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CASE REPORT | COLON

Ischemic Colitis Due to Idiopathic Myointimal Hyperplasia of the Mesenteric Veins

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ABSTRACT

Idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV) is a rare and poorly understood noninflammatory ischemic colitis. First reported by Genta and Haggitt in 1991, the disease typically presents with chronic abdominal pain, weight loss, and diarrhea with or without hematochezia in middle-aged men. IMHMV is frequently misdiagnosed as an inflammatory bowel disease. The pathophysiology of IMHMV involves the proliferation of the intimal smooth muscle in mesenteric veins leading to bowel ischemia. The etiology of this process remains unknown. There are no good medical therapies for IMHMV, and surgical resection, a curative intervention, is typically required to make the diagnosis. We present the case of a 66-year-old man with IMHMV diagnosed with endoscopic biopsies.

KEYWORDS: ischemic colitis; idiopathic myointimal hyperplasia of the mesenteric veins

CASE REPORT

A previously healthy 66-year-old man presented to a local hospital with sudden-onset left lower quadrant abdominal cramping, fevers, night sweats, and mucoid diarrhea that progressed to hematochezia. He was evaluated in the emergency department and initially diagnosed with diverticulitis based on physical examination and computed tomography (CT) scan findings. He was sent home on a course of oral ciprofloxacin and metronidazole. He returned to his local hospital a week later with progressive symptoms despite compliance with oral antibiotics. He was admitted, treated with intravenous antibiotics for 4 days, and discharged on another course of oral antibiotics. When his symptoms persisted, he presented to our large tertiary care center for further evaluation 2 weeks later. Notably, he had undergone a routine screening colonoscopy 6 years before with reportedly normal findings. There was no family history of gastrointestinal malignancy or inflammatory bowel disease (IBD).

On physical examination, he was afebrile with a heart rate of 92 beats per minute, blood pressure of 127/91 mm Hg, and body mass index of 30.4 kg/m^2, having lost approximately 30 pounds in the 4 weeks he had been ill. Abdominal examination was notable for a soft abdomen with mild tenderness to palpation in the lower quadrants and no organomegaly. There was no lymphadenopathy. Digital rectal examination revealed an empty vault without any lesions or masses. Laboratory investigations showed normal serum and liver chemistries. A complete blood count and iron studies were notable for mild microcytic anemia. Inflammatory markers, C-reactive protein and erythrocyte sedimentation rate, were only mildly elevated at 0.76 mg/dL (ref. ≤0.60 mg/dL) and 71 mm/hr (ref. <20 mm/hr), respectively. Stool analysis revealed no leukocytes or any cultured bacteria and parasites; this included a normal fecal calprotectin of 21 μg/g (ref. normal <16–50 μg/g). *Clostridioides difficile* toxin was not detected. A contrast-enhanced CT scan showed circumferential wall thickening, edema, and adjacent inflammatory changes contiguously involving the rectum through the distal descending colon. Areas of decreased enhancement within portions of the colon were indicative of possible superimposed ischemic changes. Prominent, tortuous vessels were seen scattered within the edematous submucosa and adjacent superior rectal vasculature. The remainder of the colon, appendix, terminal ileum, and rest of the small intestine appeared normal (Figure 1).

