Case Report | Pathology

Preoperatively Confirmed Idiopathic Myointimal Hyperplasia of the Mesenteric Veins: A Case Report

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Abstract

Idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV) is a rare cause of colonic ischemia. First documented in the literature in 1991, this condition has remained exceedingly rare, with only a limited number of case reports since its initial mention. It involves proliferation of intimal smooth muscle within the mesenteric veins, resulting in colonic ischemia due to venous constriction and non-thrombotic occlusion. Patients experience a prolonged clinical course and adverse outcomes due to its rarity and nonspecific symptoms. The definitive diagnosis is typically confirmed after surgical resection of the affected colon, as initial biopsies are not deep enough to confirm the diagnosis. Histological examination of these biopsies reveals arteriolization of capillaries, subendothelial hyaline deposits, and fibrin thrombi. We present a case of a patient with a one-year history of diarrhea and abdominal pain. Biopsy samples obtained during colonoscopy showed colitis and proliferation of muscularized capillaries, consistent with IMHMV, prior to colon resection. The patient ultimately underwent laparoscopic total abdominal colectomy to the distal one-third of the rectum with end ileostomy. IMHMV is commonly treated surgically and mismanagement of this condition involving corticosteroids and immunomodulatory agents may result in unfavorable outcomes. This case report is one of the few cases of IMHMV diagnosed preoperatively within existing literature, which highlights a rare etiology of nonspecific colitis, emphasizing the critical divergence in its management approach.

Keywords

Colonic Ischemia; Venous Occlusion; Chronic Diarrhea; Arteriolization of Capillaries

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Introduction

Idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV) is an extremely rare and poorly understood diagnosis, first described in the literature in 1991. A literature review conducted in 2020 revealed only 34 published reports on this condition [1]. IMHMV is caused by proliferation of intimal smooth muscle of the mesenteric veins leading to venous narrowing and occlusion, ultimately causing colonic venous ischemia [2]. In literature reviews of this diagnosis, males with a median age of 53-62 years are more likely to be affected at a ratio of 2.7:1 to 4.8:1, cardiovascular risk factors are often present, and the rectosigmoid colon is the most affected segment [1–6].

IMHMV presents with nonspecific gastrointestinal complaints, such as watery or bloody diarrhea, abdominal pain, and weight loss, and is often misdiagnosed for inflammatory bowel disease (IBD) or arterial causes of colonic ischemia [1, 5]. Symptoms are usually chronic, but it can present with acute onset of diarrhea and abdominal pain [6]. On histology, IMHMV is distinctly different from other
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causes of ischemia, and it is difficult to reach the diagnosis due to the overlap of symptoms and nonspecific findings on colonoscopy [7]. On gross examination, IMHMV causes edema, fat stranding, and friable mucosa with ulcerations, which also leads to misdiagnosis [1]. Historically, histopathology after surgical resection of the affected colon is the definitive way the diagnosis of IMHMV is made [8]. Surgery is the only known curative treatment, but because of the rarity and overlap of symptoms, treatment with corticosteroids and immunomodulatory agents is often incorrectly initiated, and there can be a diagnostic delay in up to 29% of patients [4, 5, 9]. We present a case of a 73-year-old man with a one-year history of diarrhea and significant weight loss who was found to have histologically diagnosed IMHMV prior to surgical resection of the colon.

Case Report

A 73-year-old man with a history of hypothyroidism of unknown etiology, hyperlipidemia (triglyceride predominant), and deep venous thrombosis (DVT) on rivaroxaban presented with a one-year history of watery diarrhea (10-14 episodes daily), abdominal pain, and 15-pound weight loss (starting weight of 218 pounds, 9% weight loss). The patient had no recent infectious symptoms, gastrointestinal conditions or past surgeries, and last screening colonoscopy three years earlier showed only benign polyps. His subsequent lab workup showed normal tissue transglutaminase, immunoglobulin A (IgA), fecal fat, and fecal elastase with a negative fecal immunochemical test. Calprotectin was elevated to 198.3 mcg/g (normal < 50 mcg/g) and C-reactive protein (CRP) was elevated to 9 mg/L (normal < 3.00 mg/L). Testing for giardia, cryptosporidium, Clostridioides difficile, and ova and parasites was negative. Important to note that the patient was on chronic anticoagulation after two lower extremity DVT occurrences (the first episode was provoked by total right knee replacement seven years earlier and the second one was an unprovoked instance affecting the left posterior tibial vein and left peroneal vein a few months before presentation). The patient was followed by a hematologist and underwent a full hypercoagulability workup that was negative. Additionally, the patient had no family history of similar symptoms among close relatives.

Bidirectional endoscopy revealed erythematous, edematous, and friable mucosa with superficial ulceration, most significantly involving the colon 50-70 cm from the anal verge. Computed tomography angiography (CTA) showed edematous walls extending from the mid-transverse colon to the mid-sigmoid colon with moderate inflammation and collateral arteries and veins (Fig. 1-4). The differential diagnosis included ischemic colitis, microscopic colitis secondary to non-steroidal anti-inflammatory drugs (NSAIDs), IBD, and IMHMV. Biopsies revealed an increase in lamina propria capillaries, accompanied by patchy active colitis featuring erosion and injured, and withered crypts. Two pathologists from different institutions agreed that the findings were consistent with IMHMV. Due to the rarity of this condition, there were numerous discussions at multi-

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**Figure 1.** CT findings of wall thickening in the middle to distal transverse colon.

