International Journal of Surgery Case Reports 83 (2021) 106022

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

Idiopathic Myointimal Hyperplasia of Mesenteric Veins (IMHMV) with two spontaneous bowel perforations: A case report and literature review

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ARTICLE INFO

Keywords:
Case report
Idiopathic Myointimal Hyperplasia of Mesenteric Veins
IMHMV
Colitis

ABSTRACT

Introduction and importance: Idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV) is a poorly understood disorder which poses a diagnostic challenge to clinicians and pathologists. Here we have described the case of a male patient with IMHMV along with a presumed history of ulcerative colitis for 1 year.

Case presentation: A 55-year-old male presented to the OPD with history of chronic abdominal pain. Clinical and radiological examination coupled with endoscopic findings resulted in the patient being wrongly diagnosed to be a case of ulcerative colitis and was managed accordingly. Throughout his multiple hospital visits following treatment for ulcerative colitis, the patient was persistently symptomatic. He presented with 10 days history of increasing abdominal pain and constipation following which he developed spontaneous colonic perforation for which he underwent exploratory laparotomy left colectomy and Hartman's procedure. The final pathology of the resected colon found to be consistent of Idiopathic myointimal hyperplasia of the mesenteric veins and ischemic bowel changes.

Clinical discussion: The absence of clear-cut endoscopic biopsy findings of ulcerative colitis made radiological picture to be the mainstay for diagnosis, which was inaccurate and exposed the patient to unnecessary treatment with immuno-modulators thus resulting in poor response to treatment. As the disease progressed, further narrowing of the vessels made the clinical picture to look closer to ischemic bowel pathology as the patient developed a top surgical emergency (i.e. bowel perforation). Such pathological finding (IMHMV) can only be diagnosed in a fully prepared tissue histology, but rather be considered when no other consistent alternative diagnosis was found.

Conclusion: The treating physicians must definitely consider the possibility of idiopathic myointimal hyperplasia of mesenteric veins when similar manifestations are encountered in biopsy specimens of old cases with suspected inflammatory bowel disease or non-occlusive ischemia of the distal colorectum.

1. Introduction

Idiopathic Myointimal Hyperplasia of the Mesenteric Veins (IMHMV) is a poorly understood disease condition that poses diagnostic challenge to clinicians and pathologists. It is described as an ischemic bowel disease without the presence of a thrombus along with venous occlusion caused by proliferation of the smooth muscles in the tunica intima of the veins [1]. Clinically and endoscopically, IMHMV is usually misdiagnosed as an inflammatory condition of the bowel, similar to Inflammatory Bowel Disease (IBD) with same clinical features [2]. Therefore, definitive diagnosis is possible only after surgery as biopsy is not capable of distinguishing the ischemic abnormalities from those associated with e known IBD manifestations [3]. Idiopathic myointimal hyperplasia of the mesenteric veins mostly involves the thickening of small and medium-sized mesenteric veins with the hallmark manifestation of intimal smooth muscle proliferation resulting in luminal occlusion and mucosal ischemic changes [3].

Vascular modifications involve clustered, dilated or arterialized capillaries, sub-endothelial fibrin deposits, and many fibrin thrombi in ulcerated and non-ulcerated mucosae [4]. The first case of IMHMV was described in the medical literature in 1991 by Genta and Haggit, who described 4 male patients with segmental ischemic colitis due to idiopathic myointimal hyperplasia in the small mesenteric veins and their intramural branches [5]. Since then, few cases have been reported describing this entity with different clinical presentations, however, the etiology and pathogenesis of these vascular modifications remain

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https://doi.org/10.1016/j.ijscr.2021.106022
Received 26 April 2021; Received in revised form 22 May 2021; Accepted 23 May 2021
Available online 26 May 2021

unclear. Majority of these cases ultimately end up with different degrees of bowel resection [4]. Usually, mistaken for inflammatory bowel disease, the means of definitive diagnosis for bowel ischemia and venous thrombotic disease are the pathological changes taking place, as preoperative radiological and clinical methods fail to distinguish it from these bowel and systemic diseases. To the best of our knowledge, roughly 34 cases of IMHMV have been reported in the literature with this being the 35th case.

