Inflammatory bowel disease

Objectives:

- What is the disease?
- Epidemiology of IBDs
- Pathophysiology of IBDs
- Ulcerative colitis
- Crohn's disease
-

Resources:

- Davidson's.
- Slides
- Surgical recall.
- Raslan's notes.
- 435 Teamwork

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Color index:

Notes , Important , Extra , Davidson's

Editing file

Feedback

Inflammatory bowel disease

Introduction:

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

- The distinction between the two entities can usually be established based on clinical history and examination and pathologic criteria, including:
    - History and physical examination (you can see signs of anemia and clubbing).
    - Radiologic and Endoscopic studies,
    - Gross appearance.
    - 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.
- In 20% of cases, grossly it looks like UC while microscopic features say it is CD
- Also, sometimes the biopsy looks like UC but the clinical features suggests CD.
- In medicine it's not very important to distinguish the two diseases because they have the same treatment. However, in surgery we have to distinguish the two diseases because ulcerative colitis could be treated completely with surgery. But if crohns is treated with surgery, it is very likely that the patient will relapse.

Crohn's disease Helpful Video 6:37

Definition:

- A **chronic**, **transmural** inflammatory disease of the GI 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 (terminal ileum), colon, rectum & anus.
- The most common sites of occurrence of Crohn's disease are the small intestine & colon.
- 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.
- 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.
- **If you suspect crohn's you should do a Colonoscopy.**

Etiology:

- Unknown, But Potential causes have been proposed:

  - Infectious (measles, listeria,pseudomonas and mycobacterium avium paratuberculosis).

  -Immunologic.

  -Genetic (Twins, Relatives, & children) (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.
- Most common sites of recurrence are the small intestine and colon.

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.

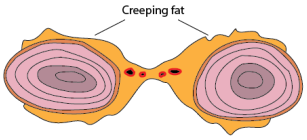
Diagnostic tests:

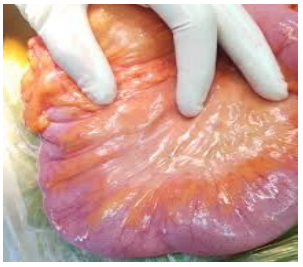
- Colonoscopy with biopsy.
- barium enema, UGI with small bowel follow-through.
- stool cultures.

Macroscopic appearance:

- The disease process is **discontinuous and segmental. (IMPORTANT MCQs!!)**
- In patients with colonic disease, rectal sparing is characteristic of **Crohn's disease** and helps to distinguish it from **ulcerative colitis**.
- Rectal sparing means that the rectum is NOT involved most of the time.
- Perirectal and perianal involvement occurs in about 1/3 of patients with Crohn's disease, particularly those with colonic involvement. Perianal disease (fissure, fistula stricture, or abscess) is common and 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. (**if a patient come with complex perianal fistula, or perianal fistula with weight loss, abdominal pain you suspect IBD until proven otherwise).

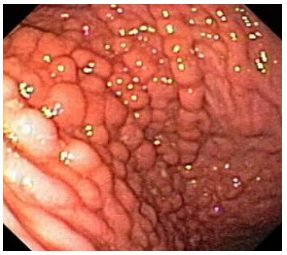
At exploration (means in the OR during Laparotomy):





- Thickened grayish-pink or dull purple-red loops of bowel, thick gray-white exudate or fibrosis of the serosa.
- **Skip areas** of diseased bowel separated by areas of grossly appearing normal bowel.
- Extensive **fat wrapping (IMPORTANT MCQs!!)** " caused by the circumferential growth of the mesenteric fat around the bowel wall. (PIC)
- If you suspect acute appendicitis in a patient and take him to the OR, if the diagnostic laparoscopy shows fat wrapping. what is the most likely diagnosis? **Crohn's.** MCQs!!
- 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 manifestation of Crohn's disease is the development of small mucosal ulcerations called **aphthous ulcers (IMPORTANT)**.
- Aphthous ulcers appear as red spots or focal mucosal depressions.
- As the inflammation progresses, the aphthous ulcers enlarge and become stellate تتجمع بنمط معين.
- 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** (pic).
- Stricturing
- 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.

Microscopic appearance:

- 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. (if you have a patient and during colonoscopy you found segmental disease not continues.what does it mean? Crohn's not UC) MCQs!!
- Well-formed lymphoid aggregates in an edematous fibrotic submucosa is a classic histological feature of the disease.
- Focal ulceration (deep fissuring ulcers).
- **Non-caseating Granuloma** with Langerhans' giant cells **(IMPORTANT)**. 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. Either in the colon or the lymph nodes.

