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CASE REPORT

Gastroenterology

Myointimal hyperplasia of the mesenteric veins mimicking inflammatory bowel disease in a 13-year-old patient

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Abstract

Idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV) is a rare entity reported in adults, frequently mimicking inflammatory bowel disease (IBD). We report the first case of intestinal ischemia secondary to IMHMV presenting as IBD in a pediatric patient with Down syndrome. Chronic intestinal ischemia is rare in children, and this case emphasizes the need to consider this in the differential, when histopathology is suggestive. Clinical findings and colonoscopy in these patients may mimic IBD. The mucosal biopsies do not show classic features of IBD. Instead, there can be variable extent of mucosal changes such as dilated small capillaries in the lamina propria, edema, and early fibrosis. These changes may in fact represent early chronic ischemia. These findings should alert for vascular imaging and a full-thickness biopsy to assess submucosal and subserosal larger vessels since medium- and large-caliber veins in the colonic wall and mesentery are affected in IMHMV.

KEYWORDS

Down syndrome, intestinal ischemia, pediatrics

1 | INTRODUCTION

Intestinal ischemia is rare in pediatrics, and the clinical presentation can range from acute intestinal obstruction presenting with shock to chronic diarrhea. It can occur due to occlusive or nonocclusive involvement of the mesenteric vasculature; arteries are affected more than veins. The latter is prone to being misdiagnosed, frequently as inflammatory bowel disease (IBD).

Chronic venous ischemia is extremely uncommon in children. Herein, we report a case of a 13-year-old female with trisomy 21, who initially presented with clinical, radiologic, and endoscopic features of IBD. Histopathology and subsequent radiologic workup demonstrated features of chronic intestinal ischemia caused by non-thrombotic and non-inflammatory occlusion of the mesenteric veins. The findings were compatible with idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV), an uncommon cause of chronic intestinal ischemia in adult patients. The youngest patients previously reported in the literature were 21 and 22.

2 | CASE REPORT

A 13-year-old female patient with trisomy 21 and a history of duodenojejunostomy as a neonate for duodenal atresia, now presented with a 1-week history of bloody diarrhea, abdominal pain, and vomiting. The inflammatory markers were elevated, and infectious workup was negative. She was initially treated with antibiotics with some improvement but later deteriorated. A computed tomography scan