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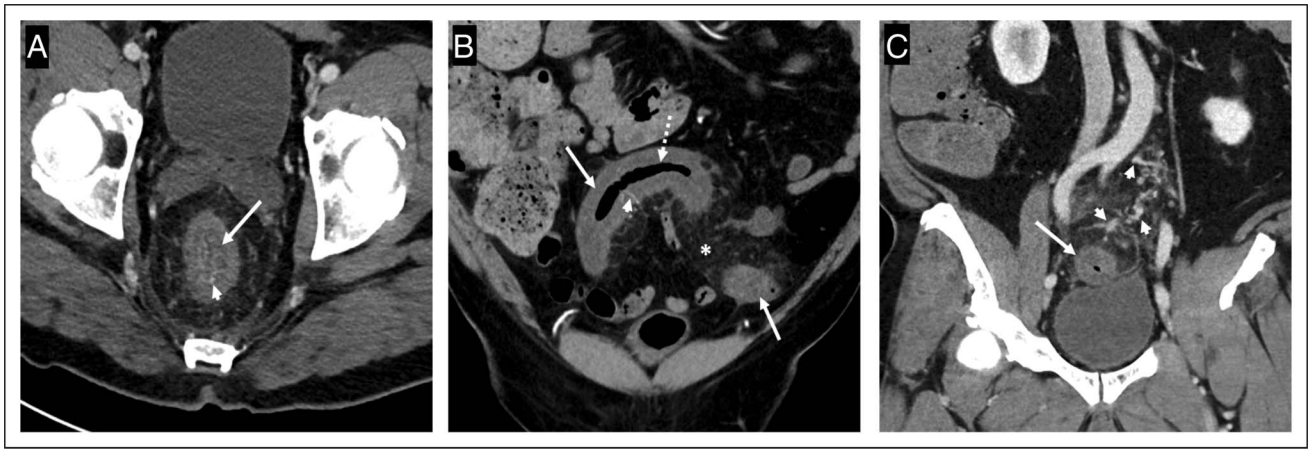

Figure 1. (A) Axial contrast-enhanced CT through the pelvis demonstrates marked inflammation and submucosal edema of the rectum (solid arrow). Prominent submucosal vessels are also seen (arrowhead). (B) Coronal contrast-enhanced CT through the pelvis demonstrates marked, long-segment inflammation and submucosal edema of the sigmoid and distal descending colon (solid arrows). There is lack of a discrete, enhancing mucosa within the inflamed sigmoid colon suggesting possible ischemia/necrosis (dashed arrow). There is inflammation of the adjacent mesenteric fat (*). Prominent submucosal vessels are also seen (arrowhead). (C) Coronal contrast-enhanced CT through the pelvis demonstrates inflammation and submucosal edema of the sigmoid colon (solid arrow). Prominent, tortuous superior rectal vasculature is seen more superiorly in the adjacent mesentery (arrowheads). CT, computed tomography.

The patient was admitted to the general medicine service and started on intravenous antibiotics, and the gastroenterology service was consulted. A flexible sigmoidoscopy was performed revealing diffuse, continuously congested, erythematous, and friable mucosa from the rectum to the mid-descending colon (Figure 2). The proximal descending colon and distal transverse colon appeared normal. No diverticula were appreciated during this examination. Several biopsies using a cold forceps were obtained and submitted for histopathologic examination. While the pathology results were pending, the patient was started on intravenous methylprednisolone given suspicion for IBD and the lack of response to IV and oral antibiotics.

Histopathologic examination of the colon biopsies demonstrated superficial mucosal necrosis and lamina propria fibrosis with dilated thickened wall capillaries containing fibrin thrombi, suggestive of ischemia (Figure 3), and based on these findings, our pathologists raised specific concern for idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV). The patient was continued on IV steroids, and a full vascular and rheumatologic workup was completed and ruled out other etiologies of ischemic colitis. The surgical service was consulted to consider proctosigmoidectomy given the suspicion of IMHMV, but this was deferred because of the patient's clinical improvement. He was subsequently transitioned to oral prednisone and discharged only to be readmitted 1 week later with recurrence and worsening of his symptoms. Repeat abdominal CT imaging was unchanged from earlier. Repeat stool studies indicated new colonization with *C. difficile*, with a positive polymerase chain reaction detecting DNA but a negative toxin assay. As a result, he was started on oral vancomycin; however, he did not show any