Major presentation:

- The onset of disease is often insidious, with a slow and protracted course (the course will go up and down and increases with time).
- 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 **colicky** abdominal pain, most commonly in the **lower Right abdomen**. The pain may be more severe and localized and may mimic the signs and symptoms of acute appendicitis.
- Watery Diarrhea with mucus is the next most common symptom and is present, at least intermittently (interspersed with asymptomatic periods of varying lengths) 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. non-bloody diarrhea (it's watery) and they don't have **tenesmus**[1]. Remember that we said there will be sparing of the rectum? sparing of the rectum protect them from tenesmus.

Systemic nonspecific symptoms:

- Low grade fever (present in about ⅓)
- Malabsorption and weight loss
- Loss of strength.
- Malaise.

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 (Rare and more common with **UC**).
- Cancer

[1] feeling of painful incomplete defecation.

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

Treatment:

Both medical and surgical treatments are **palliative not curative** (palliative means it's for the symptoms or the complications but not a cure, surgery is curative in UC).

Surgical treatment (Limited to complications)

- Intestinal obstruction
- Intestinal perforation with fistula formation or abscess
- Free perforation
- GI bleeding
- Urologic complications
- Cancer
- **Perianal disease.**

Elective surgery:

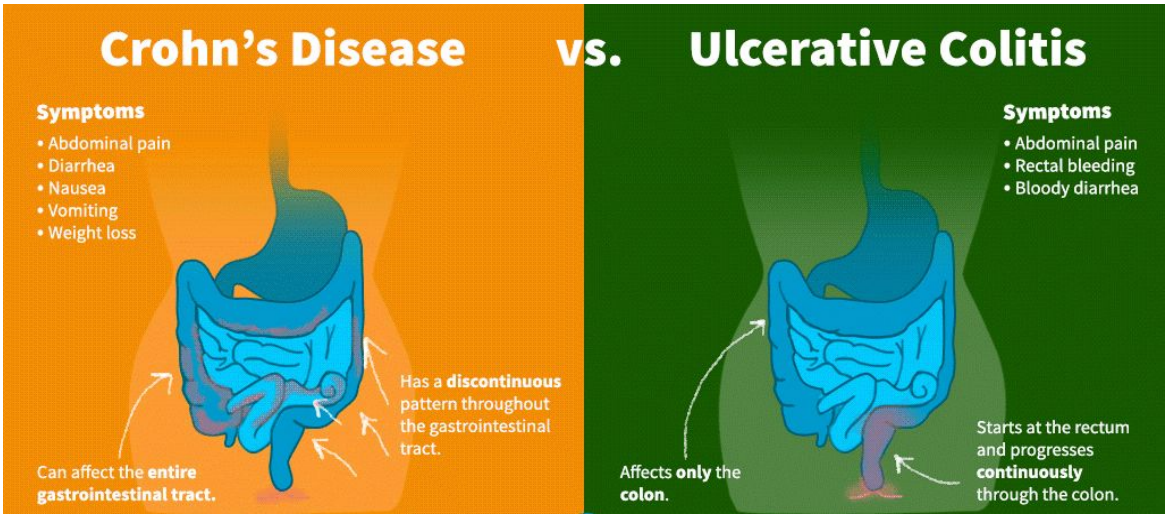
- Chronic subacute obstruction due to fibrotic stricture, adhesions or refractory disease
- Symptomatic disease unresponsive to or poorly controlled by medical management
- Chronic relapsing disease on discontinuation of medical management and steroid dependency
- Complications of medical management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

Emergency:

- Fulminant colitis or acute small bowel relapse unresponsive to medical management
- Acute bowel obstruction
- Life-threatening hemorrhage
- Abscess
- free perforation (uncommon because inflamed segments usually adhere to surrounding structures)
- Perianal abscess

Differential diagnosis:

- 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.



Ulcerative Colitis Helpful Video 6:07

Definition:

- A chronic disease that affects the mucosa and submucosa, with sparing of the muscularis of the rectum and colon (mainly the large intestine).
- Surgery is curative.
- 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 **MCQs!!**. (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).
- In the majority of cases, the disease is contiguous, affecting the rectum and extending proximally. In 5% of cases, it is segmental and the rectum is occasionally spared. “Proctitis is the most common”

Etiology:

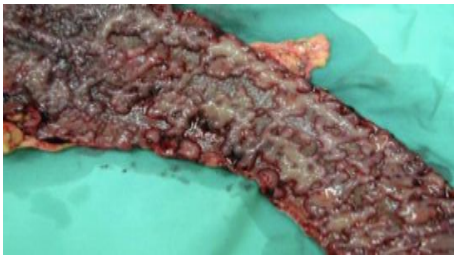
- Unknown but can be:

➔ 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.

