Idiopathic Myointimal Hyperplasia of the Mesenteric Veins as a Rare Cause of Chronic Diarrhoea

Idiopatska miointimalna hiperplazija mezenterialnih ven kot vzrok kronične driske

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ABSTRACT

Idiopatic myointimal hyperplasia of the mesenteric veins (IMHMV) is a rare and poorly understood cause of chronic ischemic bowel disease. The pathognomonic findings consist of nonthrombotic and noninflammatory occlusion of the mesenteric veins secondary to intimal smooth muscle hyperplasia. It typically affects older and middle-aged men, who are often previously healthy. The pathogenesis and etiology of IMHMV is unknown. In most patients, the disease affects the rectosigmoid colon and presents with abdominal pain, hematochezia, diarrhoea, and weight loss. It represents a diagnostic challenge for pathologists and clinicians. Colonoscopy biopsy specimens are often non representative or misleading, since they show non specific findings or even suggest other diagnosis. Surgical resection is the treatment of choice. Although preoperative diagnosis of the disease is challenging, there are some mucosal pathological features

IZVLEČEK

Idiopatska miointimalna hiperplazija mezenterialnih ven (IMHMV) je redek in slabo poznan vzrok kronične ishemije črevesja. Gre za netrombotično in nevnetno zaporo mezenterialnih ven, ki je posledica proliferacije gladkih mišic v intimi. Pogosto so bolniki moški srednjih let ali starejši moški brez predhodno znanih bolezni. Etiologija in patogeneza bolezni nista poznani. Najpogosteje prizadene rektosigmo in se kaže z bolečinami v trebuhu, hemohezijo, drisko ter hujšanjem. Ker z endoskopijo neredko pridobimo nereprezentativne biopsije, ki kažejo nespecifične spremembe ali pa sugerirajo drugo diagnozo, predstavlja diagnostični izziv tako za klinike kot patologe, zato je bolezen pogosto pozno diagnosticirana. Zdravimo jo s kirurško resekcijo prizadetega dela črevesja. Pred operacijo in histopatološkim pregledom resektata je diagnozo težko postaviti, vendar poznamo histološke elemente, ki tudi v povrhnjih

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that suggest IMHMV. We report the case of a 64-year-old man with IMHMV as a cause of chronic diarrhoea.

endoskopskih biopsijah dajejo slutiti nanjo. Predstavljamo primer 64-letnega moškega s 15-mesečno anamnezo driske.

CASE REPORT

A 64-year-old male patient with arterial hypertension, type 2 diabetes mellitus, obstructive sleep apnea syndrome, psoriasis, history of transurethral resection of a bladder tumor and umbilical hernioplasty, was admitted to our gastroenterology clinic with chief complaint of chronic diarrhoea persisting for the past 15 months. He complained of multiple loose and watery bowel movements up to 10 times per day, accompanied by abdominal pain, hematochezia and unintentional weight loss of 7 kilograms during the last month before admission.

Colonoscopy, performed 4 months after onset of symptoms, only showed mucosal edema with diffuse loss of vascular pattern in the rectum and descending colon. Histopathological examination of biopsy specimens revealed edema of lamina propria, dilated capillaries in submucosa, with numerous eosinophilic granulocytes focally infiltrating the small vessels wall, suggesting possible systemic small-vessel vasculitis. Due to numerous eosinophilic granulocytes, eosinophilic granulomatosis with polyangiitis was suspected. 6 months after onset of symptoms, he underwent contrast enhanced computed tomography, which demonstrated circumferential, symmetrical wall thickening of sigmoid and descending colon with pericolic fat stranding. He temporarily received mesalazin, which had no effect.

Based on the previous histopathological findings, initial management during hospitalization was aimed at ruling out systemic vasculitis (eosinophilic granulomatosis with polyangiitis) and causes of colonic eosinophilia (parasitic infections). The patient had normal peripheral eosinophil count and no other organ involvement (lungs, kidneys, paranasal sinuses, peripheral nervous system). The laboratory results showed no significant abnormalities, inflammatory markers levels were not significantly elevated. Blood tests for systemic vasculitis (anti-neutrophil cytoplasmic antibody, ANCA) were negative. A repeat colonoscopy was performed, which revealed numerous ulcers in the sigmoid colon (Figure 1), and multiple biopsies were taken. Based on working diagnosis of eosinophilic colitis, the patient was treated with methylprednisolone, and some clinical improvement was observed. Histopathological analysis of biopsy specimen showed the presence of thick-walled hyalinized vessels in lamina propria, with plump endothelial cells and subendothelial fibrin deposits, subintimal edema, and in some cases complete occlusion of the vascular lumen. These findings were consistent with IMHMV. Treatment with methylprednisolone was discontinued.

