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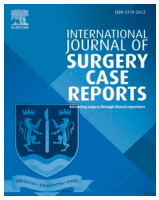

Case report

Atypical presentation of a rare disorder; idiopathic myointimal hyperplasia of mesenteric veins (IMHMV): Report of two cases

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ABSTRACT

Introduction: Idiopathic myointimal hyperplasia of mesenteric veins (IMHMV) is a rare type of chronic colonic ischemia. Patients commonly present with progressive abdominal pain, bloody diarrhea, and weight loss. IMHMV is a common mimicker of inflammatory bowel disease. However, medical management does not have a primary role and curative treatment is surgical resection.

Presentation of case: We report two cases of IMHMV with atypical presentation. The first is an 82-year-old male who had refractory, painless, explosive, and non-bloody diarrhea initially treated with antidiarrheal medications and dietary changes to no effect. Colonoscopy was not clarifying. However, CT scan had characteristic findings of IMHMV. He underwent partial colectomy and recovered well. The second case is a 59-year-old male who had recurrent episodes of sudden, massive diarrhea. He was initially treated for diverticulitis based on colonoscopy findings but did not experience relief. Eventually, MRI of the abdomen was suggestive of IMHMV. He underwent surgical resection, which confirmed the diagnosis of IMHMV. He was treated for Clostridioides difficile diarrhea five months after surgery and pulmonary embolism seven months after surgery. With over a year of follow up, neither has had disease recurrence.

Discussion: Diagnosis and treatment of rare disorders like IMHMV is challenging, especially when they mimic common entities or present in atypical ways.

Conclusion: We present two cases to highlight IMHMV as part of the differential for colitis-like symptoms. These cases demonstrate the importance of diagnostic imaging in diagnosis. Diagnostic uncertainty can lead to exposure to ineffective medical treatments and delay in curative surgery.

1. Introduction

A rare cause of colonic ischemia is idiopathic myointimal hyperplasia of mesenteric veins (IMHMV). Symptoms typically are progressive abdominal pain, bloody diarrhea, and weight loss. IMHMV mimics inflammatory bowel disease (IBD) but needs to be differentiated as medical treatment yields almost no response, while surgery is curative [1]. IMHMV is characterized by intimal hyperplasia of the extramural veins; occasionally submucosal veins and mucosal capillaries are involved [2]. Since its first description in 1991 [3], around 70 patients have been reported in the literature. The etiology is unclear. IMHMV typically affects the rectosigmoid colon of healthy males [1,3,4]. We report two cases of IMHMV with atypical symptoms, ultimately diagnosed radiographically and treated surgically. The work being reported is in line with the SCARE criteria [5].

2. Presentation of case

2.1. Case 1

An 82-year-old male with non-insulin dependent diabetes mellitus, hypertension, glaucoma, and sleep apnea experienced painless, bloodless, explosive diarrhea. The patient experienced multiple episodes of nocturnal diarrhea and incontinence to mucus discharge for two days followed by no bowel movements for several days.

Abbreviations: IMHMV, idiopathic myointimal hyperplasia of mesenteric veins; IBD, inflammatory bowel disease; CT, computerized tomography; MIVOD, mesenteric inflammatory venoocclusive disease; MRI, magnetic resonance imaging; IMV, inferior mesenteric vein; GI, gastrointestinal.

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2.1.1. Clinical findings

Stool studies were negative for enteric pathogens and parasites. Computerized tomography (CT) scan suggested mesenteric ischemia-induced colitis secondary to an infectious/inflammatory etiology, with diffuse wall thickening of the descending and sigmoid colon, mesenteric adenopathy, hyperemic changes, and no mesenteric artery stenosis. Fiber supplementation, antibiotics, and avoidance of lactose to treat small intestinal bacterial overgrowth had no effect. Loperamide led to moderate improvement of the nocturnal mucus discharge and diarrhea. The patient continued to have erratic bowel movements, bloating, urgency, and seepage of mucus. The patient had lost 35 pounds in three-months' time, had significant fatigue, and lack of appetite.