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.

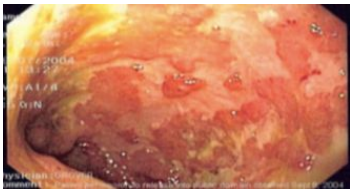


Diagnostic tests:

- Colonoscopy with biopsy.
- barium enema, UGI with small bowel follow-through.
- stool cultures.

Macroscopic appearance:

- The entire colon, including the cecum and appendix, may be involved in ulcerative colitis.
- Despite the disease's name, ulceration of the mucosa isn't always present **(IMPORTANT MCQs!!)**
- The typical gross appearance of ulcerative colitis is **hyperemic mucosa. (most important characteristic of UC, (IMPORTANT MCQs!!)**
- 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.
- **Rectum** is invariably (almost always) involved with the inflammatory process.
  In fact, rectal involvement (proctitis) is the sine qua non (يعني هو الأساس) 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. "Begins distally at rectum then extends proximally to a variable distance".
- Erythematous mucosa, has a granular surface that looks like sandpaper (enlarged ulcer⇒ sandpaper)
- Loss of haustrations. Colon becomes thick and rigid.
- 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. (the polyps here are benign and it is not an indication for surgery).



- 'Backwash ileitis' may produce a dilated and featureless terminal ileum, in which the mucosa appears granular. In contrast to Crohn's disease, ulcerative colitis does not involve
  the terminal ileum except in cases of backwash ileitis (how to differentiate between backwash ileitis and crohn's? do a contrast study in backwash ileitis it's never been fistula and fibrosis).

Microscopic appearance:

The inflammation is restricted to the mucosa and the submucosa of the large bowel. In **severe episodes**, there may be **full-thickness involvement with inflammatory infiltrate**.

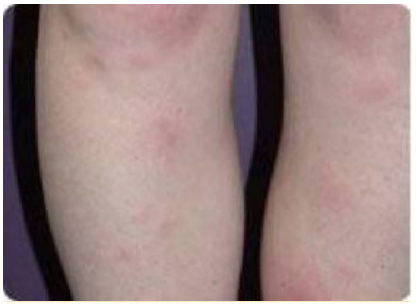
- The most characteristic lesion is crypt abscesses with goblet cell depletion (Collections of neutrophils fill and expand the lumen of individual crypts of Lieberkühn, Not specific for ulcerative colitis and can be seen in Crohn's disease and infectious colitis).

Major presentation:

- Ulcerative colitis and colonic Crohn's disease often have similar clinical presentations. (Both may present with diarrhea and the passage of mucus).
- **Diarrhea with passage of mucus/blood** (4 to more than 10), 1st DDx in bloody diarrhea.
- **Fecal urgency.** 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** (Hematochezia) is also common in ulcerative colitis.
- **Tenesmus** (feeling of painful incomplete defecation)
- 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:

Erythema nodosum



- Arthritis, ankylosing spondylitis[2], **erythema nodosum**, pyoderma gangrenosum, and primary sclerosing cholangitis[3].
- 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 (Ankylosing spondylitis and Primary sclerosing cholangitis).
- Which of the conditions associated with UC not curable by Colectomy? Ankylosing spondylitis and Primary sclerosing cholangitis (may end up with liver transplant) **(EXTREMELY IMPORTANT!!)**

Indications for surgery: (5 months, heartburn, endoscopy, for 13 hours fasting, H pylori)

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

[2] **Ankylosing spondylitis** is a type of arthritis that affects the spine. symptoms include pain and stiffness from the neck down to the lower back.
[3] Chronic liver disease characterized by a progressive course of cholestasis with inflammation and fibrosis of the intrahepatic and extrahepatic bile ducts.

Recall (EXTRA):

What is the cause of IBDs?
No one knows, but probably an autoimmune process with environmental factors contributing.
What is the differential diagnosis?
Crohn's vs Ulcerative colitis, infectious colitis (e.g. C. difficile, amebiasis, shigellosis), ischemic colitis, irritable bowel syndrome, diverticulitis, Zollinger-Ellison syndrome (ZES), colon cancer, carcinoid, ischemic bowel
What are the extra-intestinal manifestations seen in both types of IBDs? "A PIE SACK"

- **A**phthous ulcers
- **P**yoderma gangrenosum
- **I**ritis
- **E**rythema nodosum
- **S**clerosing cholangitis
- **A**rthritis, **A**nkylosing spondylitis
- **C**lubbing
- **K**idney (amyloid deposits, nephrotic syndrome)