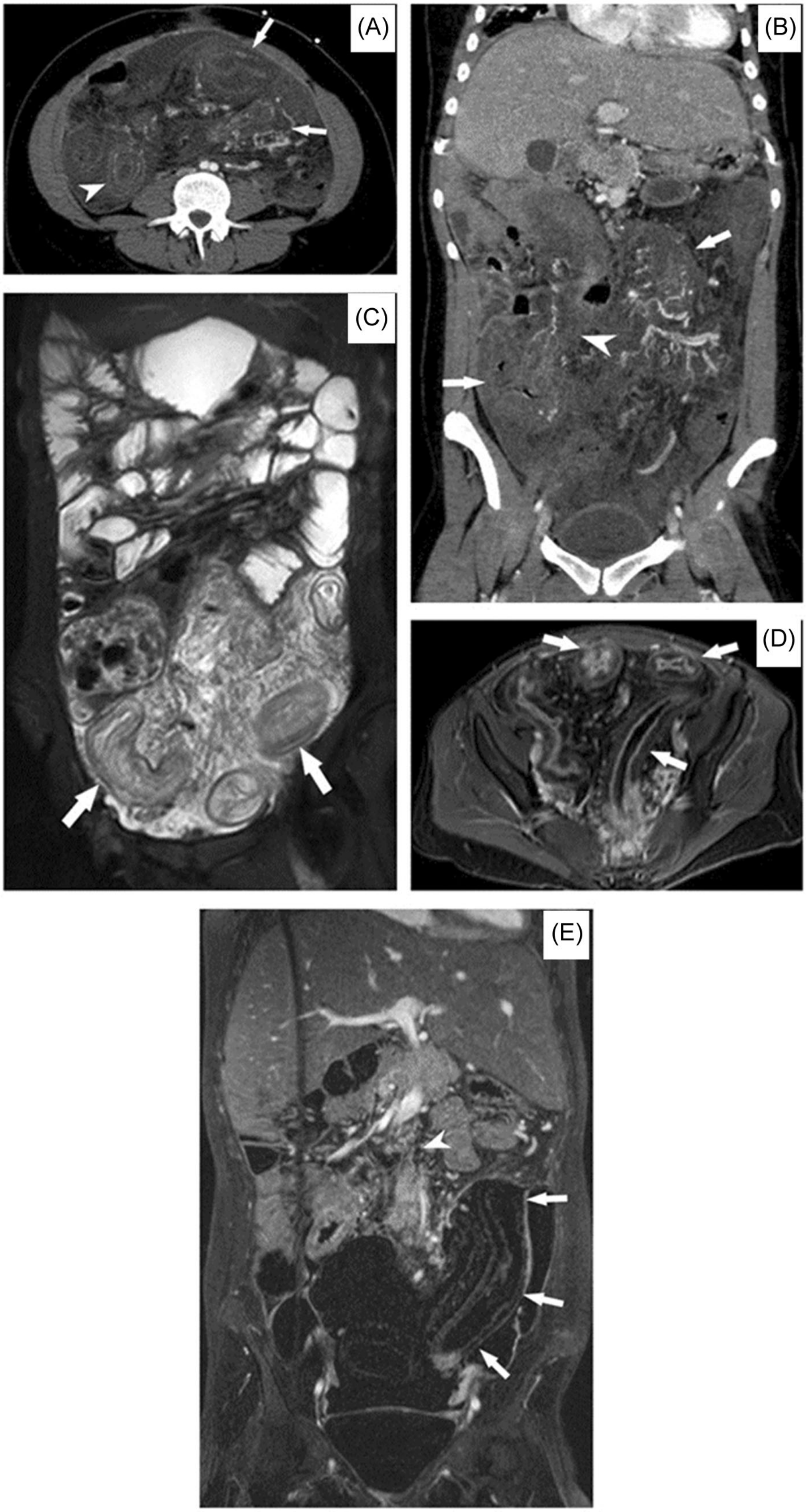

FIGURE 1 (See caption on next page).

