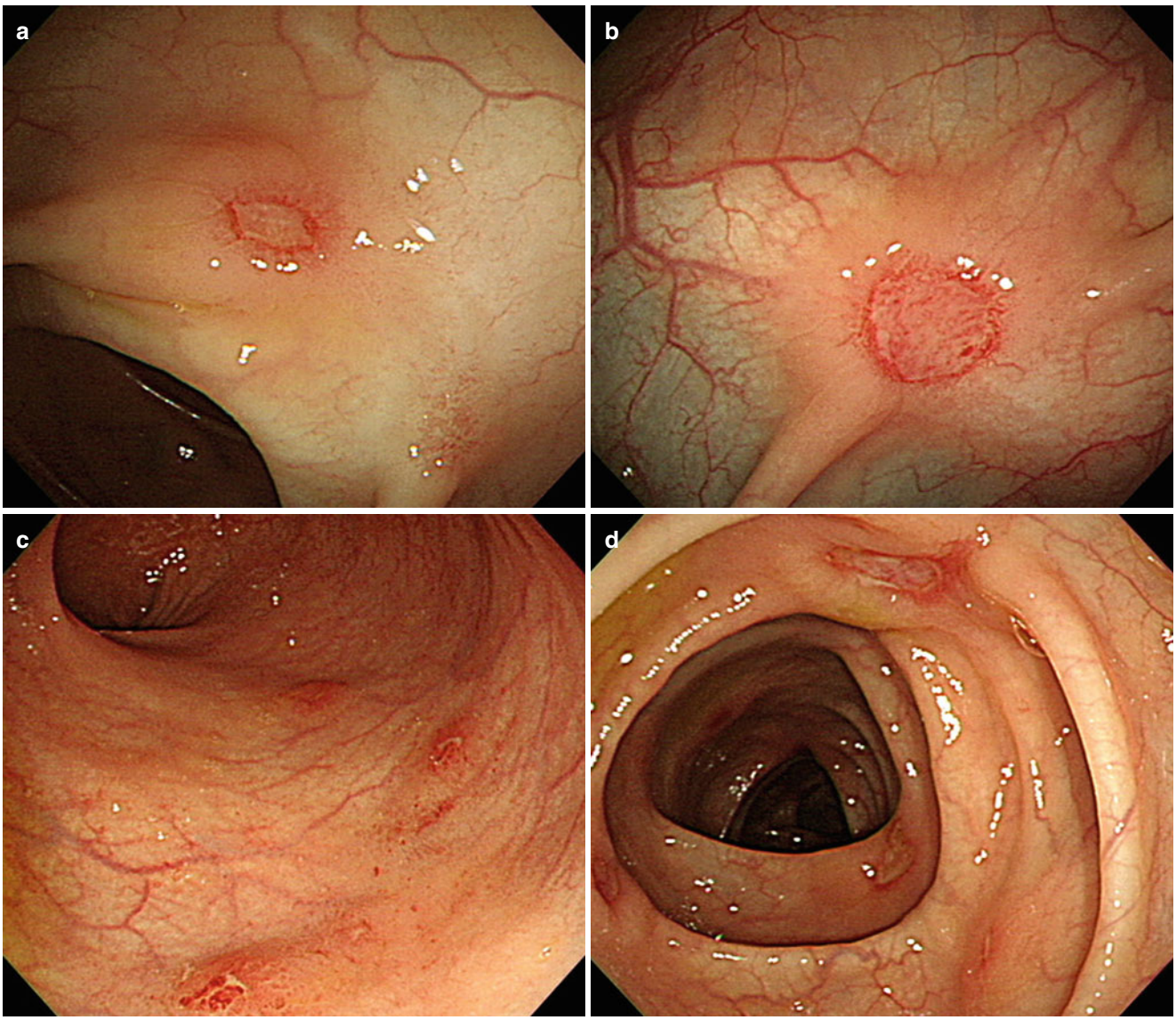

Fig. 19.40 Small ulcers of GI BD. (**a**, **b**) Small round ulcer with discrete margin. (**c**) Multiple aphthous ulcers with hyperemic rim. (**d**) Multiple ovoid ulcers of colon