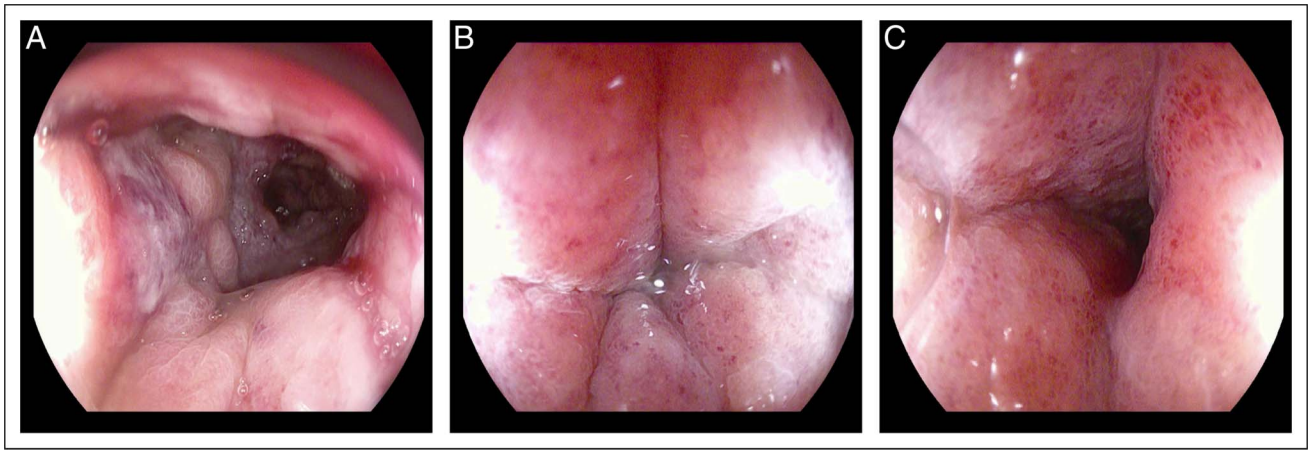

Figure 2. The patient's flexible sigmoidoscopy showed diffuse, continuously congested, erythematous, and friable mucosa from the anal verge to the mid-descending colon. Representative endoscopic images from the (A) rectum, (B) sigmoid colon, and (C) descending colon are shown.

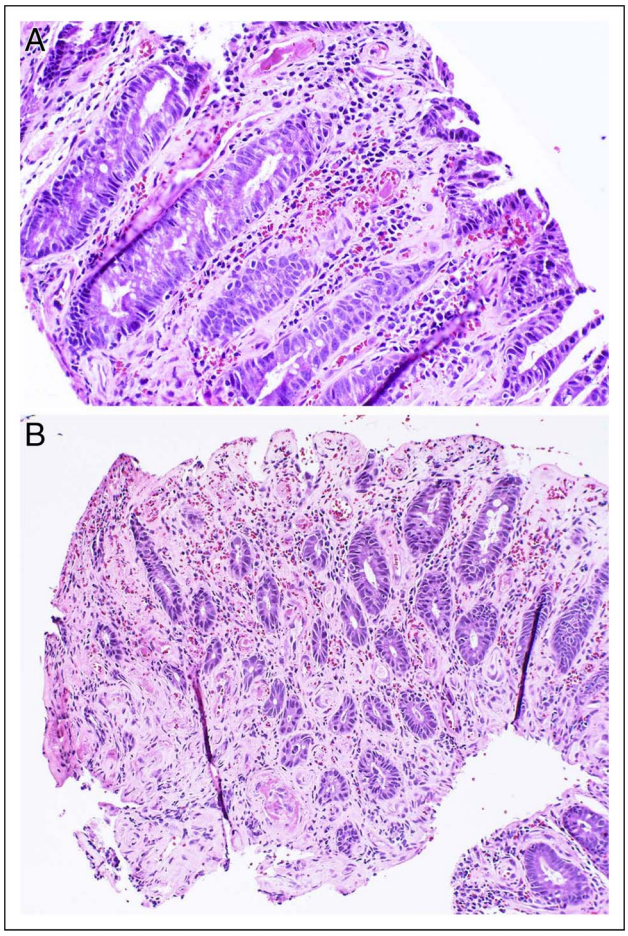

Figure 3. Hematoxylin and eosin-stained sections of the biopsy show characteristic fibrin-rich thrombi and eosinophilic deposits within vein walls (A), as well as superficial mucosal ischemic changes, lamina propria fibrosis, and dilated thick-walled capillaries (A, B) (100× [B], 200× [A]).

clear clinical improvement. Surgery was reconsulted, but colon resection was again deferred given the specter of *C. difficile*.