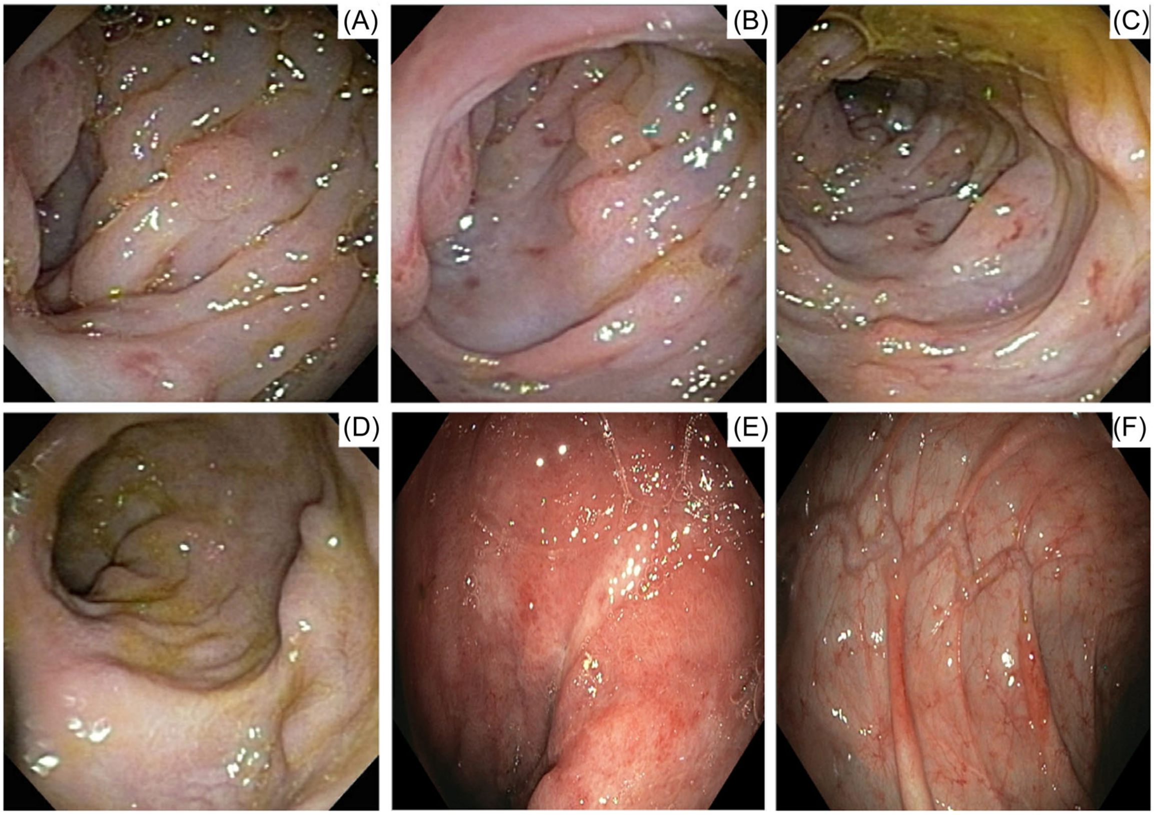

FIGURE 2 First endoscopy showed inflamed swollen mucosa involving cecum (A), ascending (B), and transverse (C), with relatively normal rectosigmoid (D). The repeat endoscopy showed a left colon with continuous erythema and some discrete moderate sized ulcers (E) and prominent vessels running along the transverse colon (F).

was performed which demonstrated mural thickening of the colon and ileum including the terminal ileum (Figure 1A,B). On endoscopy, there were discrete ulcers, patchy erythema, and pseudopolyps involving the proximal colon up to the transverse (Figure 2A–C), while the left colon was macroscopically unremarkable (Figure 2D). Colonic biopsies (cecum, ascending, transverse, descending, sigmoid, and rectum), microscopically showed a lack of an increase in chronic inflammation in the lamina propria, prominent dilated small capillaries and lamina propria edema with early fibrosis (Figure 3A).

She was started on enteral therapy in suspicion for an evolving IBD but continued to have intermittent episodes of bloody diarrhea and elevated inflammatory markers. At this point, the clinical diagnosis was still unclear. Thus, a repeat endoscopy was performed after 5 months. Macroscopically, the left colon showed erythema and discrete moderate-sized ulcers (Figure 2E), with prominent vessels running along the transverse colon (Figure 2F), features again raising the clinical suspicion for IBD, specifically Crohn's disease. The colonic biopsies showed similar features of the prominence of lamina propria vasculature as seen in the previous biopsies, with no significant inflammation or crypt architectural changes suggestive of IBD.

In the context of recurrent bloody diarrhea with no signs of improvement, a magnetic resonance (MR)-enterography was performed which showed features suggestive of IBD, characterized by wall thickening of the large bowel, displaying enhancement on post-contrast (Figure 1C,D). Based on these findings, oral prednisone was initiated.

FIGURE 1 Post contrast axial and coronal CT images of the abdomen demonstrate marked thickening of terminal ileum (arrowheads) and colon (arrows) (A, B). Coronal T2-weighted image shows marked thickening of large bowel loops (arrows) with submucosal edema. There is also edema of the mesentery (C). Axial postcontrast T1-weighted images show marked thickening of the large bowel wall (arrows) including sigmoid. There is avid prominent enhancement of the mucosa in these loops (D). Postcontrast dynamic T1-weighted coronal image in the portal venous phase demonstrates significant hypoenhancement of the descending colon (arrows). An inferior mesenteric vein was not clearly visualized. There is also significant attenuation of mesenteric venous branches (arrowhead) below the main stem of SMV (E). CT, computed tomography; SMV, superior mesenteric vein.