We herein describe the case of a male patient with idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV) with spontaneous double perforation of the colon (descending colon and sigmoid colon) associated with a year long history of what was thought to be inflammatory bowel disease, Ulcerative colitis (IBD).

This case report has been reported in accordance with the SCARE Criteria [6].

2. Case presentation

A 55-year-old male patient with a history of chronic abdominal pain was admitted to the hospital with chief complaints of severe abdominal pain for the past 10 days. The pain was continuous in nature and persisted throughout the day. There was history of loss of appetite, weight loss of nearly 4 kg over a period of 1 week and passing blood-stained stools occasionally. There was no history of any joint pain, skin lesions, fever or conjunctivitis, chest pain, cough and contact with a COVID-19 case. He is a known case of type −2 diabetes mellitus on oral hypoglycemic medications. Approximately 15 years back, he was admitted for left-sided lower limb deep vein thrombosis (DVT) associated with pulmonary embolism for which he was on warfarin for a year. His brother had a history of esophageal carcinoma.

The physical examination showed a non-distended, non-tender abdomen. However, the bowel sounds were exaggerated. Vital parameters were normal. Following Laboratory findings were reported: Increased Inflammatory markers with elevated, CRP = 179.07, WBC = 21.68, Hg = 12.50, platelets = 287.00 and neutrophil = 19.67. Fecal calprotectin was 877.8 μg/g.

Colonoscopy was performed, which revealed a congested and inflamed rectal mucosa (Fig. 1). Salt and pepper appearance was seen in the sigmoid region and the mucosa bled on touch. The transverse colon showed a whitish appearance with loss of vasculature. Biopsy findings showed mixed inflammatory cells in the lamina propria and the cells were negative for granuloma, dysplasia or malignancy, and there was no evidence of crypt abscess and distortion. None of the findings were specific for Ulcerative colitis.

Followed by the colonoscopy, computed tomography (CT) scan of the abdomen and the pelvis was done with suspicion of complications. However, CT findings revealed diffuse circumferential thickening of the sigmoid and descending colon associated with pericolic and pelvic stranding reaction that was suggestive of infective/inflammatory left-sided colitis. Based on the presentation along with the investigations, he was diagnosed to be a suspected case of inflammatory bowel disease (IBD) and was started on prednisolone 30 mg once daily, Adalimumab, oral 5-Amino salicylic acid, intravenous broad-spectrum antibiotics and then later on he was discharged. Two weeks post-discharge, he had complaints of fever, constipation, decreased oral intake along with lower right and left quadrant abdominal pain of 8/10 severity without vomiting or diarrhea. He was treated as an outpatient and was scheduled for day-case Flexible sigmoidoscopy which revealed severely inflamed mucosa with pseudo polyps that caused significant narrowing of the lumen (Fig. 2). Multiple biopsies were taken to rule out dysplasia.

