Clinical History

- Previously healthy 61-year-old man reports 10 poorly-formed bowel movements per day
  - Normally has 3 bowel movements per day
  - Mild abdominal pain
  - No blood in stool
  - No weight loss

Clinical Differential Diagnosis

- Irritable bowel syndrome with diarrhea (IBS-D)
- Infection
- Evolving inflammatory bowel disease
- Microscopic colitis

First Colonoscopy

- Normal colon
- Transition zone at splenic flexure
- Venous congestion with dilated microvasculature
- Edema and lack of vascular markings
**Pathologic Diagnosis**

- Colonic mucosa with features suggestive of chronic injury and ischemia.
- No chronic, active, or microscopic colitis

**New Clinical Differential Diagnosis**

- Irritable bowel syndrome
- 3-4-fold risk of ischemic colitis
- Inflammatory bowel disease
- Infection
- Microscopic colitis
- Medications
  - Not being treated for any other medical problems
  - Other drugs (e.g. cocaine)
  - Patient denied use of recreational drugs or nutritional supplements
- Coagulopathy or vascular abnormality

**Clinical History**

- Onset of change in bowel habits: October 2014
- Upper endoscopy and colonoscopy: May 2015
- Normal CT angiogram coagulopathy work-up: June 2015
- Second colonoscopy: Second Colonoscopy
**Summary of Findings**

- Previously healthy middle-aged man
- Bloody diarrhea and abdominal pain
- Rectosigmoid ulceration and stricture
- Ischemic mucosa with thickened, hyalinized, and thrombosed mucosal blood vessels

**Intestinal Amyloidosis**

- Vascular thickening and dilatation
- Otherwise normal mucosa

**Portal Hypertensive Colopathy**

- Architectural distortion and Paneth cell metaplasia
- No increase inflammation

**Chronic Radiation Colitis**

- Vascular dilatation and thickening
- Hyalinized lamina propria
Biopsy Differential Diagnosis

- Amyloidosis
- Portal hypertensive colopathy
- Chronic radiation colitis
- Something else?

Biopsy Interpretation

- Ischemic colitis with features of chronic venous outflow obstruction
  - In the absence of other causes, idiopathic myointimal hyperplasia of the mesenteric veins should be considered.

Clinical History

- Onset of symptoms
  - October 2014

- Upper endoscopy and colonoscopy
  - April 2015

- Normal CT angiogram coagulopathy work-up
  - May 2015

- Symptom persist
  - June 2015

- Second colonoscopy
  - September 2015

- Rectosigmoid colon resection
- Uneventful postoperative recovery

Rectosigmoid Colon Resection

- Dilated vessels
- Fibrinoid necrosis and thrombosis
Rectosigmoid Colon Resection

Idiopathic Myointimal Hyperplasia of the Mesenteric Veins

Concentrically thickened, partially occluded vein
Normal artery

Artery
Vein

Idiopathic Myointimal Hyperplasia of the Mesenteric Veins

- Segmental ischemia of the rectosigmoid colon in 4 patients

- Transmural ischemic necrosis
- Mural fibrosis
- Proliferation of smooth muscle and proteoglycan matrix in intima of veins
- Normal arteries

Proposed Etiologies

- Occurs in young, otherwise healthy patients
- Male predominance
- Often exercise enthusiasts
- No response to medical management
- Cured by resection

Extreme intravenous pressure
- Cross section of a failed saphenous vein coronary artery bypass graft showing extensive myointimal hyperplasia
- Possible arteriovenous malformations in the sigmoid mesentery


Extreme intravenous pressure
- Cross section of a failed saphenous vein coronary artery bypass graft showing extensive myointimal hyperplasia
- Possible arteriovenous malformations in the sigmoid mesentery

Proposed Etiologies

- Prolonged or recurrent trauma
- Similar changes at sites of previous injury in colonic resection specimens
- Patients are often physically active
- Mobility of the sigmoid colon
- Frequency of sigmoid volvulus
- Predisposition to traumatic arteriovenous fistulization

Biopsy Findings: Key Points

- Idiopathic myointimal hyperplasia of the mesenteric veins should be considered when colonic mucosal biopsy samples show ischemia and abnormally thickened blood vessels
- Diagnosis of exclusion, can only be confirmed in resection specimen
- Prospective suggestion allows for timely resection – only effective treatment

Resection Specimens: Key Points

- Chronic ischemic colitis with thickened veins and normal arteries
- Lack of inflammation in blood vessels
- Cause of distal colon stricture and bleeding that simulates a variety of other strictureing diseases

Differential Diagnosis of Distal Colon Stricture

- Non-inflammatory
  - Chronic ischemia
  - Chronic radiation colitis
- Inflammatory
  - Crohn disease
  - Diverticular disease-associated colitis
- Neoplastic
  - Secondary infiltration by metastatic malignancy

Ischemia

- Strictures in cases of chronic or recurrent ischemia
- Hypovolemia
- Thromboembolic disease
- Mechanical obstruction
- Vasculitis and vasculopathy

