

# IBD

Thamer Bin Traiki

Colorectal & Surgical Oncology

- *Inflammatory bowel disease* (IBD) is a term generally used to denote two diseases of unknown etiology with similar general characteristics:
  - *Ulcerative colitis* and
  - *Crohn's disease*.

- The distinction between the two entities can usually be established based on clinical and pathologic criteria, including
  - Hx & EX,
  - Radiologic and Endoscopic studies,
  - Gross appearance, and
  - Histology.

- About 10% to 15% of patients with inflammatory disease confined to the colon, a clear distinction cannot be made, and the disease is labeled *indeterminate colitis*.
- The medical and surgical management of ulcerative colitis and Crohn's disease often differ significantly.

# **CROHN'S DISEASE**

# CD

- A chronic, transmural inflammatory disease of the gastrointestinal tract of unknown cause.
- Crohn's disease can involve any part of the alimentary tract from the mouth to the anus but most commonly affects the small intestine, colon, rectum & anus.

Both medical and surgical treatments are  
palliative not curative.

# History of the Disease

- In retrospect, case descriptions of what appeared to be Crohn's disease date back to at least 1612, when Fabry reported on the death of a boy experiencing severe abdominal pain.
  - Autopsy revealed a contracted ulcerated cecum and ileum with complete bowel obstruction.



- In 1761, Morgagni described a case of an inflamed ileum with perforation and thickened mesentery in a young man with a history of diarrhea and fever.
- In 1913, the Scottish surgeon Dalziel described nine cases of intestinal inflammatory disease.

- The landmark paper by Crohn, Ginzburg, and Oppenheimer in 1932 that provided, in eloquent detail, the pathologic and clinical findings of this inflammatory disease in young adults.

# Epidemiology

- Crohn's disease primarily attacks young adults in the second and third decades of life.
  - However, a bimodal distribution is apparent with a second, smaller peak occurring in the sixth decade of life.
- The risk for developing Crohn's disease is about two times higher in smokers than in nonsmokers.

# Etiology

- Unknown.
- Potential causes have been proposed,
  - Infectious,
  - Immunologic, and
  - Genetic.
    - The single strongest risk factor for developing disease is having a relative with Crohn's disease.
- Other possibilities that have met with various levels of enthusiasm include environmental and dietary factors, smoking, and psychosocial factors.

- Smoking is known to exacerbate existing Crohn's disease and can accelerate its recurrence after resection.
- The component of cigarette smoke that is responsible for these deleterious effects on the clinical course of Crohn's disease is not known

# Pathology

- The most common sites of occurrence of Crohn's disease are the small intestine & colon.
- The involvement of both large and small intestine has been noted in about 55% of patients.
- 30 % of patients present with small bowel disease alone.
- 15%, the disease appears limited to the large intestine.

- The disease process is *discontinuous and segmental*.
- In patients with colonic disease, *rectal sparing* is characteristic of Crohn's disease and helps to distinguish it from ulcerative colitis.
- Perirectal and perianal involvement occurs in about 1/3 of patients with Crohn's disease, particularly those with colonic involvement.

- Crohn's disease can also involve the mouth, esophagus, stomach, duodenum, and appendix.
- Involvement of these sites can accompany disease in the small or large intestine, but in only rare cases have these locations been the only apparent sites of involvement.



# Microscopy

- Mucosal and submucosal edema may be noted microscopically before any gross changes.
- A chronic inflammatory infiltrate appears in the mucosa and submucosa and extends *transmurally*.
- This inflammatory reaction is characterized by extensive edema, hyperemia, lymphangiectasia, an intense infiltration of mononuclear cells, and lymphoid hyperplasia.

- Well-formed lymphoid aggregates in an edematous fibrotic submucosa is a classic histological feature of the disease.

- Characteristic histologic lesions of Crohn's disease are noncaseating granulomas with Langerhans' giant cells.
- Granulomas appear later in the course and are found in the wall of the bowel or in regional lymph nodes in 60% to 70% of patients

# Gross Pathological Features

- At Exploration:
  - Thickened grayish-pink or dull purple-red loops of bowel,
  - Thick gray-white exudate or fibrosis of the serosa.
  - *Skip areas* areas of diseased bowel separated by areas of grossly appearing normal bowel.
  - Extensive *fat wrapping* caused by the circumferential growth of the mesenteric fat around the bowel wall.

- With early acute intestinal inflammation, the bowel wall is hyperemic and boggy.
- As the inflammation becomes chronic, fibrotic scarring develops and the bowel wall becomes thickened and leathery in texture.

- Involved segments often are adherent to adjacent intestinal loops or other viscera, with internal fistulas common in these areas.
- The mesentery of the involved segment is usually thickened, with enlarged lymph nodes.

