Capturing the often-elusive diagnosis of idiopathic myointimal hyperplasia of mesenteric veins

Ga-ram Han, MD,a Anchit P. Mehrotra, MD,b Adam J. Gomez, MD,c Eric Romanucci, MD,d and Vivienne J. Halpern, MD,e Phoenix, AZ

ABSTRACT

Only 50 cases of idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV) have been reported since 1991 when it was first described. This rare etiology for chronic colonic ischemia is often debilitating to the patient’s quality of life, and no effective medical treatment is available. IMHMV is frequently confused with inflammatory bowel disease because the most common presenting symptoms include abdominal pain, diarrhea, and hematochezia. Surgical resection is curative; however, the diagnosis is rarely reached preoperatively. In the present report, we have described the seventh patient with a diagnosis of IMHMV before surgery and included a literature review to help clinicians recognize this condition. (J Vasc Surg Cases Innov Tech 2022:8:520-33.)

Keywords: Chronic colonic ischemia; Hyperplasia of mesenteric vein; Ischemic colitis; Mesenteric vascular disease; Myointimal hyperplasia

Idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV) is a rare cause of chronic colonic ischemia that is frequently mistaken for inflammatory bowel disease (IBD). It is characterized by smooth muscle hypertrophy in the mesenteric veins causing a nonthrombotic, noninflammatory occlusion leading to venous ischemia. In the present report, we have described the seventh patient to be diagnosed preoperatively with IMHMV. The patient provided written informed consent for the report of his case details and imaging studies.

CASE REPORT

A 74-year-old man had presented to the clinic with a 1-year history of diarrhea, cramping, and weight loss. The initial workup was unrevealing. The C-reactive protein level was high at 9 mg/L and calprotectin was high at 198 μg/g. Endoscopy showed erythematous, edematous, friable mucosa with superfi-

From the Department of General Surgery, Mayo Clinic Arizona,a the Department of General Surgery, Banner University Medical Center-Phoenix, The University of Arizona;b and the Department of Pathology and Laboratory Medicine Service,c Department of General and Colorectal Surgery,d and Department of Vascular Surgery,e Carl T. Hayden Veterans Affairs Medical Center.

Author conflict of interest: none.

Correspondence: Vivienne J. Halpern, MD, Department of Vascular Surgery, Carl T. Hayden Veterans Affairs Medical Center, Mail Stop 112, Phoenix, AZ 85032 (e-mail: Vivienne.halpern@va.gov).

The editors and reviewers of this article have no relevant financial relationships to disclose per the Journal policy that requires reviewers to decline review of any manuscript for which they may have a conflict of interest.

2468-4287
© 2022 Published by Elsevier Inc. on behalf of Society for Vascular Surgery. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).
https://doi.org/10.1016/j.jvscit.2022.05.014

colitis, scattered withered crypts, and increased muscularized mucosal capillaries in the descending colon lamina propria, with milder findings distal to the rectum (Fig 1). Given the unusual distribution and findings of chronic ischemic injury, the results were thought to be suggestive of IMHMV. However, a full workup was recommended to rule out the more common etiologies. Computed tomography (CT) angiography with enterography demonstrated thickening of the vascular walls and inflammatory changes from the mid-transverse to sigmoid colon with prominent collateral vessels, suggestive of an acute on chronic vascular process (Fig 2). The inferior mesenteric artery branches and venous tributaries were smaller than expected but patent, and the small bowel appeared normal.

The patient’s diarrhea and cramping worsened. He had lost 45 lb and began experiencing intermittent hematochezia. Repeat endoscopy showed severe, circumferential ulceration in the sigmoid colon with mottled, edematous mucosa and prominent, increased superficial capillaries (Fig 3). Relative rectal sparing was present, and the appearance was consistent with ischemic colitis. No evidence was found of lymphocytic or collagenous colitis. He was admitted postoperatively. Duplex ultrasound confirmed patent mesenteric vessels, and hypercoagulability workup findings were negative. Vasculitis, hepatitis, and human immunodeficiency virus were ruled out. The case was discussed in the multidisciplinary conference, and surgical resection was recommended.

The patient underwent hand-assisted laparoscopic proctocolectomy with permanent end ileostomy. The mesocolon was shortened and firm, and the bowel wall from the splenic flexure distally was thickened. The descending colon had focal hemorrhage and an 8-cm stricture. The associated vessels were patent without thrombus. Numerous medium-size mesenteric veins were present with narrowing or occlusion by circumferential myointimal proliferation and arterialized capillaries, without inflammatory cell infiltration in vessel walls, consistent with IMHMV (Fig 4). Inflammatory pseudopolyps and moderate gland architectural distortion were present. The patient was...
discharged home on postoperative day 4. At 5.5 months, he continued to do well without recurrent symptoms.

DISCUSSION

IMHMV is a nonthrombotic, noninflammatory condition causing venous wall thickening with luminal narrowing due to smooth muscle hyperplasia. It is a rare and likely underdiagnosed cause of chronic bowel ischemia with only 50 patients with IMHMV identified in the English-language literature. We excluded a case in which the pathognomonic myointimal hyperplasia of the mesenteric veins was not mentioned and a report that had found this change in one venule.\textsuperscript{1,2} Prior studies have shown that focal myointimal hyperplasia of the mesenteric veins can be found after preoperative trauma, in contrast to the diffuse distribution found with IMHMV.\textsuperscript{3}

The mean age of the IMHMV patients was 58 years, and 80% were men and 20% women (Tables I and II).\textsuperscript{4-14} The descending colon was involved in 36%, sigmoid in 79%, and rectum in 55%. In addition, 94% presented with abdominal pain and 67% had experienced hematochezia; 71% had diarrhea alone, 10% both diarrhea and constipation, and 8% constipation alone. Unintentional weight loss was noted in 25%. Some patients had experienced tenesmus, mucus discharge, or incontinence. Leukocytosis and elevated inflammatory markers could also be present.