**Figure 2.** CTA scan showing prominent collateral arteries of the proximal descending colon.

**Figure 3.** CTA findings of prominent collateral arteries along the wall of the transverse colon.
disciplinary conferences on how to proceed. In the interim, patient’s diarrhea was controlled with loperamide. No other medications were trialed, as loperamide was sufficient at controlling diarrhea.

Six months after initial colonoscopy, the patient presented with a three-week history of significantly aggravated diarrhea, occurring 20-30 times per day, accompanied by abdominal pain and rectal bleeding. The patient was symptomatically controlled in the interim between the first colonoscopy and this presentation. Abdominal CT with contrast showed colitis extending from the mid-transverse colon to the mid-sigmoid colon with occlusion of the inferior mesenteric vein and its branches. Repeat colonoscopy showed circumferentially ulcerated mucosa beginning 20 cm above the anus. The distal mucosa appeared edematous and mottled with prominent superficial capillaries. Preoperative biopsies again showed increased lamina propria vascularity with proliferation of muscularized capillaries (Fig. 5, 6). The patient was hospitalized due to the severity of ulceration. Both a vascular surgeon and a colorectal surgeon evaluated the patient and recommended colonic resection. The patient underwent laparoscopic total proctocolectomy with end ileostomy due to the predominant disease involvement on the left side of the colon. Post-operative biopsy showed both total and subtotal occlusions of the mesenteric veins with circumferential proliferation of fibrous tissues within the intimal space. The arteries displayed modest atherosclerotic changes, while the capillaries exhibited arteriolization without distinct thrombi or subendothelial hyalinization. The patient had no complications following surgery and reported complete resolution of diarrhea during a follow-up evaluation seven months later.

Discussion

Although IMHMV was first described 30 years ago, few case reports of this condition exist. In this case report, an elderly male with cardiac risk factors presented with a prolonged history of abdominal pain and episodes of bloody diarrhea, raising concerns about chronic colitis. In > 90% of cases, chronic colonic ischemia is caused by arterial atherosclerosis, but it can also result from vasculitis or venous thrombosis, which are significantly more common than IMHMV [10]. Atherosclerotic ischemic colitis ranks high in the list of differentials for an older male exhibiting cardiovascular risk factors, alongside symptoms such as abdominal pain, rectal bleeding, and diarrhea. The etiology of IMHMV is unknown, with some hypotheses suggesting that elevated venous pressures induce vascular remodeling and subsequent endothelial injury [9]. Histological similarities between IMHMV and saphenous vein grafts used in coronary artery bypass grafts suggest that vascular stress and remodeling could be contributing factors [11]. IMHMV is a histological diagnosis. Specific histological findings associated with IMHMV, not found in other types of ischemic colitis, involve arteriolization of capillaries, subendothelial hyaline deposits, and the presence of fibrin thrombi [9, 12, 13]. Angiography reveals mesenteric
vessel patency, a distinctive feature distinguishing it from other causes of colonic ischemia [1, 2]. In our case, on preoperative biopsy, there was increased lamina propria vascularity with proliferation of muscularized capillaries (Fig. 5, 6). Post-operative biopsy revealed both total and subtotal occlusions of the mesenteric veins with circumferential proliferation of fibrous tissues within the intimal space and capillaries exhibiting arteriolization without distinct thrombi or subendothelial hyalinization - all congruent with a diagnosis of IMHMV and preoperative biopsies.

Many patients are treated with steroids or immunomodulatory agents due to misdiagnosis of IBD before obtaining histological confirmation, which can lead to more complications. Our patient was diagnosed preoperatively, so steroids and immunomodulatory agents were avoided. Surgery appears to be the only viable treatment option, exhibiting no recurrence when patients were followed for up to seven years [7]. Our patient remained symptom-free seven months after surgery [8]. This case is noteworthy as it represents one of the rare instances when IMHMV was diagnosed before surgical resection. Prior to this case report, only two cases of preoperatively diagnosed IMHMV have been documented in the literature [14, 15].

**Conclusions**

IMHMV is a rare and poorly understood cause of colonic ischemia mimicking arterial colonic ischemia and IBD. Numerous cases are likely misdiagnosed and recognizing this condition while establishing a differential diagnosis for nonspecific gastrointestinal complaints, along with understanding the histological findings on biopsy, can prevent inappropriate treatment and prolonged disease course.

**Ethical Statement**

This case report was conducted in accordance with the principles outlined in the Helsinki Declaration and adhered to ethical guidelines.

**Informed Consent**

Written informed consent was obtained from the patient for publishing the details of his medical case and any accompanying images prior to drafting this case report and is available upon request.

**Data Availability**

All data generated or analyzed during this study are included in this article. Further enquiries can be directed to the corresponding author.

**Conflict of Interest**

This article is an extended version of a conference abstract given at the American College of Gastroenterology Annual Scientific Meeting (October 25th, 2022, Charlotte, North Carolina). The authors declared no other potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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