41
Ulcerative Colitis

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Introduction

Ulcerative colitis (UC) is an idiopathic inflammatory bowel disease more common in developed countries and most often presenting between ages 15 and 30 with an additional peak in the sixth decade. Many possible etiologies have been implicated, including genetic mutations, environmental factors, infectious agents, and immunologic disturbances; however, no cause has been conclusively determined. The development of ulcerative colitis appears to be multifactorial; genetic markers have been identified, the disease runs in families, and it also responds to immunosuppressive medications (Fig. 41.1). Involvement of the rectum with continuous distribution of inflammation proximally, not involving the small intestine, is the classic feature that distinguishes UC from other inflammatory bowel conditions such as Crohn's disease (CD). However, it is important to note that this presentation may vary, making the diagnosis difficult in certain cases.

Beside symptoms of UC affecting quality of life, this condition has also been found to be a strong, independent risk factor for colon and rectal cancer as well as primary sclerosing cholangitis (with subsequent cirrhosis and bile duct cancer); UC requires close surveillance to avoid these potentially fatal complications.

Clinical Presentation

Signs and symptoms of ulcerative colitis can be divided into gastrointestinal (most common) or extraintestinal. The typical clinical presentation includes crampy abdominal pain, urgency, and watery or bloody diarrhea mixed with mucus. Other symptoms include anemia, weight loss, malnutrition, and failure to thrive resulting from long-standing, severe disease. A variety of extraintestinal symptoms may be present, including but not limited to: aphthous ulcers, uveitis, arthritis, erythema nodosum, pyoderma gangrenosum, deep vein thrombosis, and pericarditis. Presentation may vary from a mild insidious course with only diarrhea and minimal abdominal pain to a more sudden onset of explosive bloody diarrhea, severe abdominal pain with fever, tachycardia, and dehydration. A small subset of patients will present with fulminant toxic colitis that may or may not involve significant colonic dilatation but increases risk of bowel perforation, sepsis, acidosis, and shock. This presentation, while rare, can be potentially fatal and requires immediate recognition and treatment.

Diagnosis

No specific laboratory tests can diagnose ulcerative colitis, and physical exam is usually nonspecific. Endoscopy remains the definitive diagnostic modality for UC and other inflammatory bowel diseases. Flexible sigmoidoscopy with biopsy is usually sufficient for diagnosis; however, a complete colonoscopy may be required in cases where uncertainty remains, allowing for evaluation of the terminal ileum. Utilizing colonoscopy in the setting of acute inflammation, however, increases the risk of perforation significantly and is contraindicated when the presentation is severe. Typical findings on endoscopy include friable mucosa, exudates, pseudopolyps, superficial ulcers, and diffuse erythema. The rectum is invariably involved, with potential continuous spread throughout the entire colon.

Histologic examination will reveal inflammation of the mucosa and submucosa, crypt abscesses, extensive infiltration of inflammatory cells, mucin depletion, and disturbed crypt architecture. Antineutrophil cytoplasmic antibody (pANCA) serum marker may be positive.

Other diagnostic methods include plain radiographs to rule out perforation in the acutely ill patient with severe abdominal pain. Computed tomography (CT) scan or barium enema may reveal shortening of the colon and loss of haustral markings; however, these findings are not sufficient for definitive diagnosis and cannot differentiate between other types

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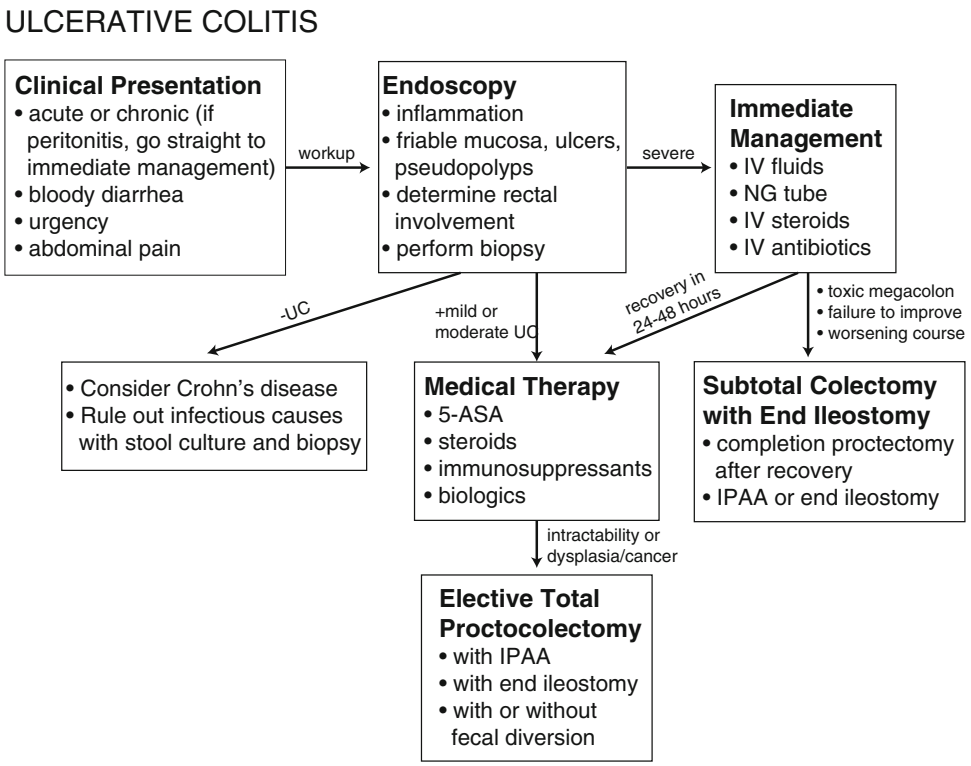