Cross-sectional imaging will reveal bowel thickening with fat stranding, usually interpreted as infectious or
inflammatory colitis. CT angiography will show patent arteries without evidence of vasculitis and might show increased collateral vessels. Some investigators have reported distended and tortuous pericolonic vessels. One study reported a thin proximal inferior mesenteric vein without distal visualization, and subsequent angiography showed distal inferior mesenteric vein occlusion with ectatic veins.15

Colonoscopy will demonstrate edematous, erythematous, friable and ulcerated walls, usually in a continuous distribution.16 Strictures and pseudopolyps can be present.17,18 The findings have frequently been mistaken for IBD, despite biopsy results inconsistent with this diagnosis. Histopathologic examination will usually show ischemic changes, congestion, and regenerative mucosal changes, with increased muscularized mucosal capillaries (so-called arterialization of capillaries) in the lamina propria.19 Myointimal hyperplasia of the mesenteric veins will not be seen on mucosal biopsy, given the extramural location of these vessels.

Of the 58 patients with IMHMVV, 59% had been misdiagnosed with IBD and 42% with infectious colitis and had undergone treatment with antibiotics, mesalamine, steroids, and/or biologic agents. The natural history of IMHMV is symptomatic progression, sometimes resulting in complications such as bowel obstruction, perforation, hematochezia requiring blood transfusions, or toxic megacolon.20-23

IMHMV can be definitively diagnosed on histopathologic review of the surgical specimen. The demonstration of myointimal hyperplasia of the mesenteric veins can only be appreciated from the resection specimen, and the use of an elastin stain might be required to distinguish these vessels from mesenteric arteries.24 The veins can be larger than their corresponding arteries, which will be spared.25 Mucosal ulceration and signs of ischemic injury with crypt distortion, regenerative changes, and “arterialization” of capillaries will be present.26 Hyalinization of the lamina propria and occasional hyaline thrombi can be found.27,28 Inflammatory cells can be present in the bowel wall but will be absent from the vessel wall, differentiating IMHMV from mesenteric inflammatory veno-occlusive disease.19

The underlying pathophysiology is poorly understood. One theory is that IMHMV is the result of chronic trauma resulting in a segmental acquired arteriovenous fistula. Intermittent colonic volvulus can cause chronic venous obstruction, resulting in increased tortuosity and dilatation of the submucosal veins. This could lead to mucosal venous and capillary ectasia, causing precapillary sphincter incompetence with occult arteriovenous fistulas and, ultimately, myointimal hyperplasia of the mesenteric veins.25 Although arteriovenous fistulas have never been found in the postoperative specimens, this theory is based on the similar appearance of the veins in IMHMV to that of veins subjected to arterial pressure.25 Another theory proposes that the mechanical stress from intermittent volvulus stimulates myointimal hyperplasia with transmission of the elevated venous pressures to the mucosal capillaries, resulting in “arterialization” and endothelial injury, with extravasation of fibrin and red blood cells into the vessel wall.26

A presumptive diagnosis of IMHMV was achieved by endoscopic biopsy for six patients and by imaging studies for one.15,16,29-32 The overall clinical course with worsening symptoms laid the groundwork through which this unusual diagnosis could be considered. Mucosal ischemia, atrophic crypts, and thickened lamina
| Age, years; gender | Presentation | Imaging findings | Endoscopic findings (histopathologic findings from endoscopic biopsies) | Affected bowel |
|--------------------|--------------|-----------------|---------------------------------------------------------------------|----------------|
| 58; M              | Pain, diarrhea, hematochezia | CTA: patent mesenteric vessels; CT: colitis with submucosal edema, pneumatosis intestinalis | Colitis: congested, friable mucosa (enterohemorrhagic colitis or pseudomembranous colitis with features of ischemia) | L colon to rectum |
| 58; M              | Pain, diarrhea, hematochezia | NR               | Mucosal granularity, edema, deep ulcers (vessels with thick, hyalinized walls, prominent endothelial lining, architectural distortion, exudate) | Sigmoid |
| 22-75; 6 M, 2 F   | Pain (n = 7), diarrhea (n = 5), hematochezia (n = 4) | CT: segmental colonic thickening or edema (n = 8); CTA, MRA, or Doppler US: patent mesenteric arteries (n = 8) | Erythema, edema, friability (ischemic changes; dilated, thick-walled, tortuous mucosal capillaries; myointimal hyperplasia of submucosal veins) | Sigmoid (n = 6) |
| 63; M              | Pain, diarrhea, elevated inflammatory markers | CT: colitis with serosal irregularity with mesocolic vascular congestion, hyperemia | Edema, erythema (nonspecific severe colitis) | L colon to sigmoid |
| 60; M              | Pain, diarrhea, hematochezia, weight loss | Angiography: patent IMA; no opacification of IMV; no definite AVF | Serpiginous circular ulcers: edematous, friable mucosa with mucoid discharge (thick-walled, medium-size blood vessels with mural hyalinization, focal thrombosis) | RS |
| 54; M              | Pain, diarrhea, weight loss | NR               | Ulcers, inflammation (CMV+; CMV – on repeat biopsy) | Transverse |
| 47; M              | Pain, diarrhea, proctalgia, malaise, elevated inflammatory markers | CT: RS thickening; dense pericolic fat with small ganglion formations, mild vascular ectasia | Rectal edema, granularity, circumferential, continuous, necrotic ulcers with nodular mucosa, sigmoid stenosis (mucosal edema; hemorrhage, fibrinoid necrosis, thrombosis of small vessels consistent with ischemia) | RS |
| 75; F              | Pain, diarrhea, hematochezia, tenesmus, weight loss, palpable mass | CT: inflammatory mass; barium enema: apple core lesion | Ischemic injury, inflammation (changes consistent with ischemic colitis) | RS |
| 32; F              | Pain, diarrhea, LLQ palpable mass, elevated inflammatory markers | CT: wall thickening, dense pericolic fat; free fluid; barium SBFT: normal; mesenteric angiography: increased collateral vessels | Bubble-like elevations consistent with pneumatosis intestinalis; fibrin-covered ulcers suggestive of pseudomembranous colitis (changes consistent with ischemic colitis) | RS |
| 30; M              | Pain, hematochezia, obstructive symptoms | Barium enema: sigmoid stricture | NR | Sigmoid |
| 38; M              | Pain, diarrhea, constipation, hematochezia, mucoid stools | AXR, US, CT: normal | Erythema, edema, ulcers (changes consistent with UC) | L colon to rectum |
| 25; M              | Pain, diarrhea, constipation, hematochezia | NR               | (Acute necrotizing inflammation; no signs of IBD) | RS |
| 67; M              | Pain, diarrhea, constipation | Barium enema: sigmoid stricture; CTA: patent mesenteric vessels | Ulcers, features of ischemic colitis (changes consistent with ischemic colitis) | RS |