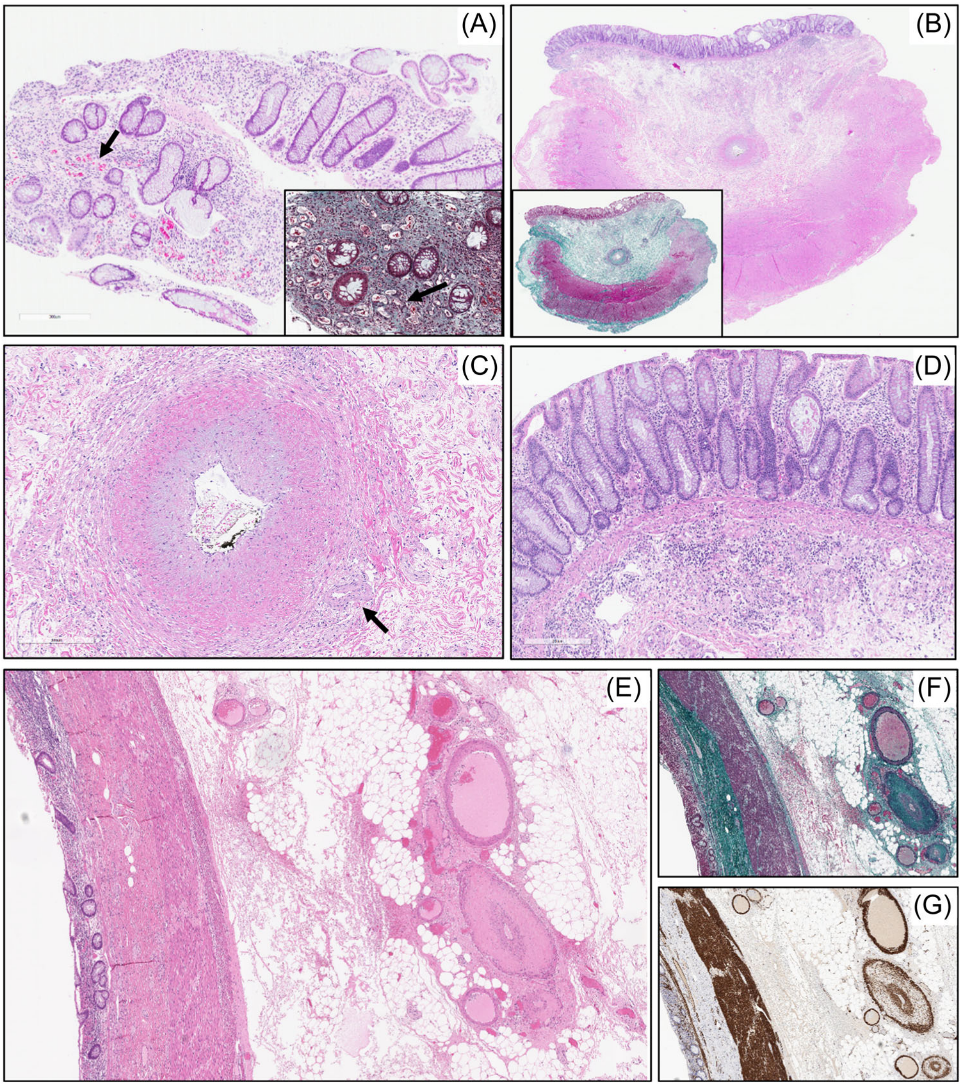

FIGURE 3 (A) H&E and Masson trichrome stain (inset) showing ascending colon biopsy with prominent blood vessels (arrows) in an edematous lamina propria with no significant inflammation and early fibrosis (×200 magnification). (B) H&E and Masson trichrome stain (inset), full-thickness colonic biopsy showing submucosal edema and fibrosis (×100 magnification). (C) H&E stain showing high magnification (×400) of medium-sized submucosal vein with marked intimal hyperplasia and near-complete luminal obliteration, the normal adjacent artery can be seen (arrow). (D) H&E stain showing the presence of numerous dilated and hyalinized "arterialized" small vessels in the submucosa (×400 magnification). (E) H&E of resected large intestine after left hemicolectomy, revealing myointimal hyperplasia of the mesenteric vein (×100 magnification); (F, G) Masson trichrome stain and smooth muscle actin immunohistochemistry highlighting myointimal hyperplasia of mesenteric veins. Note that the adjacent arteries are unremarkable (×200 magnification). H&E, hematoxylin and eosin.

A month later, the patient presented with sepsis and bowel obstruction, which required an exploratory laparotomy and diverting ostomy. Intraoperatively, the colon was diffusely edematous and thickened, worse in the rectum and sigmoid. A full-thickness open biopsy was then taken from the descending colon, which showed similar but exaggerated prominence of lamina propria capillaries (Figure 3B). Numerous dilated and hyalinized "arterialized" small vessels in the superficial submucosa were also noted (Figure 3D). Additionally, the submucosa showed marked intimal hyperplasia of medium-sized veins with near-complete luminal obliteration, while arteries were spared (Figure 3C). There was no significant chronic or acute inflammatory infiltrate. These histologic features were similar to IMHMV and were suspected to be related to a veno-occlusive abnormality within the larger veins (i.e., mesenteric and/or portal).

Subsequently, an MR-angiography was performed which demonstrated a markedly abnormal large bowel with significant hypoperfusion and ischemia, presenting with nonvisualization of the entire inferior mesenteric vein (IMV) and attenuation of the lower segment of superior mesenteric vein (Figure 1E). Two years later, the patient developed strictures and underwent two subsequent surgeries, a left hemicolectomy followed by jejunal and ileal segmental resection with stoma revision. The resected

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TABLE 1 Clinical, radiologic, and histopatholgic differences between Crohn's disease and IMHMV.

	Crohn's disease	IMHMV
Age	Childhood to adulthood (<16–>40 years of age) with a second peak at the fifth to sixth decade.	*13–83 years (mean age 53) males (80%).