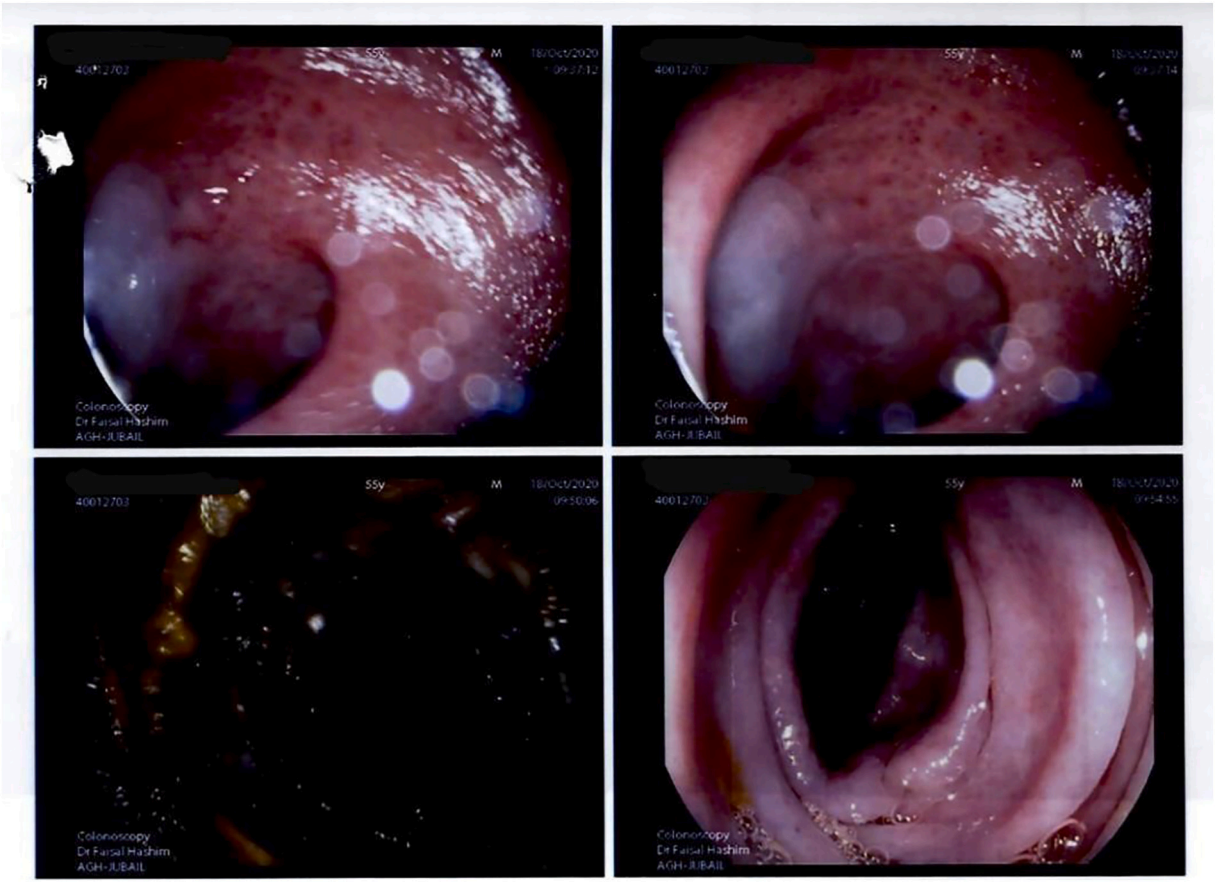

Fig. 1. Colonoscopy images showing congested mucosa in sigmoid (Top-left) and whitish appearance indicating loss of vasculature in transverse colon suggestive of IBD.