(Continued on next page)
| Age, years: gender | Presentation | Imaging findings | Endoscopic findings (histopathologic findings from endoscopic biopsies) | Affected bowel |
|-------------------|--------------|-----------------|-------------------------------------------------------------------|---------------|
| 68; M             | Constipation, mucous stools, elevated inflammatory markers | CT: edematous walls, adjacent mesocolon; barium enema: tubular narrowing with thumb printing | Edema: circumferential segmental ulcers with luminal narrowing consistent with chronic venous ischemic disease (eg. IMHMV or MIVOD) | L colon to sigmoid |
| 59; F             | Pain, diarrhea, weight loss | US: wall thickening | NR | Ileum |
| 57; M             | Pain, diarrhea | CT: wall thickening with mesocolic edema; angiography: normal SMA; ileocolic, R colic veins not seen; quick opacification of dilated, tortuous veins around R colon | Edema with stricture (normal) | TI to R colon |
| 38; M             | Pain, constipation, hematochezia, mucoid stools, proctalgia, weight loss | Defecography: normal; CT: severe RS edema; free air, fluid | Moderate proctosigmoiditis; focal mild colitis at ileocecal valve (ischemic colitis with ulceration, suggestive of infectious/ischemic etiology) | RS |
| 62; M             | Pain, hematochezia, weight loss | NR | Nodularity, loss of vascular markings, pseudopolypsis with bridging, luminal narrowing, friability; rectal sparing (minimal inflammation) | NR |
| 59; M             | Pain, diarrhea, constipation | CT: wall thickening, inflammation; CT venography: patent mesenteric veins | Circumferential edema, erythema (lamina propria fibrosis with scattered microthrombi, atrophic crypts consistent with ischemia) | RS |
| 62; M             | Pain, diarrhea, hematochezia, weight loss, elevated inflammatory markers | NR | Inflammatory changes with extensive pseudopolypsis, initially with rectal sparing, subsequently involving rectum with stricture, mild, patchy friable mucosa (mildly active colitis; chronic colitis with patchy mild activity on repeat biopsy) | Entire colon |
| 62; F             | Pain, diarrhea | CT: terminal ileal inflammation | Inflammation, ulceration (inconclusive) | Ileum |
| 63; M             | Diarrhea, weight loss, normal inflammatory markers | CT: wall thickening; CTA: extensive colitis with dilatation of transverse. R colon: no arteriopathy or mesenteric thrombus; engorged vessels | Inflamed, cobblestoned mucosa; rectal sparing but unusual vascular pattern (consistent with ischemia: ulcers, granulation tissue; retrospective: fibrin thrombi, arterialized small vessels, subendothelial fibrin deposits consistent with IMHMV) | L colon to upper rectum |
| 62; M             | Pain, diarrhea, hematochezia, proctalgia, elevated inflammatory markers | CTA: patent mesenteric arteries | Inflammation, congestion (ischemic colitis with fibrinoid microvascular wall necrosis, fibrin thrombi) | Transverse to rectum |
| Age, years; gender | Presentation | Imaging findings | Endoscopic findings (histopathologic findings from endoscopic biopsies) | Affected bowel |
|-------------------|--------------|-----------------|---------------------------------------------------------------------|---------------|
| 65; M | Pain, tenesmus | CT: wall thickening with pericolic inflammation | Inflammation with mucosal cobblestoning, erythema, ulcers; congested lamina propria; stricture (dilated mucosal capillaries without active colitis; repeat biopsy showed ischemic injury consistent with IMHMV) | RS |
| 76; M | Pain, diarrhea, weight loss, elevated inflammatory markers | CT: colonic edema, fat stranding | Circumferential sigmoid edema, narrowing, deep longitudinal ulcers (features consistent with ischemic colitis) | RS |
| 22; M | Pain, diarrhea, tenesmus | NR | Inflamed, nodular, friable mucosa; whitish exudate (inflammation consistent with UC; fibrosis of lamina propria, arteriolar sclerosis, fibrin thrombi consistent with ischemia) | RS |
| 25; F | Pain, diarrhea, hematochezia, tenesmus, weight loss | CTA: wall thickening, fat stranding | Features consistent with UC (mucosal edema, rectal aphthous ulcers consistent with ischemia; retrospective: vascular changes consistent with venous obstruction; eg, IMHMV) | L colon to rectum |
| 59; M | Pain, diarrhea, constipation, bloating | CT: edema, mucosal thickening with fat stranding; MRE: minimally active inflammation | Patchy mild inflammation with adhesions, strictures (no active inflammation or dysplasia) | Ileum to RS |
| 62; M | Pain, diarrhea, hematochezia, tender palpable LLQ mass | CT: wall thickening | Circumferential ulcers | RS |
| 62; F | Pain, diarrhea, hematochezia | Angiography: patent mesenteric vessels with minimal irregularities of distal IMA branches; no vasculitis | Continuous mucosal edema, erythema, friability, ulcers (cryptitis, capillary thrombi, glandular dropout, fibrin deposits consistent with ischemia; repeat biopsy: small vessel myointimal thickening in lamina propria consistent with IMHMV) | L colon to rectum |
| 53; M | Pain, bloody mucus per rectum, tenesmus, weight loss, elevated inflammatory markers | CT: pericolic edema with patent, engorged vasculature; CTA: serpiginous, small venous structures in RS with absence of centrally draining IMV | Congested, friable mucosa; stenosis (superficial hemorrhagic necrosis of mucosa; architecturally preserved but atrophic appearing crypts; thickened lamina propria vessels containing thrombi consistent with IMHMV) | Splenic flexure to rectum |
| 81; F | Pain, emesis, elevated inflammatory markers | CT: TI stenosis, wall thickening causing obstruction (retrospective: dilated, tortuous ileocecal veins) | Benign | TI |

