Case Report

Small Bowel Ischemic Necrosis Secondary to Idiopathic Intimal and Medial Hyperplasia of Mesenteric Vessels: A Case Report

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Small bowel ischemic necrosis secondary to idiopathic intimal and medial hyperplasia of mesenteric small vessels is rare. The mesenteric vascular diseases were recently classified as two vascular diseases: fibromuscular dysplasia (FMD) of artery and mesenteric arteriovenous dysplasia/vasculopathy (MAVD/V). FMD usually involves medium size mesenteric arteries in younger individuals. In contrast, MAVD/V tends to affect multiple small mesenteric arteries and veins without vascular lesions in other organs. We reported that a 54-year-old female presented to emergency room with acute onset abdominal pain, nausea and vomiting. CT angiography showed "string of beads" in mesenteric vessels, but did not reveal any vascular narrowing elsewhere in the full body scan. The patient had small bowel ischemic necrosis secondary to idiopathic intimal and medial concentric smooth muscle hyperplasia in mesenteric vessels. The overall picture is consistent with MAVD/V.

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INTRODUCTION

Fibromuscular dysplasia (FMD) is an uncommon, non-inflammatory arteriopathy that most commonly affects the renal and internal carotid arteries, but has been reported to affect nearly every arterial bed in the body.[1,2] FMD is categorized into intimal, medial and adventitial type and also classified as local and multifocal disease. In the United States registry of FMD, however, only 8 cases were reported to cause mesenteric ischemia.[1] More mesenteric FMD cases in children and adults were also reported.[3-14] Recently, Patil and colleague reported 11 cases with non-inflammatory, non-atherosclerotic mesenteric arteriovenous dysplasia/vasculopathy (MSVD/V) that is distinct from typical FMD.[15] They set up three criteria for MSVD/V:(1) concentric/eccentric smooth muscle collarette around the tunica media of both the artery and the vein in ≥2 foci, (2) varying degrees of intimal and medial hyperplasia and adventitial fibrosis, and (3) lack of inflammation or thrombi.[15] Only 2 of 11 cases showed small bowel transmural ischemic necrosis. Here we reported one case with acute transmural ischemic necrosis in whole small bowel secondary to idiopathic intimal and medial hyperplasia in small mesenteric arteries and veins, but the patient does not have history of Crohn's disease, mass lesions, operation and heart attack.

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CASE REPORT

A 54-year-old female had acute onset abdominal pain, nausea and vomiting. Her past medical history was significant for anxiety for which she was being treated with sertraline (Zoloft). She was a former smoker (30 pack years). She did not use oral contraception pills (OCPs) and had no known prior history of venous thromboembolism. On presentation her blood lactate level was 4.9 with normal WBC count. Imaging (CT angiography) was read as bowel ischemia of ileal loops with intraluminal hemorrhage and beaded appearance of the mesenteric vessels, suggesting a possible vasculitis **(Figure 1, A and B)**. She was originally managed with NG tube for decompression and antibiotic coverage in addition to intravenous fluids. Coagulation profile was not concerning for a hypercoagulable state. Complete lab work up was normal and ANA screen was negative. A thorough evaluation for thrombosis by imaging and laboratory work was negative for thromboembolism. Due to worsening pain, she was taken to operating room for an exploratory laparotomy resulting in small bowel resection with primary anastomosis. About 120 cm of the small bowel was resected and the small bowel necrosis was felt to be in an arterial distribution. However no thrombosis was appreciated in gross examination.