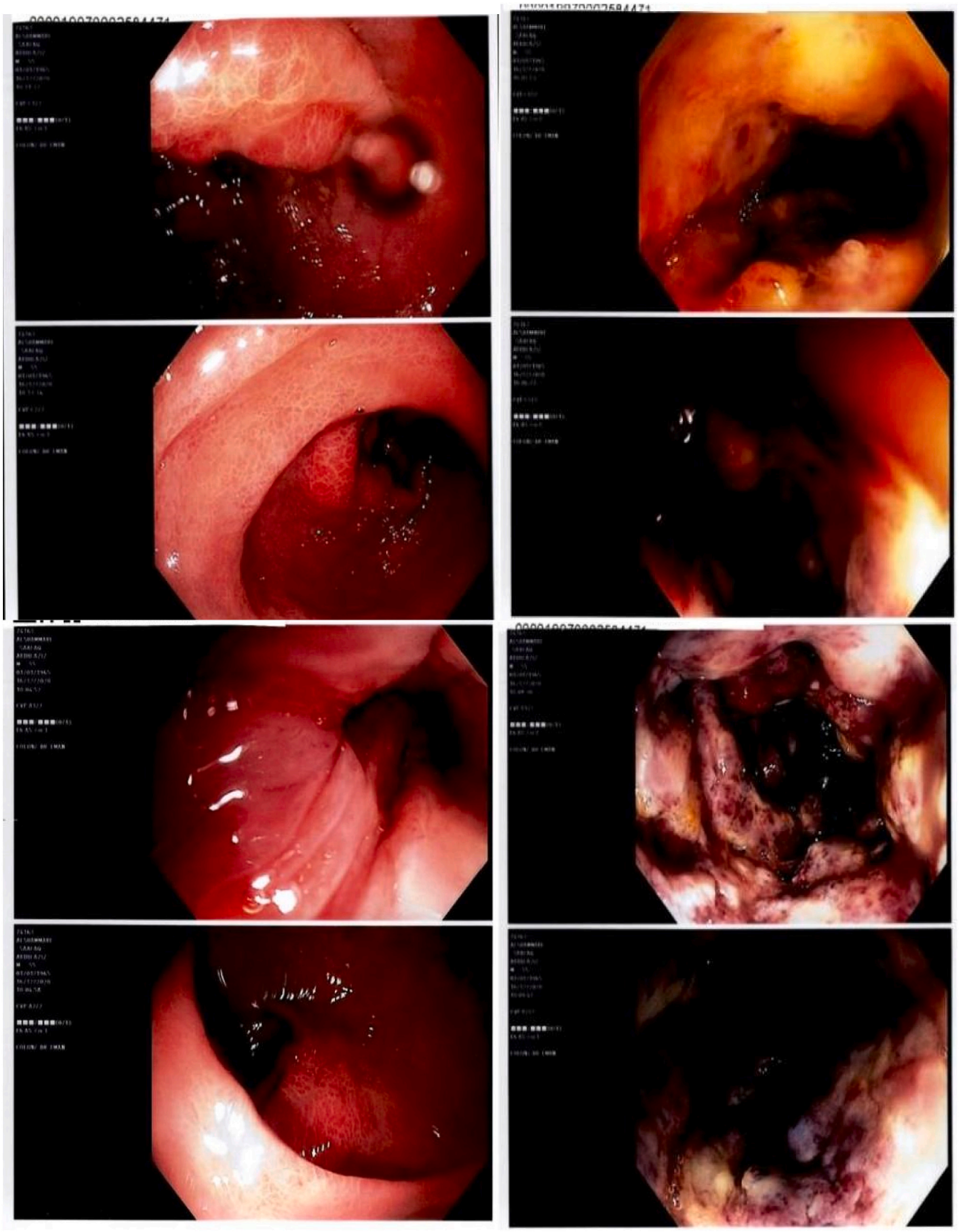

Fig. 2. Flexible sigmoidoscopy images showing severely inflamed mucosa (Top-left) with pseudo-polyps (top-right) with significant narrowing of the lumen (bottom-left and -right).

Almost 2 months later, the patient got readmitted to the hospital with severe abdominal pain and fever for 2 days associated with a weight loss of around 12 kg in 2 months. The patient's appearance was uncomfortable with normal vital parameters. Physical examination of the abdomen showed a non-distended mild tenderness. Laboratory reports were WBC = 13.37, NEU = 78.30, hemoglobin = 13.90 and C - reactive protein = 179.07. He was admitted and started on intravenous antibiotics and intravenous hydrocortisone as flare-up of the underlying disease was suspected. Over the next 3 days, the patient failed to show any significant improvement along with a proportionate increase in the abdominal pain, watery blood-stained stool, tachycardia and hypotension. However, there was no fever. To rule out any new bowel-related complications, contrast-enhanced computed tomography (CECT) was performed which had similar findings of previous CT and there was no evidence of bowel perforation. Blood works and *Clostridium difficile* toxin screen was done and was positive (Fig. 3).

It was decided to follow him with a conservative management approach, but the symptoms never subsided. Rather abdominal pain got aggravated and the abdomen was distended two days later. Tenderness was present all over the abdomen with rebound tenderness and an X-ray was done which showed free air under the diaphragm. CT scan was repeated to re-evaluate the presence of any perforation and the findings demonstrated circumferential thickening of the descending colon with a small air locule traversing the wall of the lower part of descending colon