(Continued on next page)
propria vessels with capillary fibrin thrombi are suggestive of IMHMV. In the radiographically detected case, angiography had demonstrated patent inferior mesenteric artery and distal inferior mesenteric vein occlusion with venous ectasias. A suggested algorithm for the workup of these patients is presented in Fig 5.

Table I. Continued.

| Age, years: gender | Presentation | Imaging findings | Endoscopic findings (histopathological findings from endoscopic biopsies) | Affected bowel |
|--------------------|--------------|------------------|-----------------------------------------------------------------------|----------------|
| 71; M              | Pain, diarrhea, hematochezia | NR               | All had mucosal erythema; friability; 8 had ulcers, 6 had strictures. 1 had pseudomembranes (with positive C. difficile toxin assay); 1 had prominent mucosal veins, tortuous, dilated submucosal veins proximal to colitis (numerous "arteriolized" capillaries in mucosa, many with signs of endothelial injury; 6 had mucosa showing ischemic colitis with capillaries containing subendothelial fibrinoid deposits, swollen endothelial cells, apopotic nuclear debris in vascular walls, some causing occlusion; 3 had extensive hyalinization of lamina propria; 7 had mild crypt architectural distortion with dilated, shortened or branched crypts, clustered, thin-walled capillaries; patient with C. difficile infection had pseudomembranes, neutrophilic cryptitis) | L colon to rectum |
| 83; M              | Pain, diarrhea, hematochezia, palpable LLQ mass | NR               | L colon to rectum |
| 63; M              | Pain, diarrhea, hematochezia, weight loss | NR               | L colon to rectum |
| 78; M              | Pain, diarrhea, hematochezia | NR               | L colon to rectum |
| 73; F              | Pain, diarrhea, hematochezia | NR               | L colon |
| 65; M              | Pain, diarrhea, hematochezia | NR               | L colon to rectum |
| 64; M              | Pain, diarrhea, hematochezia | NR               | L colon to rectum |
| 25; M              | Pain, diarrhea | NR               | Sigmoid |
| 71; M              | Pain, diarrhea, hematochezia | NR               | L colon to rectum |
| 83; M              | Pain, diarrhea, hematochezia | NR               | NR |
| 64; M              | Pain, diarrhea, hematochezia, elevated inflammatory markers | CT: aneurysm-like lesion near L colon; thick, poorly enhancing walls, fat stranding (retrospective: distal IMV not seen; thin, cord-like proximal IMV without thrombi or luminal irregularities); angiography: patent IMA but distal occlusion with ectatic veins | Continuous mucosal edema, wall thickening, erythema, shallow ulceration (features consistent with IBD) | Transverse to distal rectum |
| 74; M              | Pain, diarrhea, hematochezia, weight loss, elevated inflammatory markers | CTA with enterography: thick walls, inflammatory changes; prominent collateral vessels Small, patent IMA, IMV branches; duplex US: patent mesenteric vessels | Friable mucosa with erythema, edema, ulcers; increased superficial capillaries; relative rectal sparing (patchy, active colitis; scattered withered crypts; increased muscularized mucosal capillaries in lamina propria) | Splenic flexure to rectum |

AVF, Arteriovenous fistula. AXR, abdominal radiography. C. difficile, Clostridioides difficile. CMV, cytomegalovirus. CT, computed tomography. CTA, computed tomography angiography. F, female. IBD, inflammatory bowel disease. IMA, inferior mesenteric artery. IMV, inferior mesenteric vein. L, left. LLQ, lower left quadrant. M, male. MIVOD, mesenteric inflammatory veno-occlusive disease. MRA, magnetic resonance angiography. NR, not reported. R, right. RS, rectosigmoid. SBFT, small bowel follow through. SMA, superior mesenteric artery. TI, terminal ileum. UC, ulcerative colitis. US, ultrasound. *All reported weight loss was unintentional.
| Age, years, gender | Initial diagnoses (treatment) | Time to OR | Indication for surgery | Surgical procedure | Intraoperative findings, gross review of specimen | Histopathologic examination results of surgical specimen | Outcome | Follow-up, months |
|-------------------|-------------------------------|------------|------------------------|-------------------|-----------------------------------------------|------------------------------------------------|---------|-----------------|
| 58; M             | Infectious or ischemic colitis, IBD (steroids, antibiotics) | NR         | Worsening symptoms     | Hartmann procedure | Inflamed L colon to upper rectum with hosophpe rigidity; mucosal edema with fat necrosis, ulcers | IMHMV | NR | NR             |
| 58; M             | IBD (steroids, 5-ASA)         | >1 year    | Persistent symptoms    | Sigmoid colectomy | Otherwise normal colon                         | Edematous, congested mucosa, submucosa; thick-walled vessels in lamina propria with fibrin thrombi; ulcers with superficial necrosis, fibrinous exudate; IMHMV with luminal stenosis veins more prominent than arteries | NR | NR             |
| 22-75; 6 M, 2 F  | IBD in 3 (steroids, mesalamine, infliximab) | 1-6 months | Persistent symptoms    | NR | NR | Venous intimal hyperplasia with walls as thick or thicker than adjacent arteries, seen in extramural, submucosal veins; thickened mucosal capillaries | NR | NR             |
| 63; M             | Infectious colitis (antibiotics, bowel rest) | 1 month    | Worsening symptoms     | Extended left colectomy with end transverse colostomy. low Hartmann pouch | Signs of ischemia with indurated brown-reddish bowel wall, bulky, hardened mesenteric fat; fibrinous layer at inflamed mucosa | Mucosal inflammation, fibrosis with rarefaction of crypts; proliferation of small vessels in lamina propria, submucosa, pericolic fat; some vessels showed fibromyxoid wall thickening; venous intimal hyperplasia causing stenosis, focal secondary thrombosis | Doing well | 60             |
| 60; M             | IBD (steroids, mesalamine, balsalazide, antibiotics) | 2 months   | NR | Hartmann procedure | Diffuse mucosal ulcers with fibrinopurulent exudate | Intramural, extramural IMHMV with near-total occlusion, focal recanalization, arterial sparing | Doing well | 4             |
| 54; M             | CMV colitis (antiviral agents) | 4 months   | Persistent symptoms    | Partial transverse colectomy | NR | Chronic colitis with IMHMV | Doing well | NR             |
| 47; M             | IBD (steroids, infliximab)    | 9 months   | Persistent symptoms    | Hartmann procedure | Ulcer, 13 cm long | IMHMV with luminal stenosis, arterial sparing | NR | NR             |
| 75; F             | Ischemic colitis, IBD (steroids, 5-ASA, antibiotics) | >6 months  | Persistent symptoms    | Hartmann procedure | NR | Ulcerative chronic ischemic injury; IMHMV without vasculitis or arterial involvement | NR | NR             |
| 32; F             | Primary pneumatosis intestinalis, pseudomembranous colitis (oxygen, antibiotics) | 3 months   | Worsening symptoms     | Hartmann procedure | Well-demarcated firm bowel wall with ulcers, thickened pericolic fat, bluish areas in serosa with bubble, suggestive of vascular etiology | Superficial ulcer with fibrosis, hyalinization of lamina propria, marked proliferation of veins with myointimal hyperplasia in submucosa, muscularis propria, serosa | Doing well | 24             |