Gross examination revealed a segment of small bowel with ischemic necrosis (120 cm in length x 2 cm in diameter). The specimen was opened to reveal almost entire intestinal wall with intense dark purple hemorrhagic mucosa that was sharply demarcated from the adjacent viable pink appearing normal mucosa **(Figure 1, C and D)**. The small bowel and mesenteric vessels were extensively sampled for microscopic examination. Microscopic examination revealed small bowel with transmural hemorrhage, inflammation, edema, ulceration, mucosal necrosis, consistent with acute ischemic change **(Figure 2, A)**. Focal mesenteric small arteries and vein were narrowed with intimal thickening and smooth muscle hyperplasia (**Figure 2, B-E**). No inflammation or any lipid accumulation was identified. Smooth muscle and collagen hyperplasia was mixed together in intimal and medial layers **(Figure 2, B-E)**. A few mesenteric small arteries had concentric smooth muscle collarette around the tunica media of the artery **(Figure 2, F-I)**. The trichrome, elastin special stains, and smooth muscle actin (SMA) immunohistochemical stains highlighted extensive smooth muscle hyperplasia **(Figure 2, G-I)**. There was complete absence of inflammatory cells or post-inflammatory scarring together with the architectural preservation of the vascular wall. Based on the small bowel ischemic change and small mesenteric arteriovenous wall hyperplasia, the diagnosis of mesenteric arteriovenous dysplasia/vasculopathy was rendered.

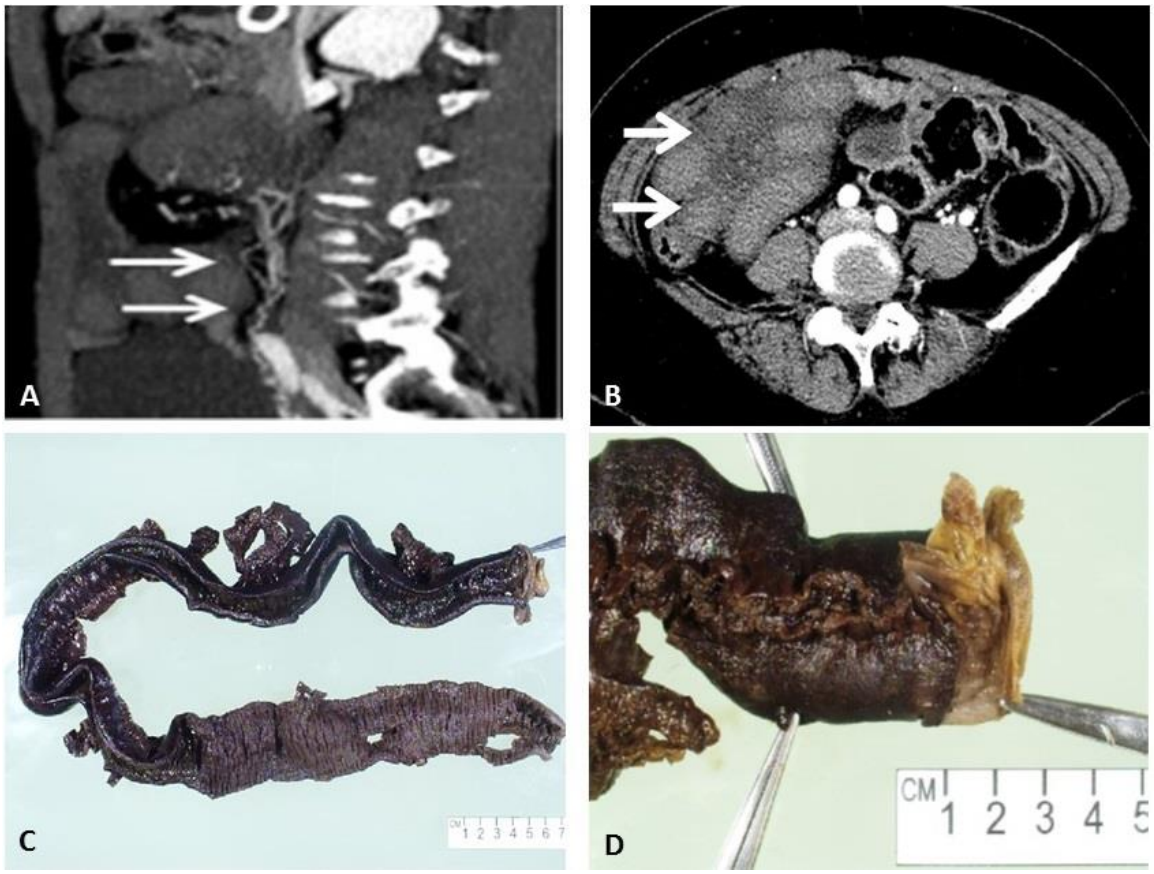

Figure 1. (A) CT angiography sagittal section shows beaded appearance of the mesenteric vessels. **(B)** CT scan (Axial section) shows thickened edematous small bowel loops suggesting ischemic changes (horizontal arrows). **(C)** The ischemic necrosis of small bowel is dark and congested; **(D)** Ischemic necrosis of small bowel sharply demarcated from adjacent viable bowel.