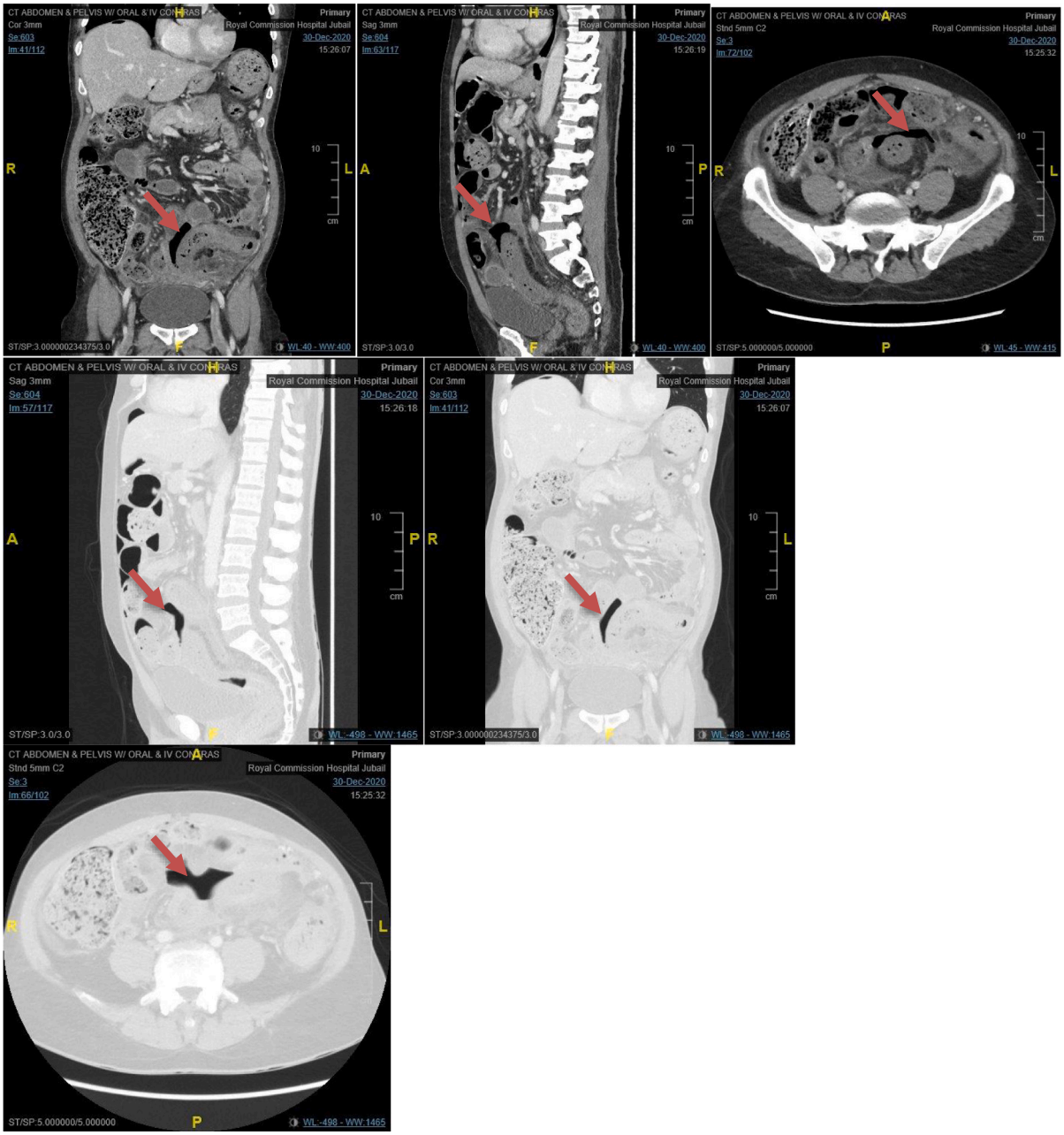

Fig. 3. Contrast enhanced computed tomography of the abdomen, abdominal and lung windows are shown. Red arrows denote the areas of pneumoperitoneum.
Operation report:
Intraoperative findings:
Severe colitis with a scattered area of dusky patches along with the bowel, extending from the upper rectum to mid of transverse colon with perforations at 2 sites (one in sigmoid and another one in descending colon below the splenic flexure). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

and a large air pocket adjacent to sigmoid colon depicting the perforated bowels. It was decided to proceed with a left colectomy and Hartman's procedure (Fig. 4).

Left hemicolectomy and Hartman's procedure were performed. Surgical specimens from mid of transverse colon, descending colon, and sigmoid up to mid rectum were sent for examination.

