EDORIUM Journals

Int J Case Rep Images 2023;14(2):17–20.
www.ijcasereportsandimages.com

CASE REPORT PEER REVIEWED | OPEN ACCESS

Idiopathic myointimal hyperplasia: An unusual cause of colonic ischemia

Samantha Phillips, Ranah Lim, Corina Behrenbruch, Julien Schulberg, Penelope McKelvie, Michael Johnston

ABSTRACT

Introduction: Ischemic colitis is a commonly encountered surgical condition with a variety of causes, including atrial fibrillation, atherosclerosis, hypoperfusion, vasculitis, and rarely drug causes such as methamphetamines.

Case Report: Here we describe the case of a 47-year-old male who presented with non-specific abdominal symptoms. He was initially treated for severe ulcerative colitis (UC), before biopsies were re-reviewed and specimens showed changes pathognomic for idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV). He underwent surgical resection of affected segments and recovered well.

Conclusion: This case illustrates a rare diagnosis presenting with common symptoms and the importance of considering this diagnosis in patients not responding to usual treatment.

Keywords: Idiopathic myointimal hyperplasia of the mesenteric veins, Inflammatory bowel disease, Ischemic colitis, Mesenteric inflammatory veno-occlusive disease

Samantha Phillips[1], FRACGP, Ranah Lim[1], FRACS, Corina Behrenbruch[1,2], Julien Schulberg[3], FRACP, PhD, Penelope McKelvie[4], FRCPA, Michael Johnston[1], FRACS

Affiliations: [1]Colorectal Unit, St. Vincent's Hospital, Melbourne, Australia; [2]University of Melbourne, St. Vincent's Hospital, Melbourne, Australia; [3]Gastroenterology Department, St. Vincent's Hospital, Melbourne, Australia; [4]Anatomical Pathology Department, St. Vincent's Hospital, Melbourne, Australia.

Corresponding Author: Dr. Samantha Phillips, St. Vincent's Hospital, Melbourne, 41 Victoria Parade, Fitzroy, VIC 3065, Australia; Email: samantha.phillips@svha.org.au

Received: 25 April 2023
Accepted: 26 June 2023
Published: 27 July 2023

How to cite this article

Phillips S, Lim R, Behrenbruch C, Schulberg J, McKelvie P, Johnston M. Idiopathic myointimal hyperplasia: An unusual cause of colonic ischemia. Int J Case Rep Images 2023;14(2):17–20.

Article ID: 101403Z01SP2023

****

doi: 10.5348/101403Z01SP2023CR

INTRODUCTION

Ischemic colitis is a common surgical condition. Potential causes include atrial fibrillation, atherosclerosis, hypoperfusion due to cardiac failure or sepsis, vasculitis and rarely drugs such as methamphetamine or cocaine [1]. Here we present an unusual case of left sided colitis due to idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV).

CASE REPORT

A 47-year-old male presented with a four-month history of progressively worsening generalized abdominal pain and diarrhea. His past medical history included atrial fibrillation on anticoagulation, epilepsy, hypercholestrolemia, hypertension, and a splenectomy at the age of 4 years for idiopathic thrombocytopenic purpura. He is self-employed, an ex-smoker with a 20-pack year history and consumed 35 units of alcohol per week.

The patient initially had two weeks of non-specific abdominal pain for which he was admitted and discharged for outpatient investigation. A computed tomography (CT) scan demonstrated a 20 cm segment of colitis

involving the descending colon. He was commenced on intravenous (IV) antibiotics for a presumed infectious cause and his symptoms improved markedly. He was discharged the following day and planned for an urgent outpatient colonoscopy.

Two weeks after discharge the patient presented to hospital a second time with ongoing abdominal pain and new rectal bleeding with mucus. His repeat CT abdomen demonstrated worsening of the descending colon colitis. A flexible sigmoidoscopy revealed only mild endoscopic mucosal changes with an edematous sigmoid colon with loss of vascular pattern (Figure 1A). Biopsies reported mild capillary dilatation. Treatment for presumed ulcerative colitis (UC) was commenced, initially IV hydrocortisone and then a weaning dose of oral prednisolone on discharge.