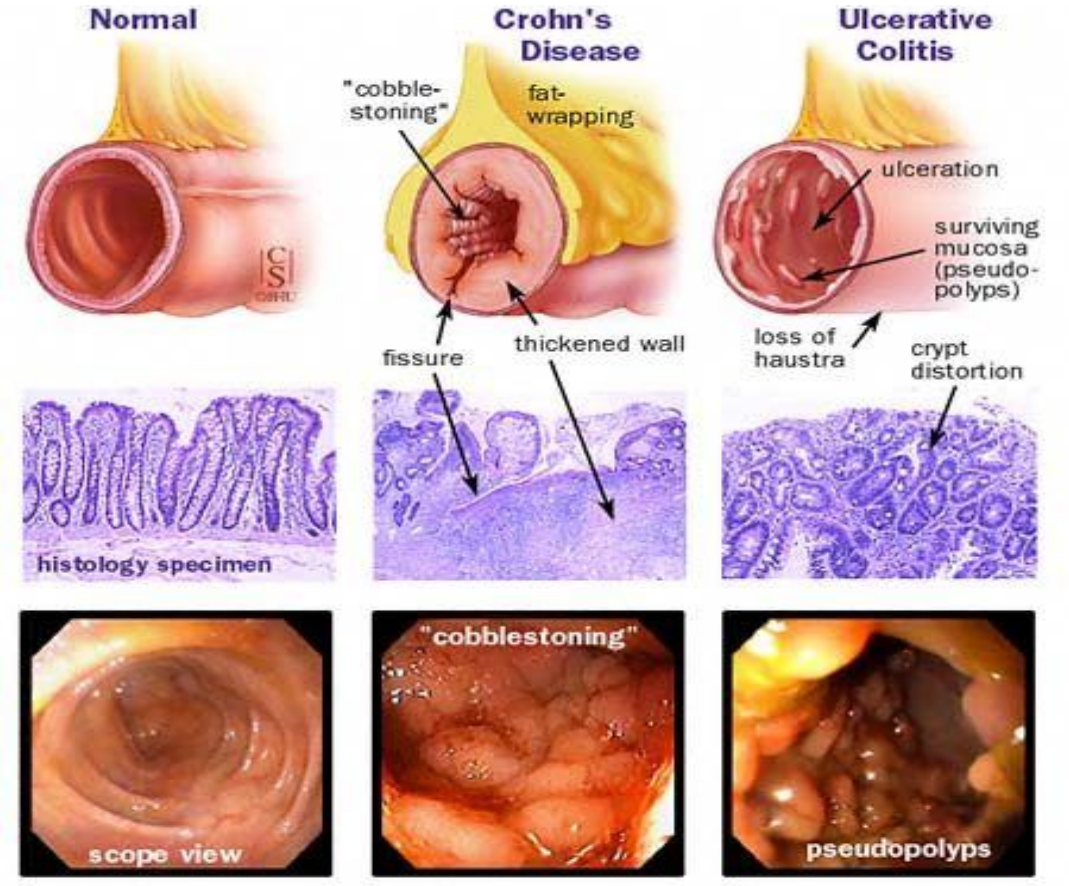
UC at risk population:

- High in jewish population.
- Low in african americans.
- Positive family history

What is toxic megacolon?
Toxic: sepsis febrile, abdominal pain. Megacolon: acutely and massively distended colon
Which disease has cobblestoning more often in endoscopic exam?
Crohn's disease (Think Crohn's = Cobblestoning)
Which disease has pseudopolyps on colonoscopic exam?
Ulcerative colitis; pseudopolyps are polyps of hypertrophic mucosa surrounded by mucosal atrophy
Which disease has a "lead pipe" appearance on barium enema?
Chronic ulcerative colitis
Rectal bleeding/bloody diarrhea is a hallmark of which disease?
Ulcerative colitis (rare in crohn's)
What is the most common indication for surgery in patients with Crohn's disease?
Small bowel obstruction (SBO)
Why do fistulas and abscesses with Crohn's and not UC?
Crohn's is transmural
What is it called when the entire colon is involved?
Pancolitis

Crohn's VS Ulcerative Colitis

	Crohn's disease	Ulcerative colitis
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%)
Anal fissure, fistula, abscess	common	Rare
Rectal sparing	Common (50%)	Rare (5%)
Ulceration	Linear, deep, scattered	Superficial, universal
Distribution	Skip areas	Rectum extending proximally



IMPORTANT

Crohn's disease:

- A **chronic**, **transmural** inflammatory disease of the GI tract of **unknown** cause.
- Macroscopic appearance: **discontinuous and segmental.**
- At exploration: Extensive **fat wrapping .** If you suspect acute appendicitis in a patient and take him to the OR, if the diagnostic laparoscopy shows fat wrapping. what is the most likely diagnosis? **Crohn's.**
- The **earliest** gross manifestation of Crohn's disease is the development of small mucosal ulcerations called **aphthous ulcers.**
- Microscopic appearance : **Non-caseating Granuloma** with Langerhans' giant cells.