Histopathological findings showed colon segment with a concentric proliferation of smooth muscle cells in the intima and media of small to medium-sized intramural veins. The arteries appeared normal. The mucosa showed dilated capillaries, focal small muscular vessels with thickened hyaline walls, and extensive ischemic changes with ulceration. No evidence of thrombosis or malignancy. The features were consistent with venous insufficiency due to idiopathic myointimal hyperplasia. And the final diagnosis was confirmed as Idiopathic myointimal hyperplasia of mesenteric veins (IMHMV) with perforation.

3. Discussion

Idiopathic myointimal hyperplasia is a rare cause of intestinal ischemia. As compared to other inflammatory conditions of the gastrointestinal tract, idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV) is not caused by arterial thromboembolism, venous thrombus or vasculitis such that its etiology remains poorly understood. Given that the case presented with double perforation in the colon with a year long history of presumed ulcerative colitis (UC) and the similarity of clinical manifestations of idiopathic myointimal hyperplasia of mesenteric veins and ulcerative colitis, the histological manifestations were completely distinct and dissimilar. The former often results in mucosal modifications due to non-specific ischemic injury than an immune-mediated inflammatory process with the absence of plasma cell-rich inflammation [4].

Evidence showed that most of the affected cases are middle-aged, previously healthy males without prior gastrointestinal complaints but presenting with recurrent, progressive abdominal pain accompanied by bloody diarrhea. Similar findings were encountered in this case [4]. Symptoms start with mild cramping pain and loose stools that progress over a variable duration (months or years), and finally result in a severe recurrent pain with hematochezia which was the presentation in our patient [4]. The non-specific presenting symptoms include bloody diarrhea, lower abdominal pain and weight loss resulting in misdiagnosis of ulcerative colitis [7]. Our patient had a history of deep vein thrombosis associated with pulmonary embolism which is explained by previous studies reporting cardiovascular risk factors commonly seen in many cases [2,7–9].

Although majority of the cases involve the recto-sigmoid colon, extension of the disease proximally along with the descending colon to involve the jejunum, ileum, and entire colon sparing the rectum is also seen [3]. In the present case, the mucosa showed dilated capillaries, focal small muscular vessels with thickened hyaline walls and extensive ischemic changes with ulceration. Evidence showed that idiopathic myointimal hyperplasia of mesenteric veins can also affect the mucosa. This seems to be characteristic of the disease where veins in the colonic submucosa and muscularis propria are also involved [5]. A study conducted by Abu Alfa and his colleagues on a patient with idiopathic myointimal hyperplasia of mesenteric veins showed that biopsy analysis revealed the presence of thick-walled hyalinized vessels [10].

Other recent studies have also reported In accordance with their findings, we also noted the presence of focal small muscular vessels with thickened hyaline walls.

Characteristic manifestations include radiographically evident structures of the distal colorectum combined to mucosal friability, ulcers, and pseudo polyps similar to the features of the inflammatory