At initial presentation to our institution, three months after symptom onset, abdominal and rectal exams were unremarkable. The patient was anemic (hemoglobin 12.9 g/dL), but otherwise his laboratory evaluations were normal. Colonoscopy showed ischemic-type mucosal injury with erythema and loss of vascularity, moderate severity edema, and continuous, circumferential friability from the distal rectum to the descending colon (Fig. 1). Biopsy showed vascular dilation and congestion, consistent with ischemic colitis. The findings were not specific, with a broad differential including ischemia, IMHMV, drug-induced injury, mesenteric inflammatory venoocclusive disease (MIVOD) and IBD. The CT enterography demonstrated severe colitis involving the descending through sigmoid colon with occluded inferior mesenteric vein and tortuous pericolonic collaterals, suspicious IMHMV or MIVOD (Fig. 1). Both of these rare disorders are often clinically confused with ischemic colitis and IBD. Neither respond to medical treatment and both are treated with surgical resection [6].

2.1.2. Surgical intervention

The patient was referred to our colorectal surgery service. Given the patient's incontinence and low rectal involvement, resection of the affected bowel with creation of an end colostomy was recommended. The initial operative approach was laparoscopic. However, due to inflammatory obscuring of tissue planes, he ultimately underwent open resection of the distal descending colon to the rectum with creation of end colostomy. Pathology revealed thick-walled, muscularized veins in the subserosa and submucosa with normal-appearing arteries. The mucosa had patchy erythema, areas of erosion, focal ischemic-type changes, ulceration, reactive small vessel proliferation, fat necrosis, and subserosal fibrosis. Obstruction of the mesenteric veins secondary to hyperplasia of intimal smooth muscle, in the absence of thrombosis or inflammation, was characteristic of IMHMV. The absence of lymphocytic inflammatory infiltrate excluded MIVOD [6,7]. He was discharged four days postoperatively.

2.1.3. Follow-up

Over one year after surgery, he was doing well, without issue.

2.2. Case 2

A 59-year-old male with hypertension presented after several episodes of urgency followed by massive diarrhea. Colonoscopy and biopsy suggested acute diverticulitis, and the patient was prescribed antibiotics. The patient subsequently developed frequent tenesmus with copious gas and clear liquid discharge. Because of symptom progression despite antibiotics, CT scan was done and showed concentric sigmoid thickening and periaortic lymphadenopathy suggestive of smoldering colitis or IBD. The patient had limited improvement with chlordiazepoxide/Clidinium bromide and Imodium.

2.2.1. Clinical findings

Due to COVID-19 restrictions, international travel for evaluation at our institution was limited, so the patient was advised to undergo repeat colonoscopy and MRI locally given persistence of symptoms. Colonoscopy showed marked worsening of the sigmoid colitis, pseudopolyp formation, and necrotic areas. The MRI showed enhancing polypoidal mucosal thickening of the rectosigmoid with pericolic hyperemia and small, reactive lymph nodes. The inferior mesenteric vein (IMV) was thrombosed, and there was collateral circulation (Fig. 2). The findings suggested IMHMV. Thrombophilia workup, done given the IMV thrombosis, revealed Factor II Prothrombin G2021A mutation carrier, and MTHFR mutation A1298C homozygote. The gastrointestinal (GI) pathogen panel was negative.

Eventually, the patient was able to travel to our clinic for surgical consultation and treatment.

2.2.2. Surgical intervention

Laparoscopic low anterior resection with anastomosis was performed (Fig. 2). Pathology showed secondary ischemic-type mucosal changes, ulceration, and polypoid areas of mucosal regeneration. Verhoeff–Van Gieson stain was used, which identifies vascular elastic laminae and helps differentiate arteries and veins. Staining is absent in areas of myointimal hyperplasia (Fig. 3) [8]. Mesenteric veins with prominent myointimal hyperplasia and areas of luminal obliteration in the pericolonic soft tissue and submucosa were found, confirming IMHMV. The patient was discharged on postoperative day six.