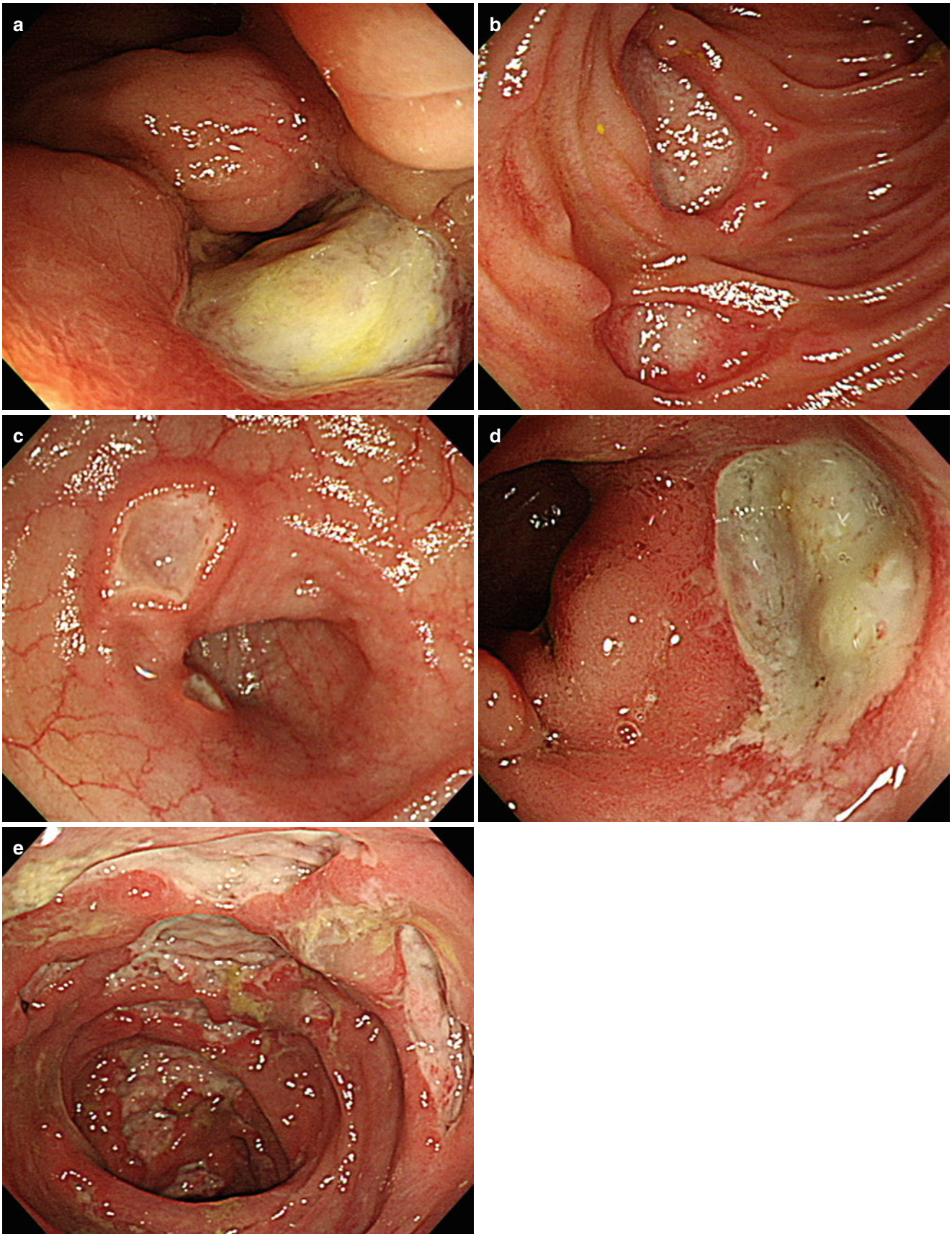

Fig. 19.41 Various ulcers of GI BD. (**a**, **b**) Ovoid ulcers with discrete margin and clean base. (**c**) Ovoid ulcers of terminal ileum. The surrounding mucosa shows slight elevation. (**d**) Ovoid ulcer of terminal ileum just proximal to ileocecal valve. The base of ulcer is covered with thick mucus. (**e**) Multiple deep ulcers with various shapes