The patient was seen in surgery clinic 3 weeks later to discuss elective resection but was noted to have an acute abdomen, so he was admitted for urgent proctosigmoidectomy with end colostomy. Unfortunately, his postoperative course was complicated by small bowel obstruction leading to aspiration and cardiopulmonary arrest. The patient ultimately died on postoperative day 4.

Posthumous evaluation of surgical and autopsy specimens confirmed the diagnosis of IMHMV (Figure 4). Interestingly, similar findings of intimal hyperplasia were also present in the small-to-medium-sized mesenteric veins and periprostatic vessels on autopsy. The etiology of his small intestinal obstruction was determined to be because of adhesion to the rectal stump 30 cm from the ileocecal valve and not because of IMHMV.

DISCUSSION

Ischemic colitis due to IMHMV is exceedingly rare, with only a few dozen cases reported in the existing literature. At the time of our patient's diagnosis, there were only 38 known cases of IMHMV published in the literature. IMHMV differs from other ischemic colitides in that it is not due to venous thromboembolism, arterial thrombus, vasculitis, or watershed ischemia from hypotension. Although it was first described in 1991 by Genta and Haggitt,[1] the etiology of this nonthrombotic occlusion of the mesenteric veins remains unclear.

Patients diagnosed with IMHMV are typically otherwise healthy middle-aged men.[2] IMHMV is often mistaken for IBD owing to patients presenting with chronic abdominal pain, diarrhea with or without blood, and imaging/endoscopic findings of colitis.[3,4] Endoscopic biopsy specimens usually show ischemic colitis without a definite etiology. In all published cases of IMHMV, patients showed no response to immunosuppression or antibiotics and are ultimately managed with either segmental or total colectomy, on average, 6 months after initial presentation.[5,6] In the case of our patient, the improvement initially seen with steroids was subsequently believed to be subjective and likely due to the symptom-masking effects of glucocorticoids. The diagnosis of IMHMV is most typically made postoperatively by evaluation of gross surgical specimens.[5–7] Our case is unique in its acute presentation and rapid progression. To our knowledge, this is the only reported case of IMHMV to have such an abrupt and brisk course.

The etiology of IMHMV remains unknown, but is believed to be due to secondary arterialization of mesenteric veins because of increased intravascular pressure, perhaps venous hypertension related to prior trauma or intermittent sigmoid volvulus.[3,8,9] While the rectosigmoid colon is most commonly affected, IMHMV has, in rare cases, involved the transverse colon and even the small bowel.[10–17] Severe IMHMV can lead to potentially life-threatening complications such as bowel obstruction and perforation.[1,18–21]

In this case, we highlight the possibility of diagnosing IMHMV early using mucosal biopsies, which, to our knowledge, is only the second time the diagnosis has been suspected based on histopathologic examination of endoscopic biopsies rather than a gross surgical specimen.[7] A recent study showed 3 specific small vessel histopathologic findings from mucosal biopsies and resections that help distinguish IMHMV from other ischemic or vasculitic colitides.[22] These changes are frequently and specifically seen in ischemic colitis due to IMHMV and consist of subendothelial fibrin deposits, arterialized capillaries, and perivascular hyalinization.[22] Ultimately, IMHMV should be considered on the differential diagnosis in previously healthy patients presenting with acute, noninfectious bloody diarrhea and imaging/endoscopic findings suggestive of ischemic colitis. In cases like this, we advise histology review by an expert gastroenterology pathologist