DISCUSSION

We report one case with acute whole small bowel ischemic necrosis secondary to small mesenteric vessels narrowing and occlusion. Idiopathic intimal and medial hyperplasia was present in small mesenteric arteries and veins. There was complete absence of inflammatory cells or post inflammatory scarring together with the architectural preservation of the vascular wall. This excludes any possibility of a vasculitis or a healed previous inflammatory lesion. The small bowel ischemic necrosis is not rare disease. However, the small bowel ischemic necrosis secondary to idiopathic intimal or medial hyperplasia of the small arteries and veins is rarely reported.

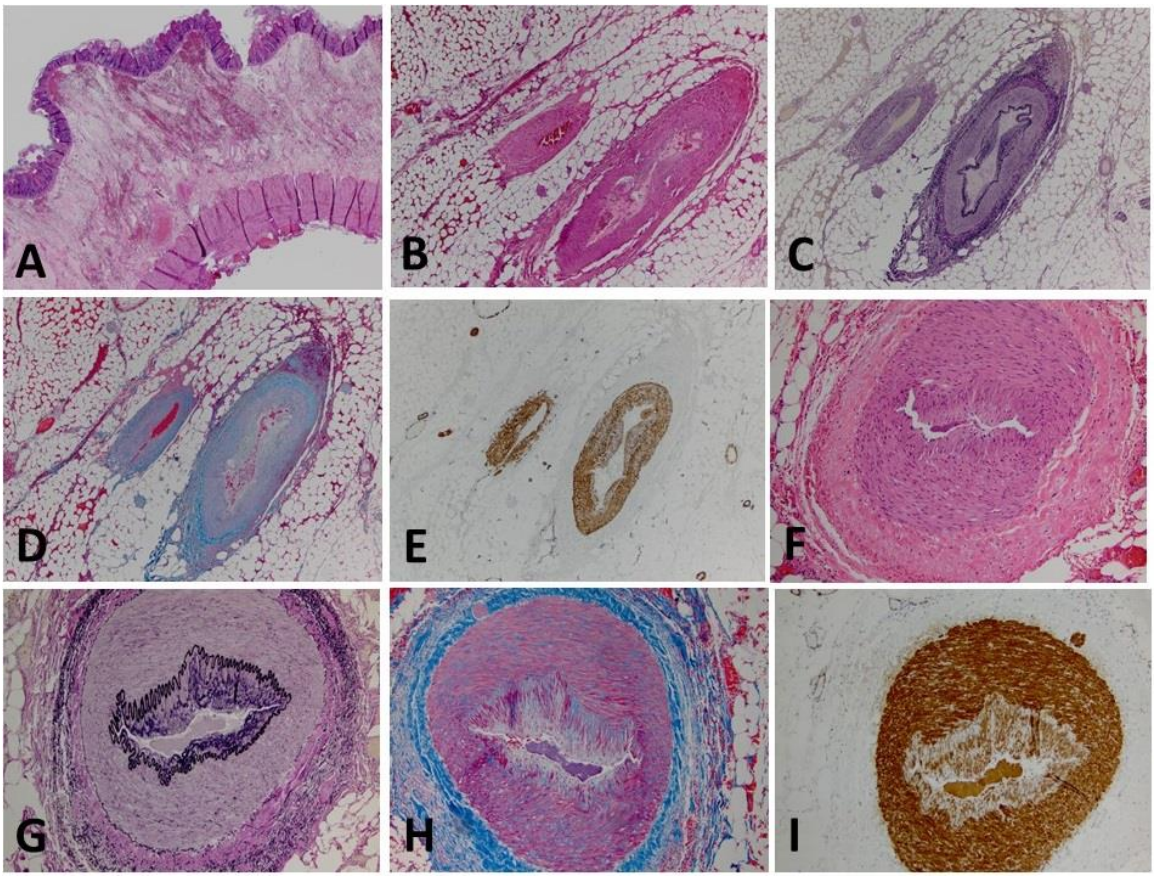

Figure 2. Histology of small bowel and vessels. **(A)** Transmural ischemic necrosis in small bowel; Small artery and vein showed intimal hyperplasia and lumen narrowing with H&E stain **(B)**, elastin stain **(C)**, trichrome stain **(D)** and smooth muscle actin immunostain **(E)**. Small artery with concentric medial smooth muscle hyperplasia and lumen narrowing with H&E stain **(F)**, elastin stain **(G)**, trichrome stain **(H)** and smooth muscle actin immunostain **(I)**.

The terminology of mesenteric vascular diseases is not very clear. FMD has been reported previously to involve focal and multiple organs.[1,2] Focal disease is usually caused by intimal hyperplasia; multifocal disease manifests with the classic "string of beads" appearance and manifests as vascular stenosis, aneurysms or dissections in medium-sized arteries.[1] The histological characteristics include the deposition of collagen fibers amidst degenerating elastic fibrils involving the intimal, medial, or adventitial compartments of the artery and the medial smooth muscle hyperplasia.[16,17] In the United States FMD registration, only 8 cases had mesenteric ischemia in adults, but no child was reported for mesenteric ischemia.[1] Recently, Patil et al. reported a group of distinct MAVD/V, which is characterized by (1) concentric/eccentric smooth muscle collarette around the tunica media of both the artery and the vein in ≥2 foci, (2) varying degrees of intimal and medial hyperplasia and adventitial fibrosis, and (3) lack of inflammation or thrombi.