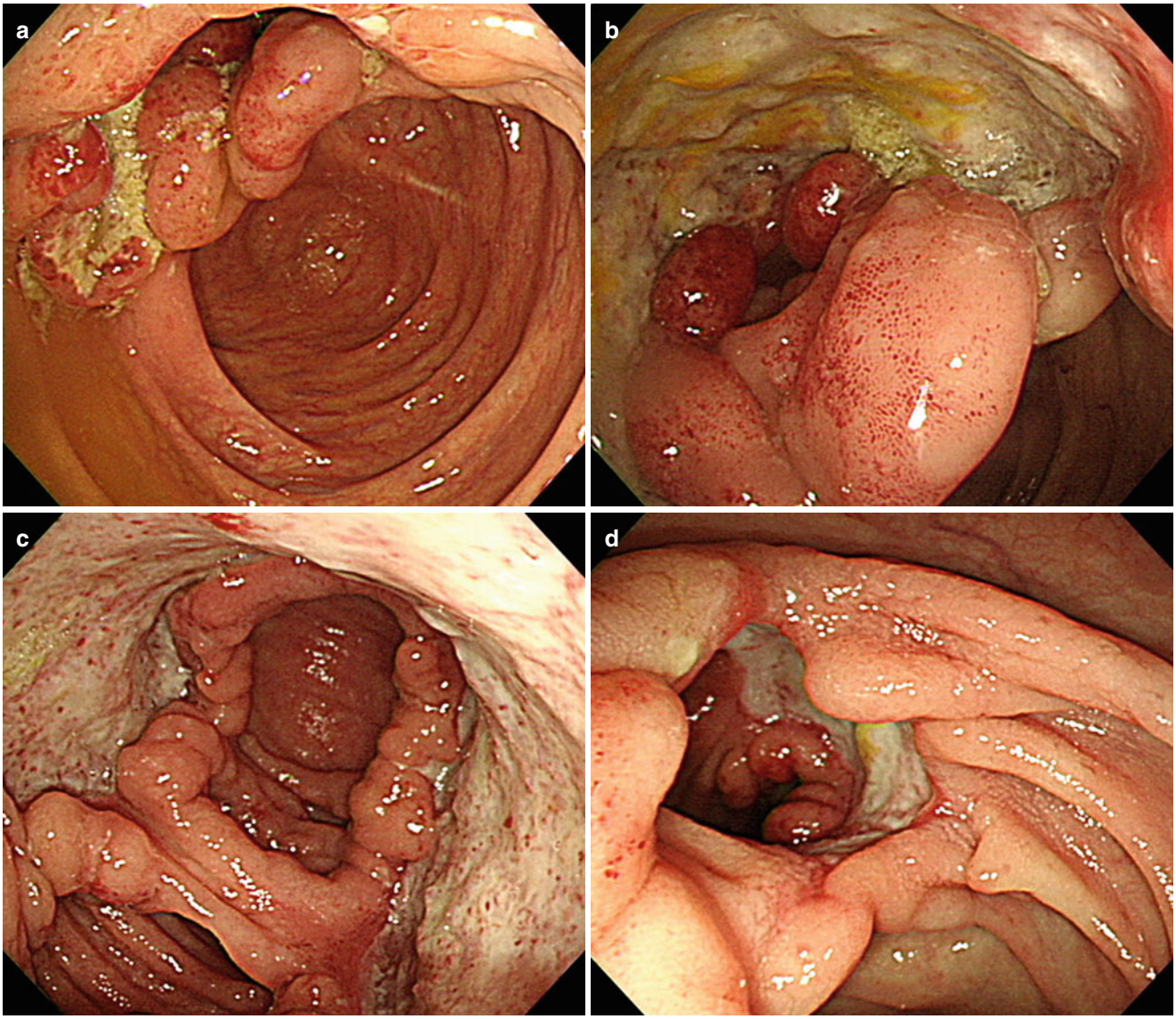

Fig. 19.42 Huge ulcers of GI BD. (**a**) Geographic ulcer is observed in ileocecal valve. (**b**) After intubation into ileocecal valve, huge circumferential deep ulcer is observed. (**c**) In terminal ileum, huge circumferential ulcer with discrete margin is observed. (**d**) Discrete ulcer is observed in the lateral side of ileocecal valve. (**e**) Closer view of (**d**). The margin of ulcer is sharp and nodular elevation of the surrounding mucosa is noted. (**f**) After healing of (**d**) and (**e**), patulous ileocecal valve with scar is observed. (**g**) Discrete ulcer of ascending colon. Converging folds to lesion suggest that the ulcer is a chronic lesion. Due to the elevated surrounding mucosa, lesion can be misdiagnosed as ulcerofungating cancer. However, in contrast to malignant ulcer, the margin is clear and friability of mucosa is not noted. (**h**) After healing of (**g**), scar with luminal deformity is noted

