inflammation and bile ductule degeneration. Small-duct sclerosing cholangitis may progress to cirrhosis.

Biliary tract disease includes an increased incidence of gallstones and of primary sclerosing cholangitis (PSC). PSC is a chronic cholestatic liver disease marked by fibrosis of the intrahepatic and extrahepatic bile ducts; it occurs1% to 4% of patients with UC and less often in those with Crohn's disease. Overall, about 70% of patients with PSC have UC. Fibrosis leads to strictures of the bile ducts, which in turn may lead to recurrent cholangitis (with fever, right upper quadrant pain, and jaundice) and progression to cirrhosis. In addition, about 10% of patients develop cholangiocarcinoma. Medical or surgical therapy for the IBD does not modify the course of PSC, and most patients progress to cirrhosis and may require liver transplantation.

The two classic dermatologic manifestations of IBD are pyoderma gangrenosum and erythema nodosum. Pyoderma gangrenosum occurs in about 5% of patients and is characterized by a discrete ulcer with a necrotic base, usually on the legs. The ulcer may spread and become large and deep, destroying soft tissues. Pyoderma parallels the bowel activity in 50% of cases. Treatment is usually with systemic or intralesional steroids, or both. Other treatment options include dapsone, cyclosporine, and the anti-TNF agents. Erythema nodosum occurs in 10% of IBD patients, usually with peripheral arthropathy, and produces raised, tender nodules, usually over the anterior surface of the tibia. Erythema nodosum responds to treatment for the underlying bowel disease. A less common dermatologic manifestation of IBD is Sweet's syndrome, or acute febrile neutrophilic dermatosis. This condition is characterized by the sudden onset of fever, leukocytosis, and tender, erythematous, well-demarcated papules and plaques that show dense neutrophilic infiltrates on histologic examination.

Ocular manifestations of IBD, including uveitis and episcleritis, occur in 1% to 5% of patients. Uveitis (or iritis) is an inflammatory lesion of the anterior chamber that produces blurred vision, photophobia, headache, and conjunctival injection. Local therapy includes steroids and atropine. Episcleritis produces burning eyes and scleral injection without vision deficits and is treated with topical steroids.

DIAGNOSIS AND DIFFERENTIAL DIAGNOSIS

The diagnosis of IBD is based on a constellation of clinical features, laboratory tests, and endoscopic, radiographic, and histologic findings. Laboratory tests are not specific and usually reflect inflammation (leukocytosis) or anemia. Perinuclear antineutrophil cytoplasmic antibody (pANCA) is positive in up to 70% of patients with UC but is rarely positive in patients with Crohn's disease, whereas anti–*Saccharomyces cerevisiae* antibodies (ASCA) are common in Crohn's disease but rarely found in UC (Table 37-2). Additional markers have improved the sensitivity and specificity of serologic testing, including antibodies to OmpC (*Escherichia coli* outer membrane porin C) and antibodies to bacterial flagellins CBir1, FlaX, and A4-Fla2.

Colonoscopy in patients with UC reveals a granular mucosa, decreased vascular markings, decreased mucosal reflection, exudate, and superficial ulcerations (Fig. 37-4). In more severe cases, the mucosa is friable, with deeper ulcerations. Patients with long-standing disease have *pseudopolyps*, which represent islands of normal tissue in regions of previous ulceration. In Crohn's disease (Fig. 37-5), endoscopic examination may show aphthoid erosions, deep linear or stellate ulcers, edema, erythema, exudate, and friability with intervening areas of normal mucosa (skip lesions). However, a diagnosis of *indeterminate* colitis is occasionally made because of an overlap of findings. For example, colonic Crohn's disease may produce superficial continuous rectal involvement similar to that seen in UC. Similarly, chronic UC can infrequently result in inflammation of the terminal ileum, called *backwash ileitis*. In many patients with indeterminate colitis,

TABLE 37-2 DIFFERENTIATING FEATURES OF ULCERATIVE COLITIS AND CROHN'S DISEASE

	ULCERATIVE COLITIS	CROHN'S DISEASE
Site of involvement	Involves colon only Rectum almost always involved	Any area of the gastrointestinal tract Rectum usually spared
Pattern of involvement	Continuous	Skip lesions
Diarrhea	Bloody	Usually nonbloody
Severe abdominal pain	Rare	Frequent
Perianal disease	No	In 30% of patients
Fistula	No	Yes
Endoscopic findings	Erythematous and friable Superficial ulceration	Aphthoid and deep ulcers Cobblestoning
Radiologic findings	Tubular appearance resulting from loss of haustral folds	String sign of terminal ileum RLQ mass, fistulas, abscesses
Histologic features	Mucosa only Crypt abscesses	Transmural Crypt abscesses, granulomas (about 30%)
Smoking	Protective	Worsens course
Serology	pANCA more common	ASCA more common

ASCA, Anti–*Saccharomyces cerevisiae* antibodies; pANCA, perinuclear antineutrophil cytoplasmic antibody; RLQ, right lower quadrant.

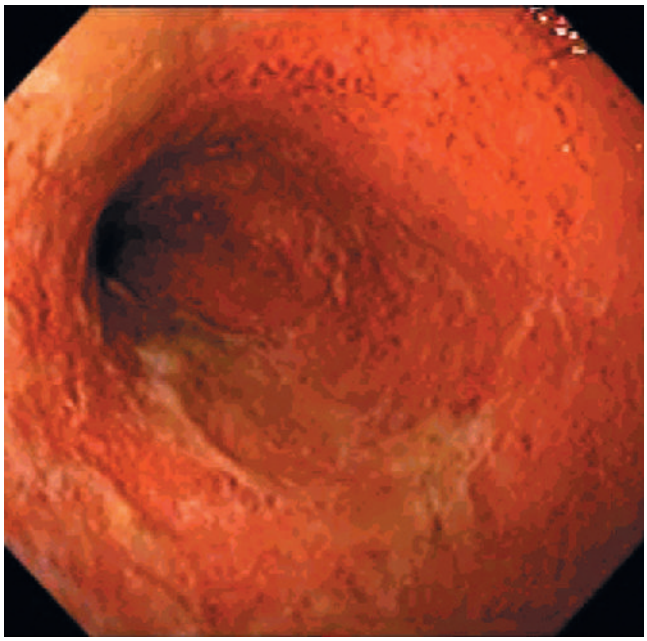

FIGURE 37-4 Endoscopic image of ulcerative colitis demonstrates diffuse inflammation characterized by erythema, edema, friability, and hemorrhage.