(Continued on next page)
| Age, years; gender | Initial diagnoses (treatment) | Time to OR | Indication for surgery | Surgical procedure | Intraoperative findings; gross review of specimen | Histopathologic examination results of surgical specimen | Outcome | Follow-up, months |
|-------------------|--------------------------------|------------|------------------------|--------------------|--------------------------------------------------|----------------------------------------------------|---------|------------------|
| 30; M             | (Scheduled for elective surgery) | 1 month    | Obstruction            | Emergent sigmoid resection | Structure with mural thickening, transmural ulcer, firm, yellowish-white serosal exudate | Features consistent with ischemic colitis with normal arteries, no primary vasculitis; ischemic lesions ranged from superficial mucosal necrosis with regenerative epithelial hyperplasia to transmural necrosis, vascular congestion, RBC extravasation in bowel wall, ulcers, focal fibrosis of lamina propria, muscularis mucosae, muscularis propria; myointimal hyperplasia of small mesenteric veins, their intramural branches, usually circumferential but occasionally eccentric, with some thrombosis or occlusion, only present in abnormal segments at mesentery, muscularis propria, submucosa, localized secondary necrotizing vasculitis, fibrin thrombi | Doing well | 84 |
| 38; M             | IBD (steroids, antispasmodic agents) | 2 months   | Toxic megacolon        | Total colectomy with ileostomy, Hartmann pouch | Indurated mesenteric fat, necrotic, hemorrhagic mucosa with thickened muscular wall, pseudopolyps | Doing well | NR |
| 25; M             | IBD | >6 months | Acute abdomen | Hartmann procedure | Edematous, hemorrhagic, focally necrotic colon with fibrinopurulent exudate, indurated mesocolon | Doing well | 48 |
| 67; M             | IBD (sulfasalazine) | 3 months | Worsening symptoms    | Hartmann procedure | Submucosal thickening, mucosal erythema with granular lesions, fibrotic, focally necrotic mesocolic fat | Doing well | 18 |
| 68; M             | Mesenteric panniculitis (steroids) | NR | Endoscopy consistent with IMHMV or MIVOD, persistent symptoms | Left colectomy with Hartmann procedure | Segmental ulcer with stenosis, contraction of L colon | Thickened vein walls due to intimal hyperplasia in submucosa, subserosa, without inflammatory cell infiltrates | NR | NR |
| 59; F             | NR | 6 months | Obstruction           | Small bowel resection | Appearance similar to Crohn disease; palpation far from ileal stenosis revealed intramural nodules, ileal stricture with thickened walls, nodular areas on bowel wall with ulcer | Well-differentiated neuroendocrine tumors, stenotic area with ischemic mucosal changes (edema, fibrosis, ulcers), IMHMV with near-total occlusion of ~30% mesenteric veins, some recanalization, no inflammatory cells or thrombosis, arterial sparing | Doing well | 3 |
| 57; M             | IBD (unspecified treatment) | >10 months | Persistent symptoms   | Right colectomy | Thick wall, firm mesocolic fat, mucosal edema, congestion | IMHMV with narrowing, arterial sparing, submucosal veins with thickened walls appearing larger than arteries | NR | NR |
| 38; M             | IBD (antibiotics, steroids) | 5 months  | Perforation           | Open Hartmann procedure | Large sigmoid perforation with well-demarcated ulcer causing fecal spillage | Ischemic necrosis, IMHMV with total or subtotal obstruction, recanalization, hemorrhage, arterial sparing | Doing well | 18 |
| Age, years, gender | Initial diagnoses (treatment) | Time to OR | Indication for surgery | Surgical procedure | Intraoperative findings: gross review of specimen | Histopathologic examination results of surgical specimen | Outcome | Follow-up, months |
|-------------------|-----------------------------|------------|-----------------------|-------------------|-----------------------------------------------|------------------------------------------------------|---------|------------------|
| 62; M             | C. difficile, IBD (antibiotics, mesalamine, steroids, infliximab) | >10 months | Persistent symptoms | Laparoscopic total proctocolectomy, ileostomy | NR | Thick, ectatic submucosal mucosal vessels; IMHMV of small, medium veins with occlusion; patchy hyalinization of lamina propria, crypt withering, submucosal fibrosis consistent with chronic ischemia | NR | NR |