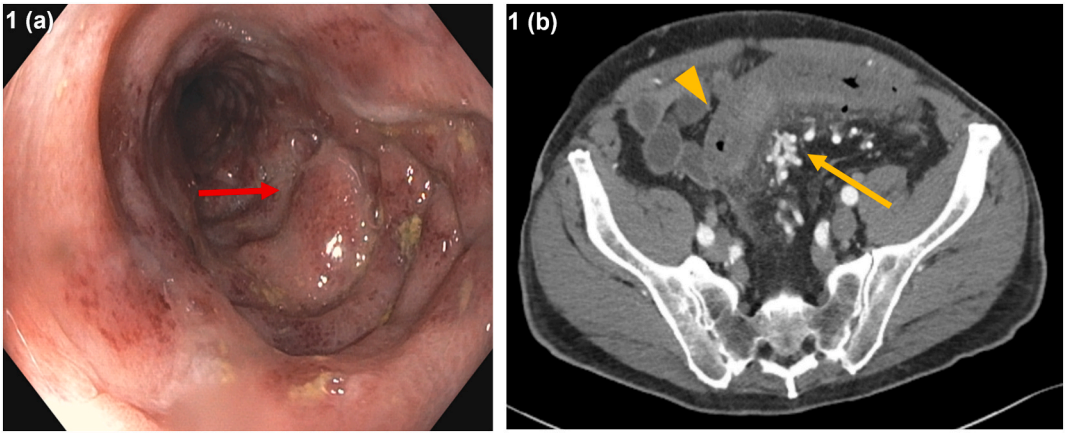

Fig. 1. Case 1 — Endoscopic and radiographic findings of IMHMV.
(a) Colonoscopic images showing erythema, edema, and psuedopolyp (red arrow) in the colon; (b) CT scan, axial image showing the thickened colon wall (yellow arrowhead) and multiple venous collaterals (yellow arrow) characteristic of IMHMV. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

