A rare cause of ischemic proctosigmoiditis: idiopathic myointimal hyperplasia of mesenteric veins

Ischemic proctosigmoiditis is an uncommon entity in colon ischemia because it is usually associated with an extensive arterial network. Here we report a case of idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV), a very rare cause of mesenteric ischemia, which typically contributes to proctosigmoiditis.

A 60-year-old man presented with a 1-month history of lower abdominal pain, bloody diarrhea, and weight loss of 10 kg. A first colonoscopy in another hospital reported ulcerative colitis. The patient was treated with mesalazine initially, but with little clinical improvement.

Repeated colonoscopy 2 weeks later showed multiple ulcerations in the sigmoid colon and rectum up to 30 cm from the anal verge. Pathological analysis after endoscopic biopsy reported inflammatory cell infiltration of rectal and colonic mucosa. The patient received steroid therapy, balsalazide, and antibiotics, but his symptoms persisted.

Follow-up colonoscopy 10 days later continued to reveal multiple serpiginous circular ulcers between 30 and 10 cm from the anal verge (Fig. 1). Pathological analysis after endoscopic biopsy showed the presence of thick-walled, medium-sized blood vessels at the base of the ulcer, with mural hyalinization and focal thrombosis. Angiography showed no demonstrable opacification of the inferior mesenteric vein (IMV) (Fig. 2). Finally, a Hartmann’s procedure of the affected part of the rectosigmoid colon was carried out. Grossly, the mucosal surface of the resected specimen showed diffuse ulceration with fibrinopurulent exudates (Fig. 3). Histologically, myointimal hyperplasia of the intramural and extramural mesenteric veins was noticed, with nearly total occlusion of the lumens and without an associated inflammatory infiltrate (Fig. 4).

The postoperative course was uneventful; the patient was free from abdominal cramps and bloody diarrhea. Follow-up colonoscopy 4 months later revealed normal colonic mucosa.

Non-thrombotic occlusion of the mesenteric veins is a rare cause of mesenteric ischemia, and IMHMV is the rarest entity among these causes. Patients with IMHMV usually present with a subacute course of weight loss, lower abdominal pain, diarrhea/constipation, and bloody stools. Clinically, these patients are often diagnosed with idiopathic inflammatory bowel disease (IBD), evidenced by the features under colonoscopy, and treated for IBD without an effective response. Mucosal biopsies, interestingly, show thick
walled vessels, focal ulcerations, or fibrin thrombi in the mucosa and submucosa, which do not suggest the diagnosis of IBD [1]. The mainstay of therapy is resection of the involved segment of colon, after which the prognosis is usually smooth. Extensive myointimal hyperplasia of the mesenteric veins in the submucosa, the adventitia, and the mesocolon – which reduces the lumens of the involved vessels, even to nearly total occlusion – is the pathognomonic feature [1]. To avoid misreading veins as arteries, a special elastin stain (Elastica Van Gieson) may be required [2].

The etiology of IMHMV remains unclear. Secondary arterIALIZATION of the mesenteric veins due to increased intravascular pressure may be the cause of myointimal hyperplasia, because of the close similarity with histological findings in patients with failed saphenous vein coronary grafts and dialysis patients with stenosis of their arteries [1,3]. Although the postoperative course is uneventful in most cases, perforation of the resected colon has occurred in several cases [2,4,5]. The average duration from onset of symptoms to surgery is about 6 months. This indicates the difficulty of diagnosing IMHMV.

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References

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Fig. 4 Histopathological findings for the rectosigmoid resection specimen. a The mesenteric vein (arrow) shows near-total occlusion, marked myointimal hyperplasia, and even focal recanalization, but without an associated inflammatory infiltrate. The accompanying artery is spared (Hematoxylin and eosin stain × 100). b Elastica van Gieson stain (× 200); this special elastin stain distinguishes the artery (arrow) from the vein, and is particularly helpful in the diagnosis of idiopathic myointimal hyperplasia of the mesenteric veins (IMHMV).