Just over a month later, after four months of symptoms, the patient re-presented to Emergency Department with very severe abdominal pain associated with frequent bloody and bowel motions with mucus 10–15 times per day, nausea, vomiting, anorexia, and weight loss. His vital signs were within normal range, the white cell count normal and c-reactive protein (CRP) 44. Computed tomography abdomen demonstrated colitis from the splenic flexure to the rectum (Figure 2) and a repeat flexible sigmoidoscopy confirmed severe inflammation with spontaneous bleeding, mucosal erythema, and loss of vascular pattern (Figure 1B). Treatment for severe UC was initiated with IV hydrocortisone and later infliximab.

Unfortunately, the patient did not respond IV steroids or infliximab. The colonic mucosal biopsies were re-reviewed by pathology and showed changes of ischemic colitis with subendothelial fibrin rings, fibrin rings, and arterialized capillaries which are pathognomonic mucosa changes for IMHMV (Figure 3A). An emergency Hartmann's procedure was performed, with a matured mucous fistula. Intra-operatively, gross edema, and congestion of the left colon down to the anus was noted, with a densely fibrotic colonic mesentery and mesorectum with fat necrosis (Figure 4). The patient recovered well despite a short post-operative ileus and a small intra-peritoneal collection, which was drained radiologically. Histopathology of the resection specimen confirmed prominent myointimal thickening of the mesenteric veins with narrowing of the lumen which is typical for IMHMV (Figure 3B).

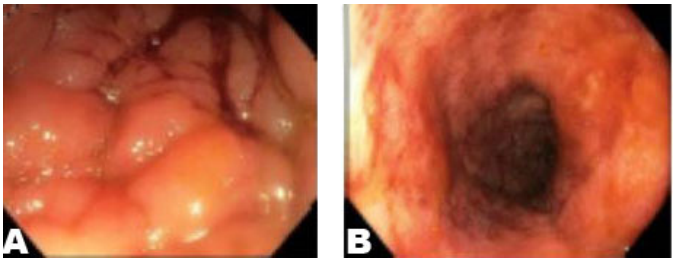

Figure 1: (A) Comparison of the initial flexible sigmoidoscopy (left) and (B) the repeat flexible sigmoidoscopy approximately one month later (right).

After a period of recover the patient's stoma was closed and intestinal continuity restored. There was no residual IMHMV found in the same taken from the stoma and proximal rectum, suggesting a period of diversion improved the condition. The patient had no further abdominal symptoms.

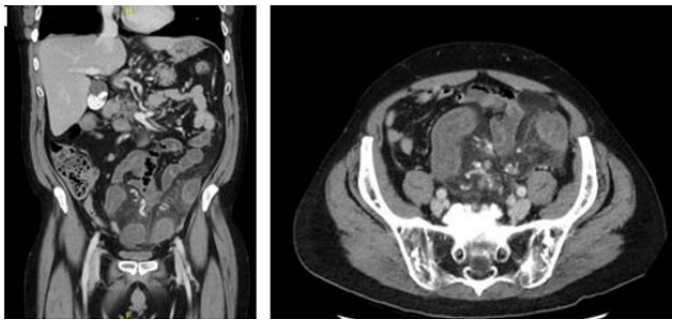

Figure 2: CT abdomen demonstrating colitis from splenic flexure to rectum.

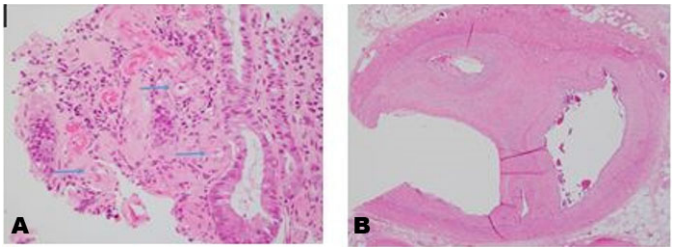

Figure 3: (A) Endoscopic biopsies showing arterialized capillaries, and subendothelial fibrin rings. (B) Mesenteric vein (from the resection specimen showing markedly thickened intima).