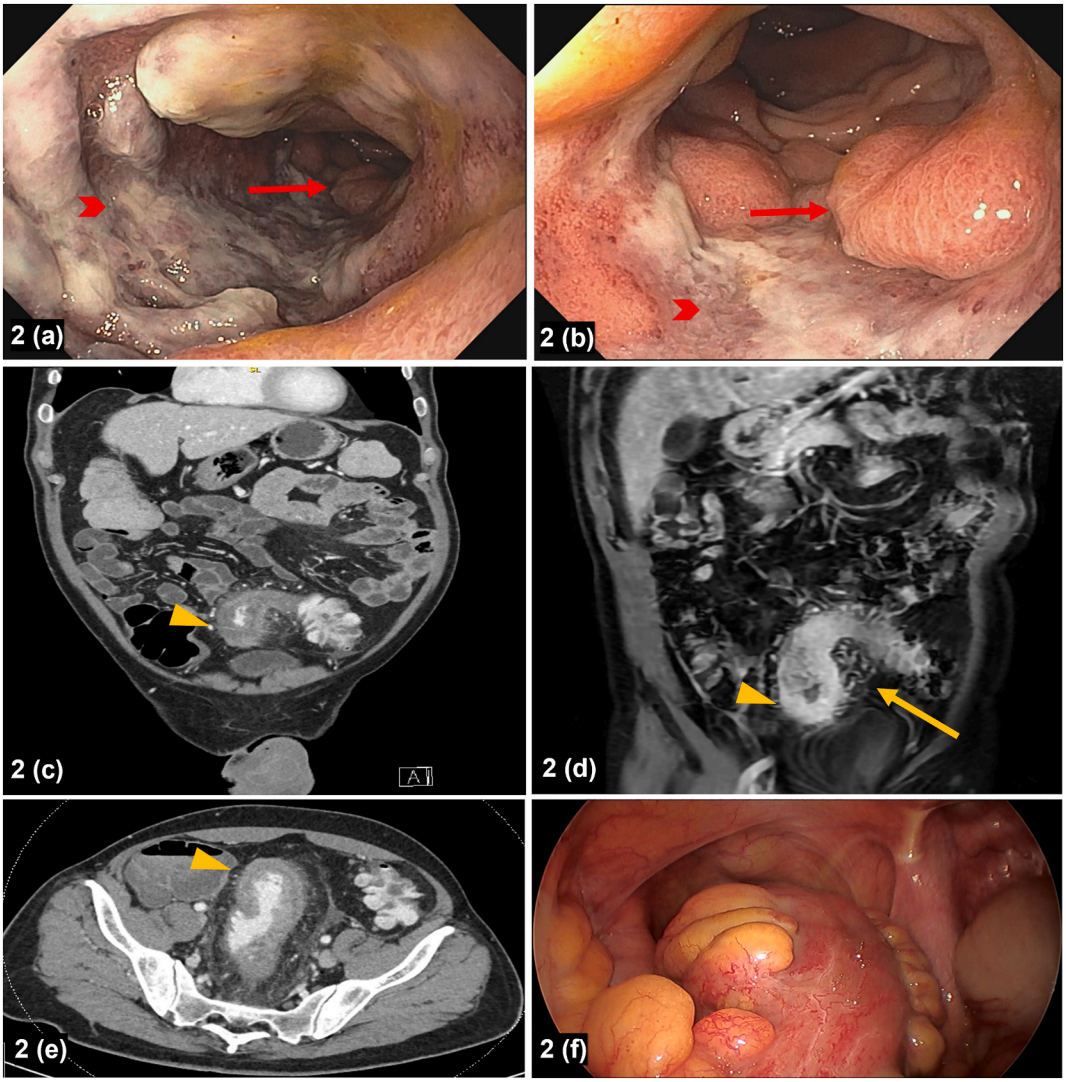

Fig. 2. Case 2 — Endoscopic, radiographic, and intraoperative imaging findings of IMHMV.
(a–b) Colonoscopic images showing ischemia (red double arrowhead) and pseudopolyps (red arrow). (c, e) CT scan, coronal, and axial images, showing the thickened colon wall (yellow arrowhead). (d) Coronal MRI LAVA-flex images showing prominent pericolic vessels (yellow arrow) and thickened colon wall (yellow arrowhead). (f) Intraoperative image, abdominopelvic view, showing hyperemic, thickened distal sigmoid colon. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

2.2.3. Follow-up

Five months after surgery, the patient again had diarrhea. The MRI showed expected postoperative changes. Flexible sigmoidoscopy showed colonic edema and mild inflammation. A CT enterography showed no evidence of myointimal hyperplasia. Clostridioides difficile testing was positive, and the patient was successfully treated with oral vancomycin. Seven months after surgery, the patient developed a pulmonary embolism and was treated with rivaroxaban. At one year follow up, the patient still suffered from diarrhea. Testing was negative for C. diff infection. Flexible sigmoidoscopy was normal. Hydrogen breath test revealed an increase in breath hydrogen and that the patient was a methane producer. Probiotics resulted in normalization of bowel function with two formed stools per day.

3. Discussion

IMHMV is a non-thrombotic, non-inflammatory venous hyperplasia that leads to chronic colonic ischemia [3,9]. It mostly affects men of any age (21 to 83 years reported) [10]. Diagnosis is commonly delayed and medical therapy is ineffective. Treatment for this progressive disease is surgical resection of the effected colon. We report two cases of IMHMV with atypical presentations that were successfully treated surgically.

The most commonly reported presentation of IMHMV is abdominal pain, hematochezia, diarrhea, and weight loss [1,11]. Both of our patients had diarrhea, but it was not bloody. Neither had abdominal pain. The first case did experience weight loss, while the second did not. Their clinical presentations were non-specific. Both patients experienced intermittent episodes of massive or explosive diarrhea with seepage of mucus, which may suggest new clinical clues for adding IMHMV to one's differential diagnosis.

As was the case for our patients, IMHMV most commonly involves the rectosigmoid colon [1,9]. In a comprehensive report of 70 cases of IMHMV, 10 % had small intestinal involvement, and 10 % had extended colonic involvement proximal to the descending colon [1]. Endoscopic features of IMHMV are generally nonspecific [1,2,12]. While biopsies can help, diagnostic yield is low. Biopsy may show prominent thick-walled vessels in the mucosa and submucosa with associated mild mucosal fibrosis [11], severe ulceration, and granulation tissue without amyloid. Arterialization of capillaries, subendothelial fibrin deposits, and fibrin thrombi have been described [8,9,12,13]. An important point