- On opening the specimen:
  - The earliest gross manifestations of Crohn's disease are the development of small mucosal ulcerations called *aphthous ulcers*.
  - Aphthous ulcers appear as red spots or focal mucosal depressions .
  - As the inflammation progresses, the aphthous ulcers enlarge and become stellate.

- The ulcers are characteristically linear and may coalesce to produce transverse sinuses with islands of normal appearing mucosa in between, thus giving the characteristic *cobblestone appearance*



- Mucosal ulcerations may penetrate through the submucosa to form intramural channels that can bore deeply into the bowel wall and create sinuses, abscesses, or fistulas.

# Clinical Manifestations

- The onset of disease is often insidious, with a slow and protracted course.
  - Characteristically, there are symptomatic periods of abdominal pain and diarrhea interspersed with asymptomatic periods of varying lengths.
  - With time, the symptomatic periods gradually become more frequent, more severe, and longer lasting

- The most common symptom is intermittent and colicky abdominal pain,
  - Most commonly in the lower abdomen.
  - The pain may be more severe and localized and may mimic the signs and symptoms of acute appendicitis.

- Diarrhea is the next most frequent symptom and is present, at least intermittently, in about 85% of patients.
  - In contrast to ulcerative colitis, patients with Crohn's disease typically have fewer bowel movements, and the stools rarely contain mucus, pus, or blood

- Systemic nonspecific symptoms
  - Low-grade fever (present in about 1/3),
  - Weight loss,
  - Loss of strength, and
  - Malaise.

- Perianal disease (fissure, fistula, stricture, or abscess) is common
- Perianal disease may be the sole presenting feature in 5% of patients and may precede the onset of intestinal disease by months or even years.
- *Crohn's disease should be suspected in any patient with multiple, chronic perianal fistulas.*

# Extraintestinal manifestations

- Present in 30% of patients.
  - The most common symptoms are skin lesions, which include erythema nodosum and pyoderma gangrenosum,
  - Arthritis and arthralgias,
  - Uveitis and iritis,
  - Hepatitis and Pericholangitis, and
  - Aphthous stomatitis

- Amyloidosis,
- Pancreatitis, and
- Nephrotic syndrome
- These symptoms may precede, accompany, or appear independent of the underlying bowel disease.



# Complications

- Obstruction
- Perforation.
- Fistulas occur between the sites of perforation and adjacent organs, such as loops of small and large intestine, the urinary bladder, the vagina, the stomach, and sometimes the skin,
- Localized abscesses.
- Toxic megacolon in patients with Crohn's colitis.
- Cancer

# Indications for operation

- Limited to complications
  - Intestinal obstruction,
  - Intestinal perforation with fistula formation or abscess,
  - Free perforation,
  - GI bleeding,
  - Urologic complications,
  - Cancer, and
  - Perianal disease.

- Elective

- Chronic subacute obstruction due to fibrotic strictures, adhesions or refractory disease
- Symptomatic disease unresponsive to, or poorly controlled by medical management
- Chronic relapsing disease on discontinuation of medical management and steroid dependency
- Complications of medical management (e.g. osteoporosis)
- Concerns about long-term immunosuppression, risk of malignancy and viral/atypical infections
- Perianal sepsis and fistula
- Enterocutaneous fistula
- Onset of malignancy, including colorectal adenocarcinoma and small bowel lymphoma
- Rarely, control of debilitating extra-colonic manifestations such as iritis and sacroiliitis.

- Emergency

- Fulminant colitis or acute small bowel relapse unresponsive to medical management
- Acute bowel obstruction
- Life-threatening haemorrhage
- Abscess or free perforation
- Perianal abscess.

# DDx

- Acute appendicitis
- Mesenteric Lymphadenitis
- Ovarian pathology
- *Salmonella* and *Shigella*;
- Intestinal TB;
- Acute distal ileitis may be a manifestation of early Crohn's disease, but it also may be unrelated, such as when it is caused by a bacteriologic agent (e.g., *Campylobacter* or *Yersinia*).
- *UC*
- protozoan infections, such as amebiasis, may present as an ileitis.
- In the immunocompromised host, rare infections, particularly mycobacterial & CMV.

**UC**

- A chronic disease that affects the *mucosa and submucosa*, with sparing of the muscularis of the rectum and colon.
- Surgery is curative.

# History

- Hippocrates described diarrheal diseases that were colitis-like well before 360 BC.
- Late 1800s that ulcerative colitis was distinguished clinically from common infectious enteritis.
- 1875, Wilks and Walter Moxon, described ulceration and inflammation of the entire colon in a young woman who had succumbed to severe bloody diarrhea, and it is more likely the first detailed account of ulcerative colitis.