FIG. 41.1 Treatment algorithm for ulcerative colitis. *UC* ulcerative colitis, *IPAA* ileal pouch-anal anastomosis, *IV* intravenous, *NG* nasogastric, *5-ASA* 5-aminosalicylic acid

of inflammatory or infectious diseases. In order to rule out infectious causes of colitis, stool cultures should be routinely sent in addition to blood cultures in a severely ill patient.

Treatment

Initial treatment in most cases of ulcerative colitis begins with medical therapy in order to relieve symptoms and induce remission. For mild symptoms, such as watery diarrhea, treatment with medications to slow intestinal transit, such as loperamide or bulking agents, may be sufficient.

For more severe presentations, aminosalicylates, steroids, and other immunosuppressants are the mainstay of treatment. Aminosalicylates (5-ASA, delayed release, oral, suppositories, enemas) are common first-line medications for moderate acute UC as well as maintenance therapy for long-term remission. Steroid therapy is typically used for the acute severe disease, but should not be used for long-term therapy secondary to serious, well-described side effects. Immunosuppressants including azathioprine, 6-mercaptopurine, and methotrexate are most effective for long-term maintenance therapy and not for acute presentation since their treatment effects may take months to manifest. More recently, biologics (antitumor necrosis factor medications) have also been shown to treat acute and severe UC and effectively maintain remission as well.

Indications for surgery in UC include:

1. Failure or noncompliance with medical therapy
2. Inability to tolerate medication side effects
3. Massive or persistent colonic bleeding not amenable to less invasive therapy (endoscopy or embolization)
4. Fulminant colitis or toxic megacolon
5. Chronic anemia, malnutrition, failure to thrive
6. Perforation
7. Obstruction
8. Dysplasia or cancer

One advantage of surgical treatment in UC is that, unlike Crohn's, complete resection of the colon and rectum is curative. Elective surgical options include total proctocolectomy with end (permanent) ileostomy or total proctocolectomy with ileal pouch-anal anastomosis (IPAA). Both options have the advantage of removing almost all potential tissue involved in the disease process and therefore eliminating almost all risk of disease recurrence or progression to colon cancer (small amount of distal rectum remains unless mucosectomy performed or anal canal removed). The main drawback of end ileostomy is the requirement of permanent ileostomy; however, this may be necessary in patients with poor sphincter function (previous surgery, trauma, elderly patients). Total proctocolectomy with IPAA preserves the anal sphincters and creates a reservoir (the ileal pouch) that allows the patient to defecate through the

anus with relatively normal bowel function. This operation is frequently performed in two stages and can be performed laparoscopically. The first stage involves removal of the colon and rectum, formation of the ileal pouch anal anastomosis, and creation of the diverting ileostomy. The second is reversal of the loop ileostomy after the IPAA heals and the patient recovers. Alternatively, few colorectal surgeons question the need for fecal diversion and perform a one-stage procedure.

Patients presenting with fulminant colitis require immediate attention. Aggressive intravenous fluid resuscitation, nasogastric decompression, and broad-spectrum intravenous antibiotics should begin without delay. Usually, medical treatment is initiated with high-dose intravenous steroids and a trial of conservative therapy with bowel rest and possible total parental nutrition. The patient should be closely monitored for 24–48 h. If the patient's condition worsens or fails to improve during this time, surgical intervention is indicated. In these circumstances when patients are critically ill, a subtotal colectomy (colectomy without proctectomy) is the procedure of choice due to its shorter operative time and less operative morbidity. Subtotal colectomy leaves the option for future sphincter preservation and pouch creation. In patients with severe UC, malnutrition, or anemia or colonic bleeding with hemodynamic instability or persistent bleeding with failure of nonsurgical management, a subtotal colectomy is recommended. After an appropriate recovery, completion proctocolectomy can be done with IPAA or end ileostomy.