| 59; M             | Ischemic colitis (antibiotics) | >1 month | Persistent symptoms | Laparoscopic total proctocolectomy, ileostomy | Distorted, thickened, friable mucosa throughout colon with cobblestoning, pseudopolyps, most severe in L colon | No acute inflammation, patchy hyalinization of lamina propria, crypt atrophy, submucosal fibrosis consistent with chronic ischemia, thickened, ectatic mucosal, submucosal vessels; IMHMV with occlusion of small, medium-size veins | NR | NR |

| 62; M             | IBD, C. difficile (mesalamine, steroids, infliximab, antibiotics) | >10 months | Persistent symptoms | Laparoscopic total proctocolectomy, ileostomy | Erythematous, ulcerated, friable mucosa throughout colon with cobblestoning, pseudopolyps, most severe in L colon | Ulcers, ischemic changes with crypt atrophy, regenerative changes, hemorrhage, capillaries with fibrous wall thickening ('arteriolization'); subendothelial fibrin deposits in small vessels; fibrin thrombi; myxoid change with IMHMV of large veins in mesentery; subserosa causing narrowing, appearing larger than arteries; one vein with recanalization | NR | NR |

| 62; F             | NR | NR | Perforation | Emergent right colectomy | NR | Full-thickness, punched-out ulcer of small bowel; IMHMV with luminal narrowing | NR | NR |

| 63; M             | Ischemic colitis, IBD (antibiotics, steroids) | 5 months | Persistent symptoms | Open Hartmann procedure | Grossly abnormal colon from upper rectum to mid-L colon edematous mesentery adherent, fixed to RP; thickened wall, mesenteric fat with ulcers | Ulcers, ischemic changes with crypt atrophy, regenerative changes, hemorrhage, capillaries with fibrous wall thickening ('arteriolization'); subendothelial fibrin deposits in small vessels; fibrin thrombi; myxoid change with IMHMV of large veins in mesentery; subserosa causing narrowing, appearing larger than arteries; one vein with recanalization | NR | NR |

| 62; M             | (Antibiotics, steroids) | >1 year | Persistent symptoms | Total colectomy, end ileostomy | NR | IMHMV | NR | NR |

| 65; M             | C. difficile, IBD (antibiotics, steroids, mesalamine), suspected IMHMV (perforated before surgery) | 1.5 months | Perforation | Emergent Hartmann procedure | NR | Muscular thickening of intramural veins with arterial sparing | NR | NR |

(Continued on next page)
| Age, years; gender | Initial diagnoses (treatment) | Time to OR | Indication for surgery | Surgical procedure | Intraoperative findings; gross review of specimen | Histopathologic examination results of surgical specimen | Outcome | Follow-up, months |
|-------------------|--------------------------------|------------|------------------------|-------------------|-----------------------------------------------|------------------------------------------------|--------|-----------------|
| 76; M             | Ischemic colitis, infectious colitis, IBD (bowel rest, antibiotics, mesalamine, steroids) | 1 year     | Worsening symptoms     | Sigmoidectomy     | Thickened wall, circumferential 10-cm ulcer | Mucosa with fibrin deposits, active inflammation, congestion consistent with ischemia; ghost-like epithelium; submucosal vascular proliferation with hyaline thrombi; IMHMV with stenosis, mucin-like matrix deposition in intima; venous wall structure resembling arteries; no phlebitis or arteriosclerosis | Doing well | 3               |
| 22; M             | IBD (mesalamine, sulfasalazine, bowel rest, steroids, cyclosporine) | NR         | Persistent symptoms, medication side effects | Open Hartmann procedure | RS transmural inflammation with sealed perforation | Colonic ischemia due to IMHMV | Doing well | 10              |
| 25; F             | IBD, C. difficile (antibiotics, mesalamine, steroids) | NR         | Persistent symptoms, endoscopic biopsy suggestive of IMHMV | NR | NR | NR | NR | NR | NR |
| 59; M             | IBD (unspecifed treatment) | 30 years   | Persistent symptoms    | Open subtotal colectomy with end ileostomy, Hartmann pouch | Dilated colon with indurated mesentery, thickened bowel wall with otherwise unremarkable mucosa; soft submucosal colonic nodules | Muscularis propria hypertrophy; peracute pericolic IMHMV; no mucosal ischemic changes or findings of chronicity or acuity seen; submucosal lipomas | NR | NR |
| 62; M             | Infectious colitis (antibiotics) | 1 month    | Worsening symptoms     | Open sigmoidectomy | NR | IMHMV with mesenteric fibrosis, fat necrosis | NR | NR |
| 62; F             | IBD (steroids, mesalamine), IMHMV suspected from endoscopic biopsy | 2 months   | Endoscopic biopsy consistent with IMHMV, persistent symptoms | Laparoscopic RS resection with low anastomosis, diverting loop ileostomy | RS with bowel wall edema, muscular hypertrophy or thickening with surrounding mesenteric edema | Colonic mucosa with ulcers, granulation tissue, acute inflammation, congestion, hemorrhage, lamina propria fibrosis, IMHMV in mucosa, submucosa, subserosa with occlusion | Doing well | >18 |
| 53; M             | Inflammatory colitis (nortriptyline, antibiotics, steroids) | >3 months  | Endoscopic biopsy consistent with IMHMV, persistent symptoms | Open left colectomy with Hartmann pouch, end colostomy | Colonic, mesenteric inflammation extending to distal rectum, with dense, fibrotic adhesions to RP | IMHMV with luminal narrowing | Doing well | 3               |
| 81; F             | Small bowel obstruction treatment | 1 year     | Failed medical management of obstruction | Laparoscopic small bowel resection | No adhesions, telangiectasia on TI serosa, thick wall, circumferential ulcers with scarring in stenotic segment | Fibrosis with lymphohypertrophic, plasmacytic infiltration in mucosa, lamina propria, subserosa; subserosal veins with thick walls, stenosis or obstruction; venous wall structure resembling arteries; arterial sparing, no phlebitis or phlebosclerosis | Doing well | 32              |
**Table II.** Continued.