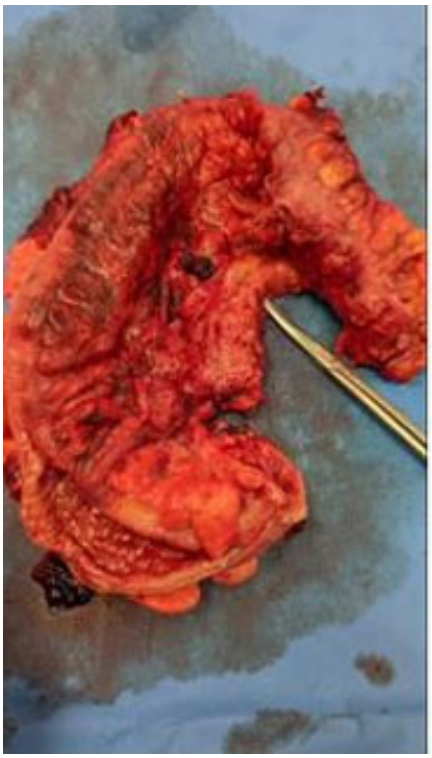

Figure 4: Intra-operative specimen.

DISCUSSION

Idiopathic myointimal hyperplasia of the mesenteric veins is exceedingly rare and defined as a "non-thrombotic, non-inflammatory occlusion" of the mesenteric veins [2]. The etiology is unknown. However, the characteristic histopathologic findings are venous intimal smooth muscle hyperplasia with arterial sparing [2, 3]. Idiopathic myointimal hyperplasia of the mesenteric veins was first described in 1991 by Genta and Haggitt and since then, 70 cases have been reported in the literature [4]. Although the pathogenesis is unknown, there are two proposed theories. The first is that intermittent volvulus of the sigmoid colon results in arteriovenous fistula formation, and the second is that IMHMV is a part of the spectrum of enterocolic lymphocytic colitis [4].

The clinical presentation of IMHMV is similar to other causes of colitis; hence, the initial presentation can often be confused UC [4]. Almost all of patients discussed in the literature progress to surgery due to failure of medical management or the development of complications. Idiopathic myointimal hyperplasia of the mesenteric veins most commonly affects middle aged men and usually the left colon; however, there are reports of involvement of descending colon, rectum, and entire colon [5].

The diagnosis of IMHMV in the majority of cases is made on pathology review of the operative specimen as the colonoscopic and CT findings are essentially indistinguishable from other forms of colitis. In this case, the pathologist was able to identify pathognomonic features of endoscopic biopsies.

CONCLUSION

There is no medical management of this condition, with surgical resection of the affected segment of the bowel the only curative treatment. Although rare, IMHMV is an important differential diagnosis to consider in patients with colitis not responding to usual medical therapy.

REFERENCES

1. Theodoropoulou A, Koutroubakis IE. Ischemic colitis: Clinical practice in diagnosis and treatment. World J Gastroenterol 2008;14(48):7302–8.
2. Platz J, Hyman N. Idiopathic myointimal hyperplasia of mesenteric veins. Gastroenterol Hepatol (N Y) 2012;8(10):700–2.
3. Martin FC, Yang LS, Fehily SR, D'Souza B, Lim A, McKelvie PA. Idiopathic myointimal hyperplasia of the mesenteric veins: Case report and review of the literature. JGH Open 2020;4(3):345–50.
4. Li H, Shu H, Zhang H, Cui M, Gao Y, Tian F. 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) 2022;9:855335.
5. Al Ansari A, Ahmed S, Mansour E, Abass MA. Idiopathic myointimal hyperplasia of the mesenteric veins. J Surg Case Rep 2021;2021(1):rjaa453.

****

Author Contributions

Samantha Phillips – Design of the work, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Ranah Lim – Conception of the work, Design of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Corina Behrenbruch – Conception of the work, Design of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Julien Schulberg – Conception of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Penelope McKelvie – Conception of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Michael Johnston – Conception of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None.

Consent Statement

Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.

Copyright