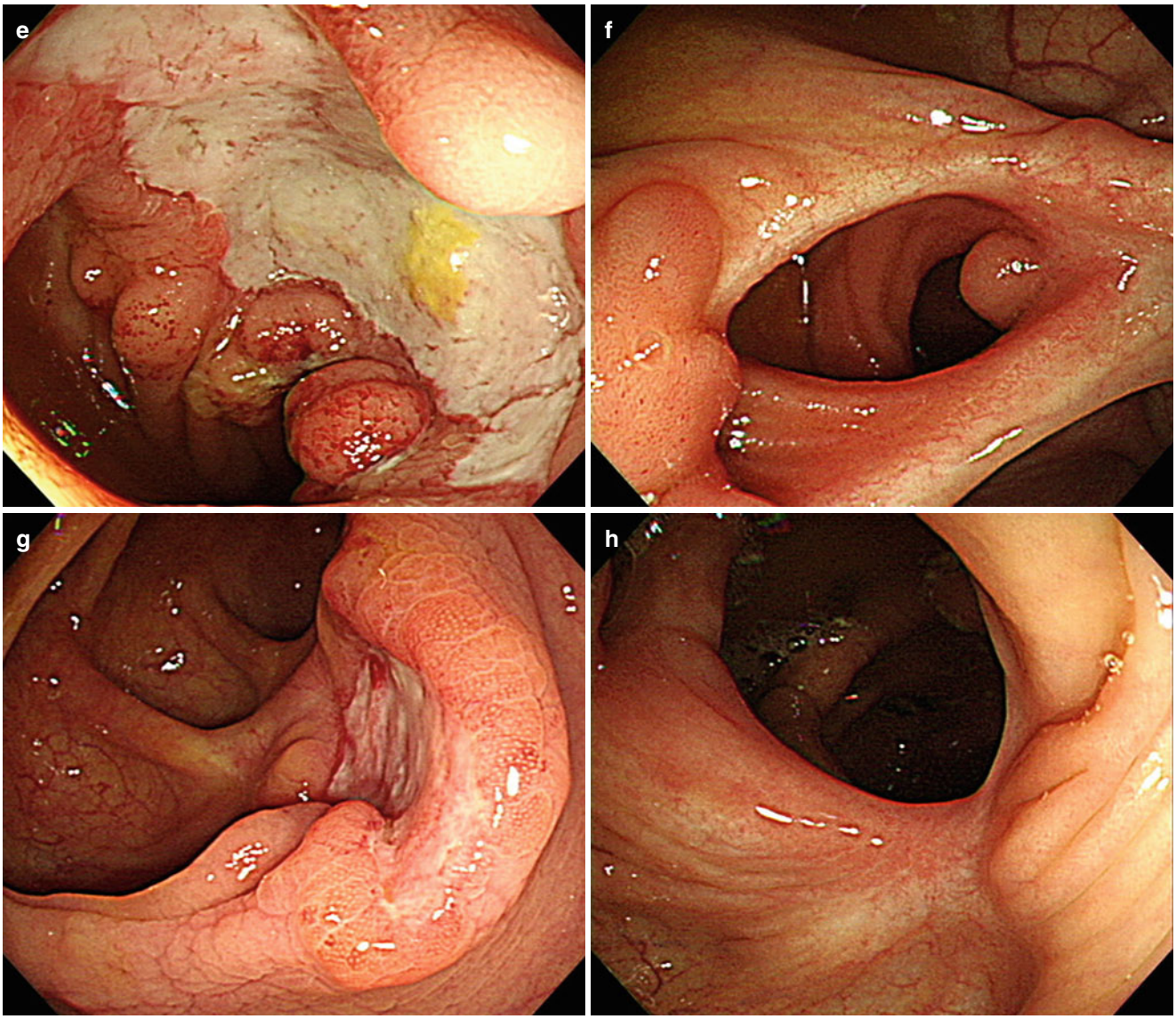

Fig. 19.42 (continued)

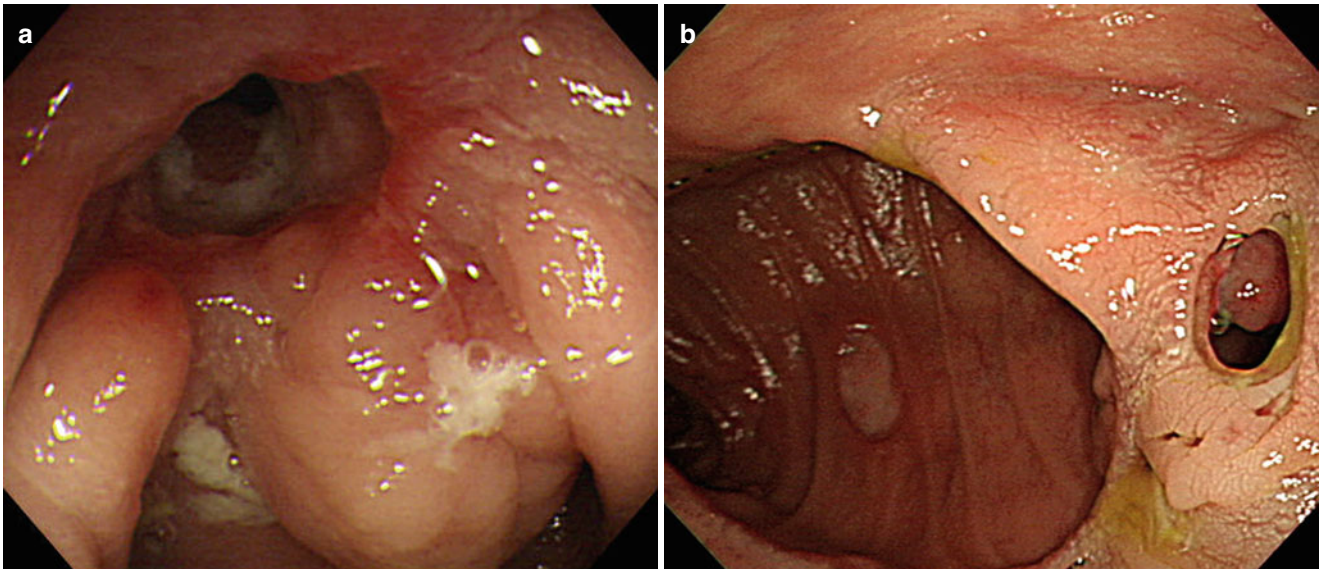

Fig. 19.43 Complications of GI BD. (**a**) Ileocecal stricture with ulceration. (**b**) Opening of fistula is observed in the terminal ileum (*right side* of the figure)