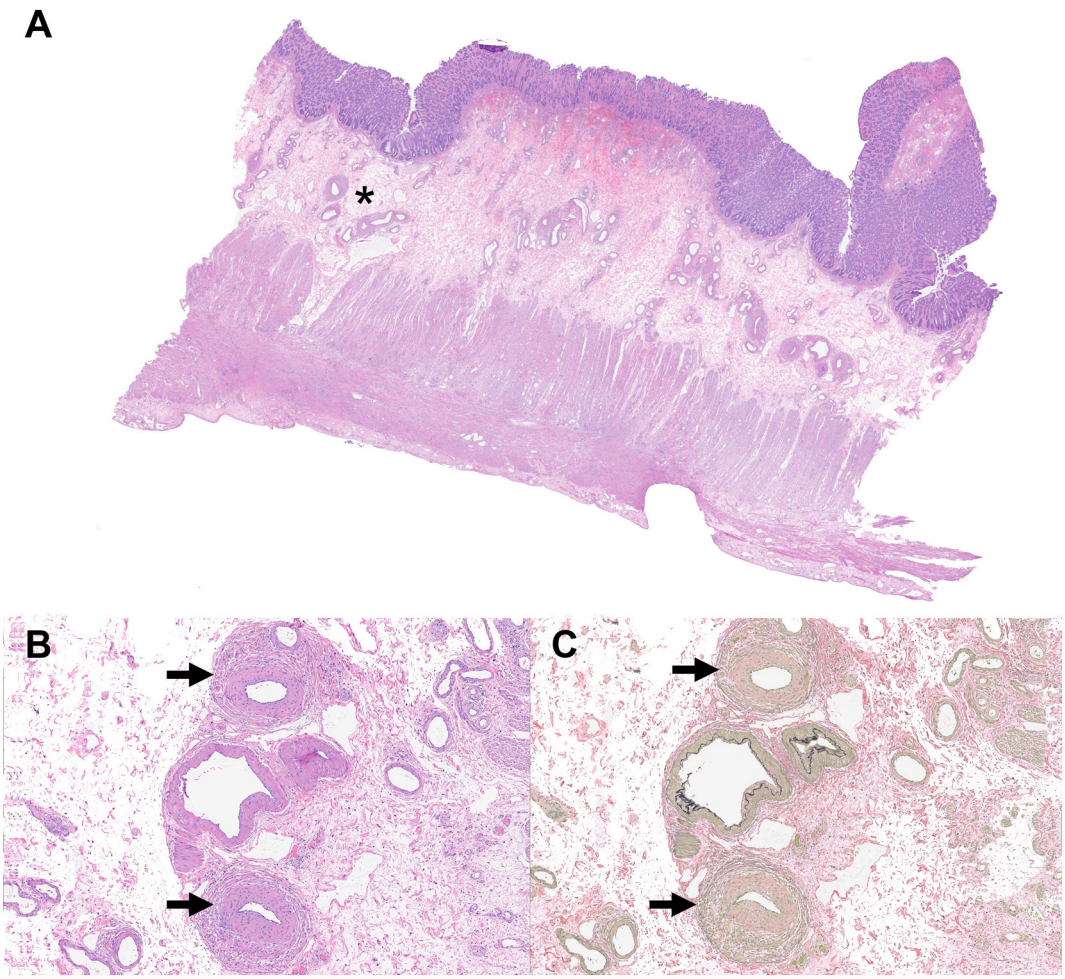

Fig. 3. Histopathologic findings in idiopathic myointimal hyperplasia of mesenteric veins. A. Routine hematoxylin and eosin (H&E) stain showing abnormal, dilated blood vessels in the submucosa (asterisk) (H&E stain, 10× digital magnification). B and C. Histologic details of the abnormal submucosal blood vessels within the submucosa, featuring arterialized veins (arrows) which show no elastic layer in Verhoeff-Van Gieson stain (B). Notice a pair of normal arteries with normal inner elastic layer (black line) in between abnormal veins (H&E and Verhoeff-Van Gieson stains, 80× digital magnification).

is that biopsy results do not support a diagnosis of inflammatory bowel disease [6]. Endoscopy and biopsy did not lead to definitive diagnosis for our patients. Cross-sectional imaging can be diagnostic of IMHMV. Focal colonic thickening and edema with normal patency of mesenteric arteries is typically reported [2]. Additional findings include pericolic fat stranding with rich, dilated, winding peripheral veins [1]. Both of our patients were diagnosed with IMHMV by cross-sectional imaging.

Delayed diagnosis of this rare entity is common given its symptomatic overlap with IBD and ischemic colitis [6]. The second case had delay from both diagnostic uncertainty and COVID-19 pandemic travel restrictions. The first case had a more rapid time to diagnosis, despite atypical symptoms, with recognition of IMHMV on imaging. Because IMHMV is refractory to medical management [6,14], timely diagnosis may decrease exposure to expensive and ineffective medical therapies. Suspicion or diagnosis of IMHMV should prompt surgical referral. Symptoms are the most common indication for elective surgery for IMHMV. Bleeding, perforation, and acute abdomen are reasons for emergent surgical treatment [9,10,13]. While there are no specified guidelines, surgery generally includes resection of affected segments to normal mucosa and grossly normal vasculature. To date, there are no reported recurrences after surgery [9,10,13]. Both of our patients are doing well and without recurrence over one year after surgery. Surgical outcomes have not otherwise been reported. Neither of our cases experienced early postoperative complications. One was treated for C. diff diarrhea five months after surgery; and subsequent, persistent diarrhea was treated with probiotics. The patient was treated for pulmonary embolism seven months after surgery.

4. Conclusion

Our cases demonstrate the importance of diagnostic imaging in differentiating IMHMV from more common causes of colitis. Including IMHMV in the differential diagnosis for those with typical colitis symptoms, as well as for atypical presentations among patients with persistent diarrhea, may allow for a quicker time to diagnosis and surgery. For both cases, cross-sectional imaging was the key to diagnosis. The postoperative course for both patients was favorable. As in other series, neither has experienced disease recurrence after surgical resection.