| Age, years; gender | Initial diagnoses (treatment) | Time to OR | Indication for surgery | Surgical procedure | Intraoperative findings; gross review of specimen | Histopathologic examination results of surgical specimen | Outcome | Follow-up, months |
|--------------------|-------------------------------|------------|------------------------|--------------------|--------------------------------------------------|------------------------------------------------------|----------|------------------|
| 71; M              | Ischemic colitis, IBD         | NR         | 5/10 due to perforation, other indications, obstruction, refractory colitis | 5/10 had urgent colectomy for perforation | Strictures, ulcers, serositis with thick mesenteric fat, medium, large submucosal mesenteric veins with narrowing due to myointimal hyperplasia, arterial sparing. 10/10 had withered, regenerative microcrypts, architectural distortion, hemorrhage, subendothelial hyaline deposits consistent with ischemia. 9/10 had fibrin thrombi, dilated thick-walled mucosal capillaries with prominent endothelium (arterialization). 1/10 had hyalinized lamina propria. 1 had IMHMV in L colon, pseudomembranous colitis in transverse colon | NR | NR |
| 83; M              | IBD                           | NR         |                        |                    |                                                  | NR | NR |
| 78; M              | IBD                           | NR         |                        |                    |                                                  | NR | NR |
| 73; F              | IBD                           | NR         |                        |                    |                                                  | NR | NR |
| 65; M              | IBD                           | NR         |                        |                    |                                                  | NR | NR |
| 64; M              | IBD                           | NR         |                        |                    |                                                  | NR | NR |
| 25; M              | Ischemic colitis              | NR         |                        |                    |                                                  | NR | NR |
| 71; M              | IBD                           | NR         |                        |                    |                                                  | NR | NR |
| 83; M              | Ischemic colitis              | NR         |                        |                    |                                                  | NR | NR |
| 64; M              | IBD (antibiotics, steroids) suspected IMHMV after angiography and CT | 2 years | Imaging consistent with IMHMV, persistent symptoms | Total proctocolectomy with IPAA, ileostomy | Continuous inflammation in rectum to distal transverse colon | Ulcers, submucosal edema with hemorrhage, chronic serositis, fat necrosis, fibrous intimal thickening, occlusion of medium to large veins, no venulitis or venous thromboarterial sparing | Doing well | 6 |
| 74; M              | IMHMV suspected from CT and endoscopic biopsy findings | 2 years | Endoscopic biopsy consistent with IMHMV, persistent symptoms | Hand-assisted laparoscopic total proctocolectomy, end ileostomy | Shortened, thickened, firm mesentery with thickened walls from splenic flexure to distal margin. L colon had hemorrhage, stenosis | Medium mesenteric veins with total, subtotal occlusion by myointimal proliferation, arterialized capillaries, without inflammatory cell infiltration, inflammatory pseudopolyps in overlying mucosa with gland-architectural distortion | Doing well | 5.5 |

5-ASA, 5-Aminosalicylic acid; C. difficile, *Clostridioides difficile*; CT, computed tomography; CMV, cytomegalovirus; F, female; IBD, inflammatory bowel disease; IPAA, ileal pouch—anal anastomosis; L, left; M, male; MIVOD, mesenteric inflammatory veno-occlusive disease; NR, not reported; OR, operating room; RBC, red blood cell; RP, retroperitoneum; TI, terminal ileum.
CONCLUSIONS

IMHMV is a rare diagnosis; however, the symptoms can be debilitating with life-threatening complications. Patients will often be misdiagnosed with IBD, delaying definitive treatment. Resection will be curative with resolution of symptoms. Careful histopathologic review of endoscopic biopsies in the context of worsening symptoms and suspicious CT findings can facilitate the preoperative diagnosis. We have described the seventh patient to be diagnosed preoperatively and provided a literature review to increase awareness and accelerate the diagnostic process to allow patients to undergo curative resection more expeditiously.

We thank Dr Wendy Lamb for assistance with obtaining the computed tomography images.