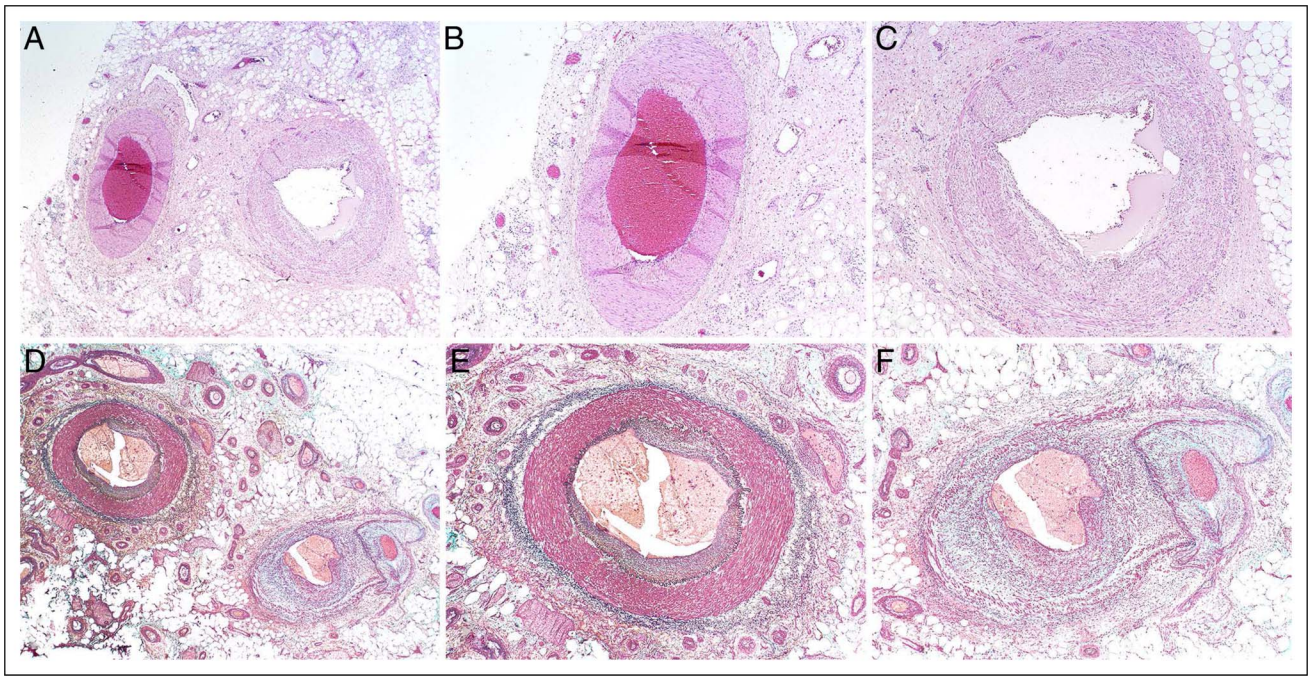

Figure 4. Hematoxylin and eosin-stained sections from the surgical resection specimen demonstrate intimal hyperplasia of mesenteric veins with relative sparing of mesenteric arteries (A–C). Movat pentachrome stain highlights marked intimal hyperplasia and arterialization of veins with concentric proliferation of the medial smooth muscle. The vein wall demonstrates lack of an internal elastic lamina compared with the normal paired artery (D–F). Movat pentachrome stain (5 colors): nuclei stain black, collagen (established) stains yellow, ground substance/mucin/subacute fibroblastic tissue stains blue to green, muscle stains red, and fibrinoid stains intense red (20× [A, D], 40× [B, C, E, F]).

because recognition of pathognomonic mucosal biopsy findings in IMHMV can lead to an earlier, preoperative diagnosis, which can prompt earlier surgical intervention, thus improving patient outcomes.

DISCLOSURES

Author contributions: MG Noujaim and D. Wild were the consulting gastroenterologists who diagnosed the patient. MG Noujaim and H. Tang wrote and revised the manuscript. K. Kalisz read the initial radiographic studies and provided the radiology images and commentary for the manuscript as well as assisted in writing the manuscript. N. Iranzad read the initial pathology slides and provided pathology images and commentary as well as assisted in the writing of the manuscript. D. Wild reviewed the manuscript and assisted in the writing of the final draft. MG Noujaim is the article guarantor.

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