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

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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 superficial ulceration in the descending colon with milder findings in the rectosigmoid. No signs of IBD were present, and cytomegalovirus testing was negative. Biopsies showed patchy active 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 postprocedurally. 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

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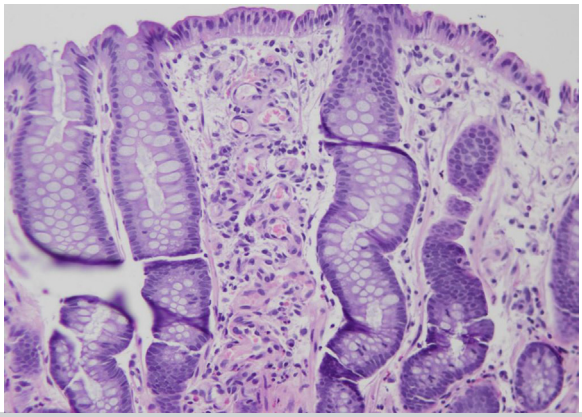

Fig 1. Preoperative biopsy of distal colon, May 2020 (similar findings were seen in the follow-up preoperative biopsy, November 2020), with sections demonstrating colonic mucosa with increased muscularized mucosal capillaries (so-called arterialization of capillaries). Additional biopsy findings (not shown) included ischemic-type changes with patchy active colitis and erosion, sparing the proximal colon. Features suggestive of inflammatory bowel disease were not appreciated. The differential diagnosis included idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV), chronic ischemic injury, and chronic medication injury. Hematology and eosin stain, original magnification ×200.

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.[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.[3]

The mean age of the IMHMV patients was 58 years, and 80% were men and 20% women (Tables I and II).[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