# Epidemiology

- There appears to be a seasonal variation in the activity of the disease, with onset as well as relapse occurring statistically more often between August and January.
- Commonly affects patients younger than 30 years.
  - A small secondary peak in the incidence occurs in the sixth decade.



# Etiology

- Unknown
  - Environmental
  - Infectious
  - Genetic
- A family history of IBD is a significant risk factor.

- Smoking appears to confer a protective effect against the development of ulcerative colitis, as well as providing a therapeutic influence; nicotine has been reported to induce remission in some cases. This is in contrast to Crohn's disease, which is more common in smokers and appears to be aggravated by the habit.
- Patients who have had an appendectomy appear to be at increased risk for developing ulcerative colitis

# Pathology

- Despite the disease's name, ulceration of the mucosa is not invariably present.
- **Rectum** is invariably involved with the inflammatory process.
- In fact, rectal involvement (proctitis) is the *sine qua non* of the disease, and the diagnosis should be seriously questioned if the rectal mucosa is not affected.
- The mucosal inflammation extends in a *continuous* fashion for a variable distance into the more proximal colon.

- Diagnostic characteristic of ulcerative colitis is *continuous, uninterrupted inflammation of the colonic mucosa beginning in the distal rectum and extending proximally to a variable distance.*
  - This is in contrast to Crohn's disease, in which normal segments of colon (skipped areas) may be interspersed between distinct segments of colonic inflammation.
- The entire colon, including the cecum and appendix, may be involved in ulcerative colitis.
  - In contrast to Crohn's disease, ulcerative colitis does not involve the terminal ileum except in cases of backwash ileitis, when the ileal mucosa may appear inflamed in the presence of extensive proximal colonic involvement.

# Gross Appearance

- The typical gross appearance of ulcerative colitis is hyperemic mucosa.
- Friable and granular mucosa is common in more severe cases, and ulceration may not be readily evident, especially early in the course of the disease.
- Ulceration may appear and vary widely, from small superficial erosions to patchy ulceration of the full thickness of the mucosa.

- Pseudopolyps, or inflammatory polyps, represent regeneration of inflamed mucosa and are composed of a variable mixture of non-neoplastic colonic mucosa and inflamed lamina propria.

# Microscopy

- The typical microscopic finding in ulcerative colitis is inflammation of the mucosa and submucosa.
- The most characteristic lesion is the **crypt abscess**,
  - Collections of neutrophils fill and expand the lumina of individual crypts of Lieberkühn
  - Not specific for ulcerative colitis and can be seen in Crohn's disease and infectious colitis

# Clinical manifestations

- Ulcerative colitis and colonic Crohn's disease often have similar clinical presentations.
  - Both may present with diarrhea and the passage of mucus.
- Patients with ulcerative colitis tend to have more urgency than those with Crohn's disease,
  - likely because ulcerative colitis is invariably associated with distal proctitis.
- Rectal bleeding is also common in ulcerative colitis;
- Patients with the acute onset of ulcerative colitis often complain of abdominal discomfort, but the pain is seldom as severe as that found in patients with Crohn's disease.



# Extraintestinal Manifestations

- arthritis, ankylosing spondylitis, erythema nodosum, pyoderma gangrenosum, and primary sclerosing cholangitis.
- Arthritis, particularly of the knees, ankles, hips, and shoulders, occurs in about 20% of patients, typically in association with increased activity of the intestinal disease.

- Ankylosing spondylitis occurs in 3% to 5% & Primary sclerosing cholangitis (PSC) occurs in 5% to 8% of patients with ulcerative colitis.
- Colectomy has no effect on the course of these 2 conditions.

# Indications for surgery

- Fulminant colitis with toxic megacolon
- Massive bleeding,
- Intractable disease, and
- Dysplasia or carcinoma.
- Malnutrition and growth retardation may necessitate resection in pediatric and adolescent patients.

|                                | CD                      | UC                          |
|--------------------------------|-------------------------|-----------------------------|
| <b>Symptoms and Signs</b>      |                         |                             |
| Diarrhea                       | Common                  | Common                      |
| Rectal bleeding                | Less common             | Almost always               |
| Abdominal pain (cramps)        | Moderate to severe      | Mild to moderate            |
| Palpable mass                  | At times                | No (unless large cancer)    |
| Anal complaints                | Frequent (>50%)         | Infrequent (<20%)           |
| <b>Proctoscopic Findings</b>   |                         |                             |
| Anal fissure, fistula, abscess | common                  | Rare                        |
| Rectal sparing                 | Common (50%)            | Rare (5%)                   |
| Ulceration                     | Linear, deep, scattered | Superficial, universal      |
| Distribution                   | Skip areas              | Rectum extending proximally |