[15] FMD usually involves in medium size mesenteric arteries such as superior mesenteric and celiac arteries in younger individuals. The majority of patients show features of ischemic colitis with "beads of string" in CT angiography. In contrast, MAVD/V involves multiple small mesenteric arteries and veins without vascular lesions in other organs and may or may not have vascular change on angiography. Our case showed "string of beads" in mesenteric vessels by CT angiography, but did not reveal vascular narrowing elsewhere in the full body scan. In addition, both small arteries and veins were involved in attached mesenteric adipose tissue and some small arteries showed concentric smooth muscle hyperplasia **(Figure 2, F-I)**. The overall picture is suggestive of MAVD/V. However, our case showed almost whole small bowel acute ischemic necrosis and "string of beads" in mesenteric vessels, which is difficult to interpret by focal small vascular occlusion. In addition, most of MAVD/V cases had Crohn's disease (45%) or mass lesions (27%). Seven patients had a previous history of surgery and 4 patients with history of cardiac disease. Our patient is a 54-year-old female and a smoker without history of chronic gastrointestinal condition or a mass. She did not use oral contraception pills and did not have deep vein thrombosis. Our patient presented to emergency room with acute onset abdominal pain, nausea and vomiting. Only 2 of 11 previously reported MAVD/V cases had acute transmural ischemic change similar to our case.

At present, the mechanism of vascular change in both FMD patients and MAVD/V is unclear. Cigarette smoking, increased estrogen levels, mechanical trauma, and genetic factors have been proposed for the pathogenesis of FMD.[18-22] From 11 MAVD/V cases, it does appear to be more common in women, none of the subjects in their cohort has a history of long-term oral contraceptive pill or hormone supplement intake. It is less likely that nonsteroidal anti-inflammatory drug use (2/11 patients), smoking history (4/11 patients), or medications alone could contribute to these vascular abnormalities. Although 7/11 patients had history of surgery, but the interval between previous history and vessel-related intestinal resection is variable from 5 months to 13 years.[15]

In conclusion, we reported a unique case that fits the new category of mesenteric arteriovenous dysplasia/vasculopathy proposed by Patil and colleague. However, we did find "string of beads" in CT angiography. It suggests the possibility of mixed vascular change in both mesenteric medium-sized arteries and small arteriovenous vessels.

CONFLICT OF INTEREST

Authors declare that they have no conflict of interest.

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