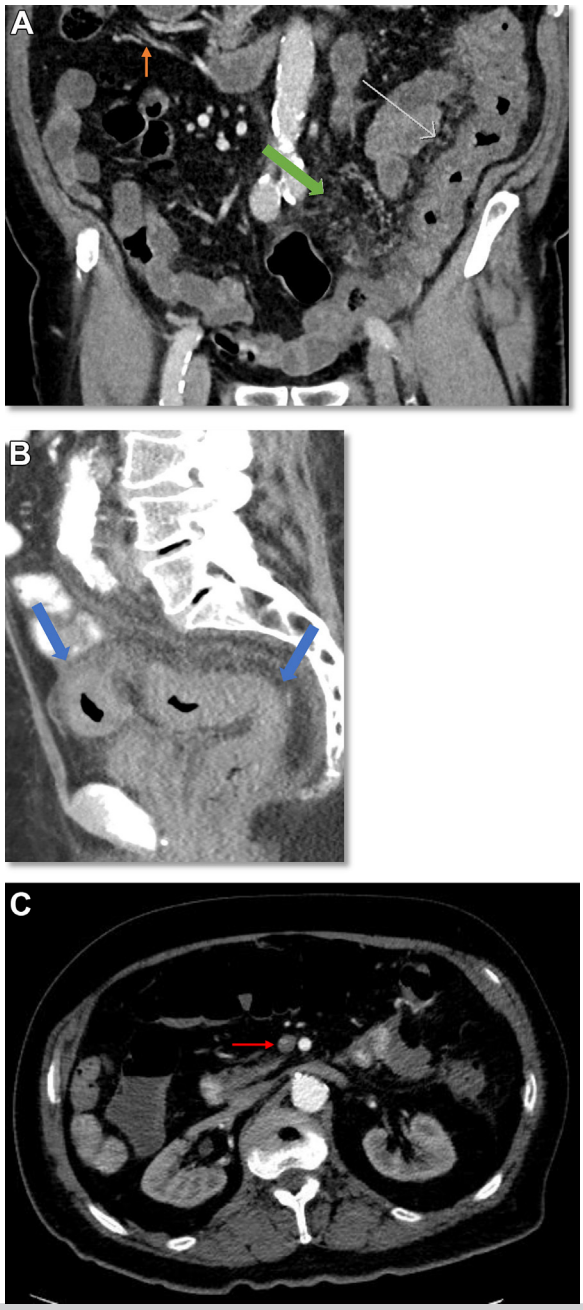

Fig 2. Computed tomography (CT) angiograms of the abdomen and pelvis demonstrating acute on chronic inflammatory changes of the descending colon. **A,** The wall of the descending colon was moderately thickened with pericolonic soft tissue attenuation stranding, consistent with active inflammation (*white arrow*). Arterial branches of the inferior mesenteric artery at the left lower quadrant had an abnormal serpiginous morphology and tortuosity (*green arrow*), in contrast to normal linear morphology arteries at the right mid-abdomen (*orange arrow*), consistent with a chronic process. **B,** Inflammation involving the rectosigmoid colon (*blue arrows*). **C,** Inferior mesenteric vein wall thickening vs thrombus with contrast filling a narrowed lumen (*red arrow*).

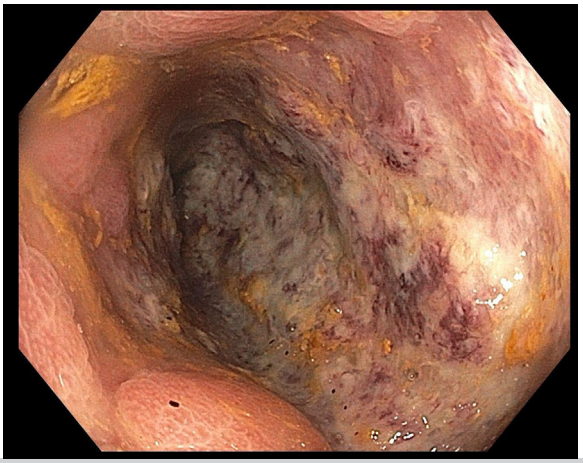

Fig 3. Colonoscopy depicting severe, circumferential ulceration in the sigmoid colon with mottled, edematous mucosa and prominent, increased superficial capillaries.

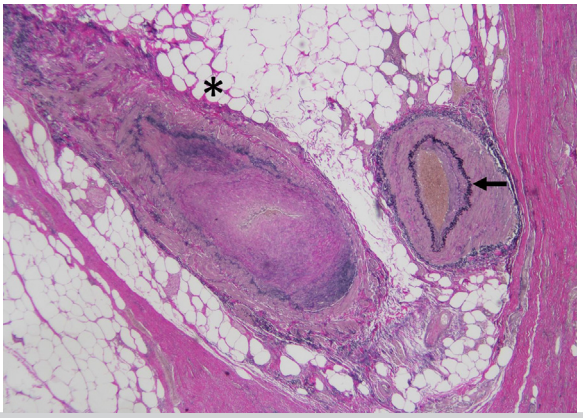

Fig 4. Mesenteric vessels of distal colon at colectomy, March 2021, with sections demonstrating myointimal hyperplasia of the mesenteric vein on the left (*asterisk*) with thickening of the vessel wall and subtotal occlusion of the lumen. The adjacent mesenteric artery to the right shows normal vessel thickness and luminal caliber with intact internal elastic lamina (*arrow*), which is absent in the mesenteric vein. Elastin stain, original magnification ×40.

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

Table I. Summary of all idiopathic myointimal hyperplasia of mesenteric veins (IMHMV) patients in the English-language literature: presentation, workup, and disease distribution[a]

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)

Table I. Continued.

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, pseudopolyposis 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 pseudopolyposis, 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

Table I. Continued.

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

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Table I. Continued.

Age, years; gender	Presentation	Imaging findings	Endoscopic findings (histopathologic 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, apoptotic 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, weight loss	NR		L colon
65; M	Pain, diarrhea, hematochezia	NR		L colon to rectum
64; M	Pain, diarrhea, hematochezia	NR		
25; M	Pain, diarrhea	NR		Sigmoid
71; M	Pain, diarrhea, hematochezia	NR		L colon to rectum
83; M	Pain, diarrhea, hematochezia	NR	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.
[a]All reported weight loss was unintentional.

propria vessels with capillary fibrin thrombi are suggestive of IMHMV.[33] In the radiographically detected case, angiography had demonstrated patent inferior mesenteric artery and distal inferior mesenteric vein occlusion with venous ectasias.[15] A suggested algorithm for the workup of these patients is presented in Fig 5.