Clinical features	Chronic abdominal pain, diarrhea, weight loss, perianal disease (abscesses, fistulas, and fissures), and extraintestinal manifestations (6–25%).	Chronic abdominal pain, diarrhea, and hematochezia. No extraintestinal manifestations.
Common site of involvement	- Ileocolonic - colonic. - ileal - any of the above with upper GI involvement in 5%–30% children and adolescents - isolated upper GI	Disease mostly localized to the sigmoid and rectum. The small bowel is rarely involved. No upper GI involvement.
Histopathology findings	Ileocolonic patchy and segmental distribution with skip lesions. Chronic colitis with increased chronic inflammation and crypt architectural distortion. Activity includes neutrophilic inflammation, cryptitis, crypt abscess, erosions, and ulceration. Deep fissuring ulcers. Granulomas (in approximately 50%). Transmural inflammation with lymphoid aggregates in the subserosal adipose tissue. Fistula and sinus tract formation.	Mucosa and superficial submucosa: arterialized capillaries, subendothelial fibrin deposits, and perivascular hyalinization of the lamina propria small vasculature. In full-thickness biopsy and resections: marked myointimal hyperplasia of submucosal and mesenteric veins. No chronic colitis or activity.
Radiological findings	Commonly used to assess small bowel disease (ultrasonography, CT scan, and MRI) Small bowel findings: asymmetrical segmental mural hyperenhancement. Other less specific findings such as wall thickening, intramural edema, strictures, proximal bowel dilation, and ulcers. Pelvic MRI: used to evaluate and define perianal fistula tracts. Colonoscopy: small aphthous and transmural ulcers, linear and serpiginous ulcers with intervening unaffected mucosa (Cobblestone appearance), and skip lesions.	CT scan: wall thickening in most patients, pericolic fat stranding, decreased mural enhancement, small bowel or colonic dilatation and rarely pneumatosis. Angiography: patent mesenteric arterial vasculature. One-third showed anatomical venous abnormalities near the affected intestinal regions. Nonopacification or absence of draining veins (i.e., IMV, left colic vein, and right colic vein), congested, tortuous, ecstatic veins, and marked collaterals.
Treatment	Medical therapy such as corticosteroids, thiopurines, methotrexate, and anti-TNF agents. Surgical therapy: when there are complications such as stricturing disease, fistulizing disease, and perianal disease (fistula or abscess).	Surgical resection is the treatment of choice.
Recurrence	Recurrence is common after 10 years of diagnosis, most commonly as ileocolonic disease. Postoperative recurrence is dependent on disease location. An increased risk for colon and small bowel carcinomas, anal squamous cell carcinoma, and lymphoproliferative diseases.	Risk of postsurgical recurrence is extremely rare.