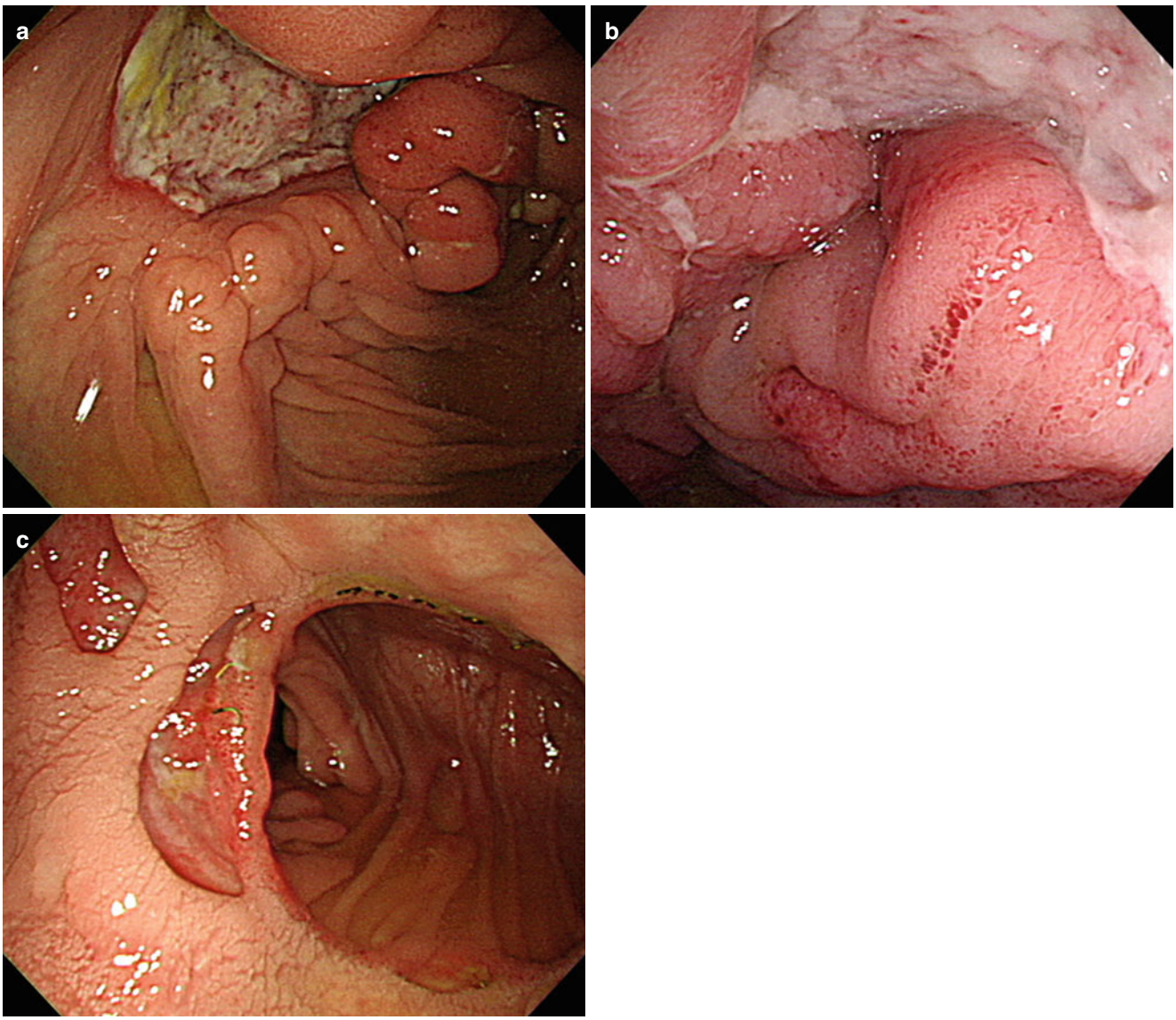

Fig. 19.44 Anastomotic recurrence. (**a**) Discrete ulcer in the ileocolic anastomosis. (**b**) Huge encircling ulcer in the ileocolic anastomosis. (**c**) Circular ulcer is noted in the ileocolic anastomosis