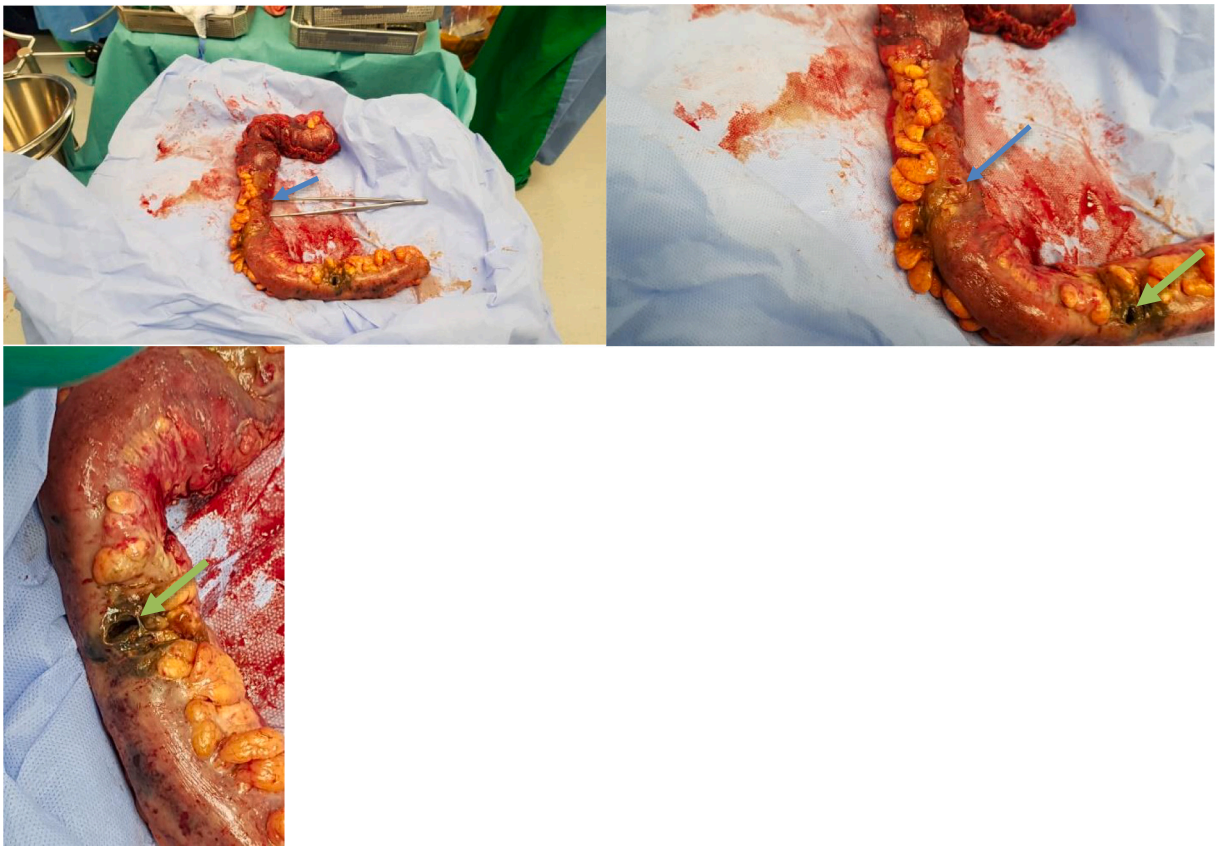

Fig. 4. Intra-operative findings: Blue arrow: descending colon perforation, Green arrow, Sigmoid colon perforation. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

bowel disease following which the patient was treated with corticosteroid therapy prior to definitive surgical management.

Absence of concrete histopathological criteria for definitive diagnosis of IMHMV makes the biopsy-based diagnosis a challenge for the pathologists resulting in early initiation of treatment with anti-inflammatory drugs rather than surgery, which at present is the only effective treatment for this condition that has been reported to be completely curative. Hence, those who underwent surgery are predisposed to a higher risk of developing intestinal perforation, specifically if they received corticosteroid therapy for a presumptive diagnosis of ulcerative colitis [4,5]. This might possibly explain the double perforation in the colon that was reported after a year long history of inflammatory bowel disease (IBD).

4. Conclusion

The present case report describes a male patient with an Idiopathic myointimal hyperplasia of mesenteric veins (IMHMV) with perforation with the presence of mesenteric veins in the mucosa. It must be noted that treating physicians must consider the possibility of idiopathic myointimal hyperplasia of mesenteric veins when similar manifestations are encountered in biopsy specimens of old cases with suspected inflammatory bowel disease or non-occlusive ischemia of the distal colorectum.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Informed consent

The study participant was at negligible risk. Care was taken to ensure no breach in the confidentiality of data. Written informed consent was obtained from the patient for publication of this Case Report and for all the accompanying images and videos. A copy of the written informed consent is available for review by the Editor of this journal. Dr. Ahmed T Almumtin, as a corresponding author and part of the case management team.

Ethical approval

The case is set to be published in full accordance with all the applicable policies and procedures of National Committee on Bio-Ethics Regulations and Royal Commission health services along with all the applicable Saudi laws and regulations including National Committee on Bio-Ethics Regulations and National Committee on Bio-Ethics Regulations and Royal Commission health services. Reference number of the committee is 2021-012/04/CR-Surg.

Source of fundings

No funding was received/required for this case report.

Author contribution

Ahmed T Almumtin: Main author: Study concept and design, data collection, data analysis and interpretation, writing the paper.

Guarantor

Dr. Ahmed Almumtin.

Research registration number

Not applicable.

Declaration of competing interest

No conflicts of interests or disclosures.

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