Note: *Indicate youngest patient reported in this case report.

Abbreviations: CT, computed tomography; GI, gastrointestinal; IMHMV, idiopathic myointimal hyperplasia of the mesenteric veins; MRI, magnetic resonance imaging; TNF, tumor necrosis factor.

colon showed similar features of venous myointimal hyperplasia of the submucosal, subserosal, and mesenteric veins (Figure 3E–G). There was no evidence of vasculitis or thrombosis. The arteries were unremarkable. The histopathology of the subsequent ileum and jejunum, showed similar changes only confined to the ileum.

3 | DISCUSSION

We report the first case of IMHMV presenting as chronic intestinal ischemia in a pediatric patient, who clinically presented as IBD. Our patient is also the first patient reported with Down syndrome and IMHMV. She was later investigated and had no other underlying condition, particularly vasculitis or thromboembolic disease. IMHMV is a non-thrombotic and non-inflammatory distinct and rare cause of chronic intestinal ischemia in adult patients. The mesenteric veins harbor myointimal proliferation leading to obliteration of the vascular lumen and chronic ischemia. Genta and Haggitt first described IMHMV in 1991.[1] In a recent large systemic review done for IMHMV cases,[2] 124 patients were identified; 80% were males with an average age of 53. The commonest symptoms were abdominal pain and diarrhea. Rectosigmoid was the most affected area. Eleven percent involved the small bowel, mainly the ileum. Approximately one-third of patients had complications due to diagnostic delay. Angiography abnormalities were identified in 35%. The most reported findings included the absence of veins, mainly IMV. Other features were congested and tortuous veins and prominent collaterals. Mesenteric arterial vasculature was patent.

On colonoscopy, the findings can range from mucosal erythema, edema, and friability to strictures, pseudopolyps, and ulcers. Radiologically, there can be intestinal wall thickening.[3–8] These clinical, endoscopic, and radiological appearances mimic IBD and less often raise the possibility of ischemic colitis.

Histologically, arterialized capillaries and subendothelial fibrin deposits can facilitate the recognition of IMHMV in mucosal biopsies.[7] Not seen in our patient. As IMHMV affects medium- and large-caliber mesenteric veins, a full-thickness biopsy or segmental resection is needed for diagnosis.

To summarize, IMHMV is a rare entity reported in adults, frequently mimicking IBD. This case underscores the critical significance of considering this diagnostic differential, irrespective of the patient's age, particularly when the histopathology is not typical for IBD and is suggestive of chronic ischemia. Moreover, early diagnosis is essential to avoid unnecessary treatment and delay in surgical intervention, which in most cases is curative. If untreated, the complications may be life-threatening (Table 1).

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

ETHICS STATEMENT

Informed patient consent was obtained for publication of this case report.

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