REFERENCES

1. Lantis S, Kontovounios C, Karaliotas C. An extremely rare small bowel lesion associated with refractory ascites. Gastroenterology 2012;142:e5-7.
2. Bryant J. Unexpected sudden death during propranolol therapy in a patient with mild mesenteric venous myointimal hyperplasia. J Forensic Sci 1998;43:905-7.
3. Sherman J, Kao PC, West AB, Blaszky H. Focal myointimal hyperplasia of mesenteric veins is associated with previous trauma in surgical specimens. Pathol Res Pract 2006;202:517-22.
4. Anderson B, Smyrk TC, Graham R, Lightner A, Sweetser S. Idiopathic myointimal hyperplasia is a distinct cause of chronic colon ischaemia. Colorectal Dis 2019;21:1073-8.
5. Al Ansari A, Ahmed S, Mansour E, Abass MA. Idiopathic myointimal hyperplasia of mesenteric veins. J Surg Case Rep 2021;2021:rrjaa453.
6. Chiang C-K, Lee C-L, Huang C-S, Huang S-H, Wu C-H. A rare cause of ischemic proctosigmoiditis: idiopathic myointimal hyperplasia of mesenteric veins. Endoscopy 2012;44(Suppl 2)E54-5.
7. Chudy-Onwugaje K, Ali O, Umoren M. Idiopathic myointimal hyperplasia of the mesenteric veins of the colon. Clin Gastroenterol Hepatol 2020;18:A19-20.
8. Costa MN, Saiote J, Pinheiro MJ, Duarte P, Bentes T, Ferraz-Oliveira M, et al. Segmental colitis caused by idiopathic myointimal hyperplasia of mesenteric veins. Rev Esp Enferm Dig 2016;108:821-6.
9. Feo L, Cheeyandira A, Schaffzin DM. Idiopathic myointimal hyperplasia of mesenteric veins in the elderly. Int J Colorectal Dis 2013;28:433-4.
10. De Hertogh C, Van Eyken P, Stessens L, Caenepeel P, Geboes K. Myointimal hyperplasia of mesenteric veins secondary to heterozygous factor V Leiden mutation. Histopathology 2005;47:322-4.
11. Korenblit J, Matro R, Goldstein S, Burkat A, Baill F, Frankel R, et al. Idiopathic myointimal hyperplasia of the mesenteric veins. Am Surg 2014;80:E152-4.
12. Laskaratos F-M, Hamilton M, Novelli M, Shepherd N, Jones G, Lawrence C, et al. A rare cause of abdominal pain, diarrhoea and GI bleeding. Cut 2015;64:214, 350.
13. Thomas BS. Myointimal hyperplasia of the mesenteric veins mimicking infectious colitis. Int J Colorectal Dis 2013;28:727.
14. Song SJ, Shroff SC. Idiopathic myointimal hyperplasia of mesenteric veins of the ileum and colon in a patient with Crohn's disease: a case report and brief review of the literature. Case Rep Pathol 2017;2017:6795031.
15. Yun SJ, Nam DH, Kim J, Ryu JK, Lee SH. The radiologic diagnosis of idiopathic myointimal hyperplasia of mesenteric veins with a novel presentation: case report and literature review. Clin Imaging 2016;40:870-4.
16. Wangensteen KO, Fogt F, Kann BR, Osterman MT. Idiopathic myointimal hyperplasia of the mesenteric veins diagnosed preoperatively. J Clin Gastroenterol 2015;49:491-4.
17. Abbott S, Hewett P, Cooper J, Ruszkiewicz A. Idiopathic myointimal hyperplasia of the mesenteric veins: a rare differential to be considered in idiopathic colitis. ANZ J Surg 2015;85:242-3.
18. Korenblit J, Burkat A, Frankel R, Klinge M, Greenbaum L, Goldstein S, et al. Refractory pancolitis: a novel presentation of idiopathic myointimal hyperplasia of mesenteric veins. Gastroenterol Hepatol (N Y) 2012;8:696-700.
19. Platz J, Hyman N. Idiopathic myointimal hyperplasia of mesenteric veins. Gastroenterol Hepatol (N Y) 2012;8:700-2.
20. Genta RM, Haggitt RC. Idiopathic myointimal hyperplasia of mesenteric veins. Gastroenterology 1991;101:533-9.

Fig 5. Algorithm for workup of a patient with idiopathic myointimal hyperplasia of mesenteric veins (IMHMV).
21. Guadagno E, Del Basso De Caro M, Del Prete E, D’Armento FP, Campione S. Coexistence of multiple ileal neuroendocrine tumors and idiopathic myointimal hyperplasia of mesenteric veins: coincidence or consequence? Case report and review of literature. Int J Surg Pathol 2016;24:627-30.

22. Kao PC, Vecchio JA, Hyman NH, West AB, Blaszyk H. Idiopathic myointimal hyperplasia of mesenteric veins: a rare mimic of idiopathic inflammatory bowel disease. J Clin Gastroenterol 2005;39:704-8.

23. Savoie LM, Abrams AV. Refractory proctosigmoiditis caused by myointimal hyperplasia of mesenteric veins. Dis Colon Rectum 1999;42:1093-6.

24. Yamada K, Hiraki M, Tanaka T, Mori D, Tanaka F, Manabe T, et al. A case of idiopathic myointimal hyperplasia of the mesenteric veins presenting with small bowel obstruction. Surg Case Rep 2021;7:1-5.

25. Abu-Alfa AK, Ayer U, West AB. Mucosal biopsy findings and venous abnormalities in idiopathic myointimal hyperplasia of the mesenteric veins. Am J Surg Pathol 1996;20:1271-8.

26. Martin FC, Yang LS, Fehily SR, D’Souza B, Lim A, McKelvie PA. Idiopathic myointimal hyperplasia of the mesenteric veins: case report and review of the literature. JCH Open 2020;4:345-50.

27. García-Castellanos R, López R, de Vega VM, Ojanguren I, Piñol M, Boix J, et al. Idiopathic myointimal hyperplasia of mesenteric veins and pneumatosis intestinalis: a previously unreported association. J Crohn Colitis 2011;5:239-44.

28. Sahara K, Yamada R, Fujiwara T, Koizumi K, Horiguchi S, Hishima T, et al. Idiopathic myointimal hyperplasia of mesenteric veins: rare case of ischemic colitis mimicking inflammatory bowel disease. Dig Endosc 2015;27:768-71.

29. Patel AD, Schneider Y, Saumoy M, Maltz C, Yeo H, Jessurun J, et al. Idiopathic myointimal hyperplasia of the mesenteric veins. ACG Case Rep J 2016;3:e64.

30. Gonai T, Toya Y, Nakamura S, Kawasaki K, Yanai S, Fujita Y, et al. Gastrointestinal idiopathic myointimal hyperplasia of mesenteric veins. J Gastroenterol Hepatol 2018;33:1939.

31. Kelly Wu W, Tombazzi CR, Howe CF, Kendall MA, Walton DB, Washington MK, et al. Idiopathic myointimal hyperplasia of the mesenteric veins: a rare imitator of inflammatory bowel disease. [e-pub ahead of print]. Am Surg, https://doi.org/10.1177/0003134820973390, accessed August 9, 2022.

32. Snell D, Shah SL, Jessurun J, Maltz C, Wan D. The great IBD imitator: a case of idiopathic myointimal hyperplasia of the mesenteric veins. J Am Coll Gastroenterol 2017;112:S782.

33. Yantiss RK, Cui I, Panarelli NC, Jessurun J. Idiopathic myointimal hyperplasia of mesenteric veins. Am J Surg Pathol 2017;41:1657-65.

Submitted Feb 2, 2022; accepted May 22, 2022.