Table II. Summary of all idiopathic myointimal hyperplasia of mesenteric veins (IMHMV) patients in the English-language literature: initial diagnoses, prior treatment, operative details, and postoperative course

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 hosepipe 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

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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
30; M	(Scheduled for elective surgery)	1 month	Obstruction	Emergent sigmoid resection	Stricture 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

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
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	1: Laparoscopic converted to open transverse loop colostomy;2: open left colectomy	Distorted, thickened, fibrotic colon with attached firm rubbery yellow-white pericolonic fat consistent with fat necrosis	1: Myointimal hyperplasia with occlusion of small, medium-size veins with ischemic mucosal changes; 2: 95% venous occlusions	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	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; 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)

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
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
59; M	IBD (unspecified 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; peri-ileal, 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 lymphocytic, 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	NR	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"); 7/10 had hyalinized lamina propria; 1 had IMHMV in L colon, pseudomembranous colitis in transverse colon	NR	NR
83; M	IBD	NR			NR		NR	NR
3; M	IBD	NR			NR		NR	NR
78; M	IBD	NR			NR		NR	NR
73; F	IBD	NR			NR		NR	NR
65; M	IBD	NR			NR		NR	NR
64; M	IBD	NR			NR		NR	NR
25; M	Ischemic colitis	NR			NR		NR	NR
71; M	IBD	NR			NR		NR	NR
83; M	Ischemic colitis	NR			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 thrombi; arterial 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, stricture	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.

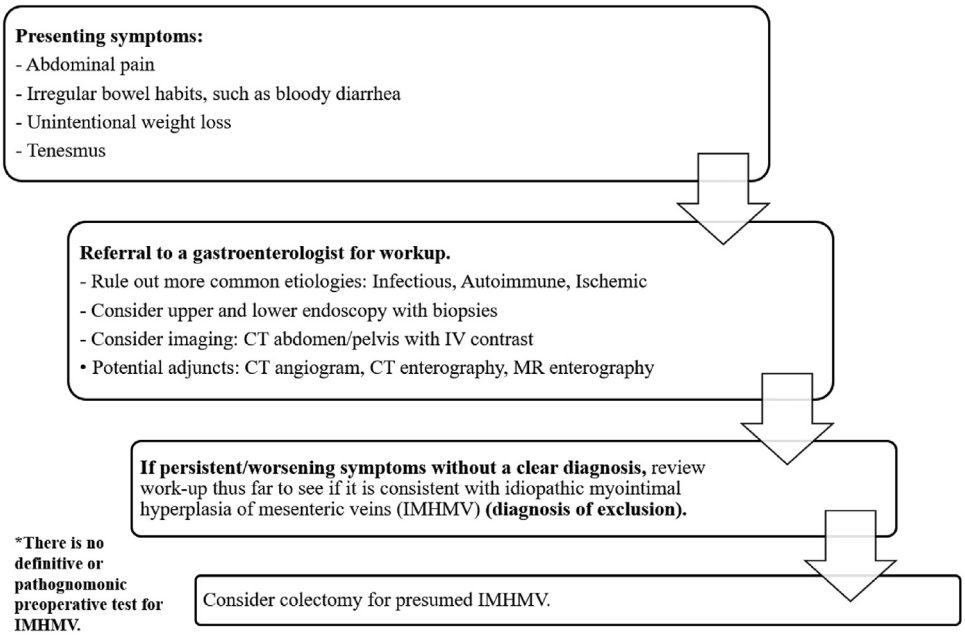

Fig 5. Algorithm for workup of a patient with idiopathic myointimal hyperplasia of mesenteric veins (*IMHMV*).

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.

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