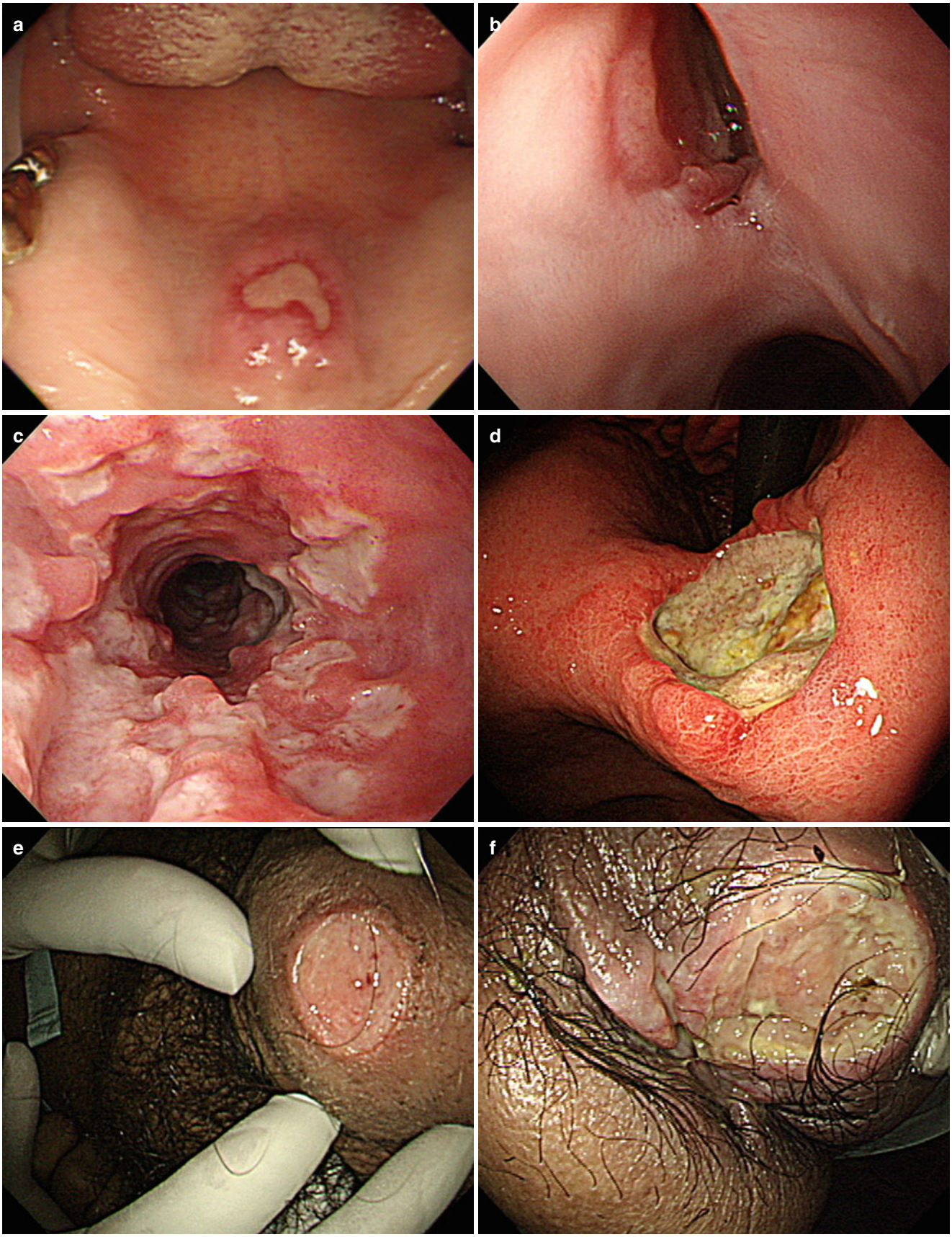

Fig. 19.45 Extracolonic ulcers. (**a**) Oral ulcer. (**b**) Deep penetrating esophageal ulcer (*upper side* of the figure). (**c**) Multiple esophageal ulcers. (**d**) Discrete gastric ulcer in the angle. (**e**) Scrotal ulcer. (**f**) Punched-out ulcer in the perianal area

19.6 Endoscopic Differential Diagnosis

19.6.1 UC *vs.* CD [3] (Table 19.5)

In some cases with pure colonic inflammation, UC and CD cannot be differentiated based on colonoscopic features. In that case, the terminology "colonic IBD unclassified (IBDU)" is used.

19.6.2 CD *vs.* GI BD [3] (Table 19.6)

Table 19.5 Endoscopic differential diagnosis of UC and CD [3]

		UC	CD
Distribution of lesions	Continuity	Continuous	Not continuous
	Symmetricity	Symmetric	Not symmetric
	Rectal involvement	Nearly always	Commonly not
Mucosal inflammation	Vascularity	Blurred	Not blurred
	Hyperemia	Usually present	Rare or not severe
	Granularity	Common	Rare
	Friability	Common	Rare
	Bleeding	Common	Rare
	Mucosa around ulcers	Inflamed	Commonly normal
Ulcers	Aphthous ulcer	Not common	Common
	Longitudinal ulcer	Not common	Common
	Serpiginous ulcer	Not common	Common
	Large ulcer (>1 cm)	Not common	Common
Others	Cobblestone appearance	Not common	Common
	Stricture	Not common	Common