Ulcerative Colitis:

- 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.
- Macroscopic appearance:
  - Despite the disease name ulcer there is no ulcer.
  - The typical gross appearance of ulcerative colitis is **hyperemic mucosa.**
- Extraintestinal manifestations:
  - Which of the conditions associated with UC not curable by Colectomy? Ankylosing spondylitis and Primary sclerosing cholangitis (may end up with liver transplant).

Summary

	Crohn's disease	Ulcerative colitis (despite the disease's name, ulceration of the mucosa is not invariably present)
Involvement	• mouth to anus • commonly small intestine and colon • discontinuous and segmental • rectal sparing	• rectum and entire colon, including the cecum and appendix, may be involved • rectum is invariably involved • continuous
Microscopic appearance	• extends transmurally • non-caseating granulomas	• affect the mucosa and submucosa • crypt abscess
Gross appearance	• skip areas • fat wrapping • aphthous ulcers • cobblestone appearance	• hyperemic mucosa
Symptoms	• intermittently : - colicky abdominal pain - diarrhea • stools rarely contain mucus, pus, or Blood. • perianal disease (fissure, fistula, stricture) is **common**. • complications : - obstruction - perforation - fistulas - abscesses - toxic megacolon - cancer	• diarrhea • passage of mucus • urgency (due to distal proctitis) • abdominal discomfort
Surgical indications	(surgery is palliative) • Limited to complications : - intestinal obstruction - intestinal perforation with fistula or abscess - GI bleeding - urological complications - cancer - free perforation - perianal disease	(surgery is curative) • fulminant colitis with toxic megacolon • Massive bleeding • Intractable disease • Dysplasia • malnutrition and growth retardation • May necessitate resection in pediatric And adolescent patients • note : - Ankylosing spondylitis and sclerosing cholangitis are considered as extraintestinal manifestations of UC, And colectomy has no effect on the course of these 2 conditions.

Questions

1-One of the indications for surgical intervention in crohn's disease is:

A- patient's wish
B- when inflammation is restricted to ileum
C- fistula formation
D- development of extra intestinal manifestations

2-Which of the following extra intestinal manifestations persist in ulcerative colitis after total colectomy:

A- Arthritis
B- Uveitis
C- Erythema Nodosum
D- Primary Sclerosing cholangitis

3- Which of the following is a feature of ulcerative colitis:

A- skin lesions
B- crypt abscess
C- transmural inflammation
D- granuloma

4- A 22 y/o male presents to the clinic complaining of abdominal pain, diarrhea and weight loss lasting for one month. He gave a history of occasional occult bleeding in stool. The most likely diagnosis is:

A- Ulcerative colitis
B- Peptic ulcer
C- Incarcerated hernia
D-Intestinal obstruction

5- Transmural inflammation of the colon is seen in:

A- Crohn's Disease
B- Ulcerative Colitis
C -Colon cancer
D- Both A & B

6- A 25-year-old female presents to your clinic complaining of 3 months history of recurrent crampy abdominal pain. Which one of the following points in history is suggestive of crohn's disease:

A- history of being non-smoker
B- history of bloody diarrhea
C- family history of inflammatory bowel disease
D- history of perianal fissure

7- 30 years old female presented with abdominal pain and bloody diarrhea, colonoscopy and biopsy were done. Which one of the following histological features will be suggested diagnosis Ulcerative Colitis?

A -Uniform crypt abscess with goblet depletion
B- Mononuclear cell infiltrate with non-caseating granuloma
C- Mucosal and submucosal thickening with fibrosis and stricture
D- Neuronal hyperplasia with vacuities and aphthous ulcer

8- Which of the following conditions associated with UC is not curable by Colectomy :
A- Ankylosing spondylitis
B- Primary sclerosing cholangitis
C- crohn’s
D- A&B

9- The typical gross appearance of ulcerative colitis is :
A- hyperemic mucosa
B- mucosal inflammation
C- thickness of the mucosa
D-Friable and granular mucosa

Answers:

1:C 2:D 3:B 4:A 5:A 6:C 7:A 8:D 9:A