CRediT authorship contribution statement

H.B., S.S., K.R. were involved in the study concept or design.
H.B., K.R. were involved in data collection.
H.B., R.M. K.R. were involved in data interpretation.
H.B., R.M. S.S., K.R. contributed to manuscript revisions.

Consent for publication

Written informed consent was obtained from both patients to publish

this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal upon request.

Ethics statement

Ethics approval was not necessary per the Mayo Clinic (Rochester, Minnesota, USA) IRBe Human Subjects Research Wizard Tool, and in accordance with the Code of Federal Regulations, 45 CFR 46.102. A copy of the document is available upon request.

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Declaration of competing interest

The authors declare no conflict of interest. All procedures were performed in studies involving human participants followed the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declare and its later amendments or compared ethical strands.

Data availability

Detailed patient information including history, examination, laboratory results, imaging studies, operative and follow up details of this case are available in the hospital as an electronic medical record and can be submitted upon request.

References

[1] H. Li, H. Shu, H. Zhang, M. Cui, Y. Gao, F. Tian, Idiopathic myointimal hyperplasia of the mesenteric veins: a case report and scoping review of previously reported cases from clinical features to treatment, Front. Med. (Lausanne) 9 (2022) 855335.
[2] B. Anderson, T.C. Smyrk, R.P. Graham, A. Lightner, S. Sweetser, Idiopathic myointimal hyperplasia is a distinct cause of chronic colon ischaemia, Colorectal Dis. 21 (9) (2019) 1073–1078.
[3] R.M. Genta, R.C. Haggitt, Idiopathic myointimal hyperplasia of mesenteric veins, Gastroenterology 101 (2) (1991) 533–539.
[4] K. Sahara, R. Yamada, T. Fujiwara, K. Koizumi, S. Horiguchi, T. Hishima, et al., Idiopathic myointimal hyperplasia of mesenteric veins: rare case of ischemic colitis mimicking inflammatory bowel disease, Dig. Endosc. 27 (7) (2015) 767–770.
[5] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (2020) 226–230.
[6] J. Platz, N. Hyman, Idiopathic myointimal hyperplasia of mesenteric veins, Gastroenterol. Hepatol. (N. Y.) 8 (10) (2012) 700–702.
[7] M.J. Flaherty, J.T. Lie, R.C. Haggitt, Mesenteric inflammatory veno-occlusive disease. A seldom recognized cause of intestinal ischemia, Am. J. Surg. Pathol. 18 (8) (1994) 779–784.
[8] M.A. Piccinin, J. Schwartz, Histology, Verhoeff stain, in: StatPearls. 2023, StatPearls Publishing LLC, Treasure Island (FL), 2023. StatPearls Publishing Copyright ©.
[9] F.C. Martin, L.S. Yang, S.R. Fehily, B. D'Souza, A. Lim, P.A. McKelvie, Idiopathic myointimal hyperplasia of the mesenteric veins: case report and review of the literature, JGH Open 4 (3) (2020) 345–350.
[10] L. Feo, A. Cheeyandira, D.M. Schaffzin, Idiopathic myointimal hyperplasia of mesenteric veins in the elderly, Int. J. Colorectal Dis. 28 (3) (2013) 433–434.
[11] A.K. Abu-Alfa, U. Ayer, A.B. West, Mucosal biopsy findings and venous abnormalities in idiopathic myointimal hyperplasia of the mesenteric veins, Am. J. Surg. Pathol. 20 (10) (1996) 1271–1278.
[12] R.K. Yantiss, I. Cui, N.C. Panarelli, J. Jessurun, Idiopathic myointimal hyperplasia of mesenteric veins: an uncommon cause of ischemic colitis with distinct mucosal features, Am. J. Surg. Pathol. 41 (12) (2017) 1657–1665.
[13] P.C. Kao, J.A. Vecchio, N.H. Hyman, A.B. West, H. Blaszyk, Idiopathic myointimal hyperplasia of mesenteric veins: a rare mimic of idiopathic inflammatory bowel disease, J. Clin. Gastroenterol. 39 (8) (2005) 704–708.
[14] R. Wong, D. Westerveld, H. Yeo, J. Jessurun, A. Jesudian, Ischemic colitis from idiopathic myointimal hyperplasia of the mesenteric veins in a post-liver transplant patient, ACG Case Rep. J. 8 (11) (2021), e00692.