Hematoxylin and eosin

Hematoxylin and eosin

*Gut crypts*

**Hemorrhage**

Hyalinized lamina propria

Regenerative crypt with increased mitotic activity
Ischemia

Atheroembolus

Clues may be found in mural or mesenteric blood vessels

Cause may not be identified, even in resection specimens

Vasculitides with Gastrointestinal Involvement

Medium vessel vasculitis

Polyarteritis nodosa

Kawasaki disease

Small vessel vasculitis

Microscopic polyangiitis

Granulomatosis with polyangiitis (Wegener granulomatosis)

Churg-Strauss syndrome

Henoch-Schonlein purpura

Systemic lupus erythematosus

Behcet disease

Enterocolic phlebitis

Vasculitis

General Features

- Biopsies reveal non-specific ischemic change
- Multi-organ involvement
- Serology: pANCA, cANCA
- Gastrointestinal involvement in a minority of cases
- Vascular inflammation outside of ulcerated and/or ischemic areas in resection specimens
- Vascular necrosis and inflammation in an ulcerated area

Patient with mesenteric ischemia following aortic dissection

Vasculitis

Many types have similar features

Vasculitis

Leukocytoclastic vasculitis: Mixed inflammation with vessel wall necrosis

Medial arterial hypeplasia: Fibrous deposits and cellular debris in vessel walls

Vasculitis

Fibrous necrosis of vessel wall

Fibrinoid necrosis of vessel wall

Fibrinoid necrosis of vessel wall

Fibrinoid deposits and cellular debris in vessel walls
**Vasculitis**

- Others have characteristic findings that point to the diagnosis

**Enterocolic Phlebitis**

- Rare idiopathic inflammatory disorder of the mesenteric veins
  - Overlapping features with idiopathic myointimal hyperplasia of the mesenteric veins
  - Segmental ischemia in otherwise healthy patients
  - Exclusively involves veins
  - Mesenteric veins in the “burnt out” stage may show myointimal thickening without inflammation

**Chronic Radiation Colitis**

- Ischemia caused by radiation-induced damage to the mesenteric vessels
  - Onset is months to years after completion of therapy
  - Overlapping features with idiopathic myointimal hyperplasia of the mesenteric veins
  - Often involves rectosigmoid colon
  - Radiation therapy for pelvic malignancies
  - Biopsies may be indistinguishable

**Distinguishing Features**

- Involves the right colon and distal small intestine
- Lymphocytic infiltration of vein walls

**Similarities are also present in resection specimens**

- Mucosal ulceration
- Mural fibrosis
- Transmural ischemic necrosis
- Concentric vascular thickening

**Courtesy S. Hoda, MD**

**Courtesy J. Jessurun, MD**

**Chronic Radiation Colitis**

- Mucoid degeneration
- Foamy macrophages
- Myxoid fibroblasts
Stricturing Disease in the Distal Colon

- Non-inflammatory
  - Ischemia
  - Chronic radiation colitis
- Inflammatory
  - Crohn disease
  - Diverticular disease-associated colitis
- Neoplastic
  - Secondary infiltration by malignancy

Crohn Disease

- Architectural distortion without increased inflammation in areas of quiescent disease
- Mucosal ulceration and adjacent preserved mucosa (pseudopolyps)

Crohn Disease

- Basal lymphoplasmacytosis
- Crypt abscesses and crypt destruction

Crohn Disease

- Transmural inflammation
- Fissures and fistulae

Diverticular Disease-Associated Colitis

- Chronic colitis involving diverticular and peridiverticular colon
- Rare complication of diverticulitis (1-2% of patients)
- Often clinically and histologically indistinguishable from Crohn disease

Diverticular Disease-Associated Colitis

- Chronic active colitis in peridiverticular colon

Chronic colitis in diverticula

Courtesy R. Yantiss, MD
**Diverticular Disease-Associated Colitis**

- Should be considered in the differential diagnosis of elderly patients with Crohn disease of the distal colon
- Cured by resection

**Differential Diagnosis of Distal Colon Stricture**

- Non-inflammatory
  - Chronic ischemia
  - Chronic radiation colitis
- Inflammatory
  - Crohn disease
  - Diverticular disease-associated colitis
- Neoplastic
  - Secondary infiltration by metastatic malignancy

**Neoplastic Infiltration**

- Metastasis or direct extension
- Mural, rather than luminal distribution

**Neoplastic Infiltration**

- Malignant cells expand lamina propria between benign colonic crypts
- Diffuse-type gastric cancer
- Lobular breast carcinoma
- Melanoma

**Neoplastic Infiltration**

- Adenocarcinoma with luminal component
- Large glands with luminal “dirty” necrosis

**Neoplastic Infiltration**

- Serous carcinoma
- Extensive intravascular component
- Papillary architecture, prominent nucleoli, bizarre mitotic figures
Summary and Conclusions

- Idiopathic myointimal hyperplasia simulates a broad range of other, more common colonic disorders
  - Endoscopically
    - Inflammatory bowel disease
  - In biopsy specimens
    - Amyloidosis
    - Portal hypertensive colopathy
    - Chronic radiation colitis
    - Other causes of ischemia
  - In resection specimens
    - Strictures of vascular, inflammatory, and neoplastic etiology

Summary and Conclusions

- Pathologists should be aware of changes produced by idiopathic myointimal hyperplasia in biopsy samples to guide timely resection of the affected segment

- Prospective identification of this disorder requires a high index of suspicion and correlation with clinical history to exclude other possibilities

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