Table 19.6 Different characteristics of ulcers between CD and GI BD [3]

Characteristics of ulcer	CD	GI BD
Distribution	More widely distributed	Usually ileocecal area
Number	Usually multiple	One/a few
Size	Various	Usually larger than in CD
Depth	Shallower	Deeper
Shape	Various: longitudinal, serpiginous, or cobblestone-like	Round or oval, thick mucus in base
Margin	Less discrete	More discrete

References

1. Waye JD. Endoscopy in inflammatory bowel disease: indications and differential diagnosis. Med Clin North Am. 1990;74:51–65.
2. Pera A, Bellando P, Caldera D, et al. Colonoscopy in inflammatory bowel disease. Diagnostic accuracy and proposal of an endoscopic score. Gastroenterology. 1987;92:181–5.
3. Yang SK, Byeon JS. Colonoscopy: diagnosis & treatment. Seoul: Koonja; 2009.
4. Schroeder KW, Tremaine WJ, Ilstrup DM. Coated oral 5-aminosalicylic acid therapy for mildly to moderately active ulcerative colitis. A randomized study. N Engl J Med. 1987;317:1625–9.
5. Yang SK. Intestinal Behçet's disease. Intest Res. 2005;3:1–10.
6. International Study Group for Behçet's Disease. Criteria for diagnosis of Behçet's disease. Lancet. 1990;335:1078–80.
7. Mizushima Y, Inaba G, Mimura Y, Ono S. Diagnostic criteria for Behçet's disease in 1987, and guidelines for treating Behçet's disease. Saishin Igaku. 1988;43:382–91 (in Japanese).