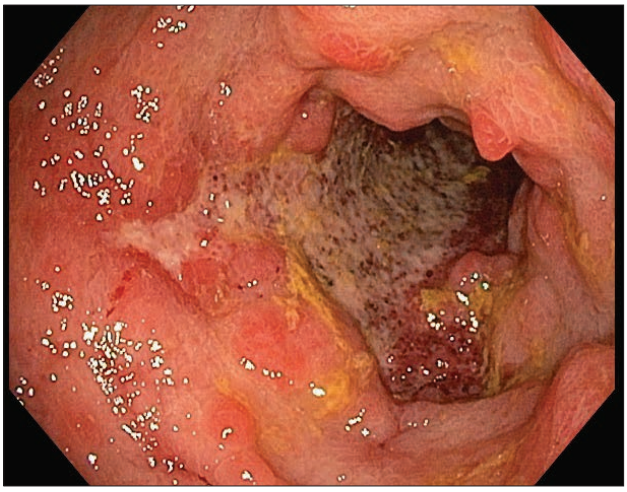

Figure 1. Endoscopic findings of sigmoid colon mucosa showed multiple ulcers

Under the guidance of the scope the bowel was tattooed distally and proximally to the abnormal region. The patient underwent laparoscopic surgical resection of the affected sigmoid segment. Histopathological examination of the resected bowel specimen confirmed the diagnosis of IMHMV. Veins with thickened

and hyperplastic wall, without signs of inflammation, were observed in subserosal fat, muscularis propria and submucosa. The findings involved irregular chronic ischemic colitis with erosions, shallow ulcers and regenerative changes in the mucosa, edema especially of the mucosa and submucosa, steatonecrosis of the pericolic fat and reactive changes in the lymph nodes in the pericolic fat. There were no ischemic changes in the resection margins. During one year follow-up period, there was no evidence of disease recurrence.

DISCUSSION

Etiology

Idiopatic myointimal hyperplasia of the mesenteric veins was first reported in 1991 by Genta and Haggitt. They described 4 patients with segmental ischemic colitis, who recovered completely after segmental resection of the ischemic portion of the colon and had no recurrence of intestinal symptoms on follow-up of up to 7 years (1). Since the histological features of the veins in IMHMV are similar to those of failed cardiac saphenous vein bypass grafts and stenosis of arteriovenous fistula (AVF) in patients undergoing dialysis (2, 3), Genta and Haggitt assumed that the formation of AVFs might increase venous blood flow and result in vascular remodeling that leads to myointimal hyperplasia (1).

Hui Li and his collegues performed a review of the literature from 1991 until February 2022 using electronic databases (Medline, EMBASE, Web of Science, PubMed, and The Cochrane Library – CENTRAL) and found 70 cases of IMHMV. Most patients were previously healthy and no arteriovenous malformation has been identified in the literature (4).

Clinical Features

The most common clinical features are recurrent, progressive abdominal pain (82,9 %), hematochezia (50 %), diarrhoea (37,1 %) and weight loss (18,6 %). Constipation alternating diarrhea (7,1 %) and constipation (7,1 %) were rarely reported. Left colon is affected in most cases, especially rectosigmoid colon, while small intestine, pancolon, ascending colon, transverse colon or only rectum are rarely affected. Clinical presentation depends on site of involvement. Regarding colonic IMHMV, the majority of patients suffer from persistent abdominal pain, hematochezia and diarrhoea. In contrast, small bowel IMHMV mostly presents with obstruction. The analysis of 70 reported cases showed that the meantime between symptom onset and surgery was 4,5 months (4).

Diagnosis

Historically, surgical resection and histopathological findings in resected specimen were considered the only method to diagnose IMHMV. Typical finding is the thickening of small and medium-sized intramural mesenteric veins, with the hallmark manifestation of intima and media smooth muscle proliferation, resulting in luminal occlusion, that leads to mucosal ischemic changes (5). The latter raises the possibility of endoscopic diagnosis. Because of the absence of concrete mucosal histopathological criteria of IMHMV, the preoperative diagnosis is still difficult, but there are some pathological features in endoscopy biopsy specimens reported in the literature that suggest the diagnosis. Those findings include the presence of ischemic pattern of mucosal damage, clustered, slightly dilated, "arteriolized" capillaries with myointimal thickening within the lamina propria, lined by plump endothelial cells and subendothelial fibrin deposits, and thickened submucosal veins (6–8). A hypothesis states that chronic mechanical stress on the mesenteric veins leads to vessel remodeling. The increased venous pressure is transmitted to the mucosal capillaries and causes endothelial injury, which results in fibrin extravasation in subendothelial space (7).

Endoscopic appearance of IMHMV is a result of ischemic changes of the mucosa and is non-specific, contributing to the high rate of misdiagnosis. The most common endoscopic findings are ulcers and mucosal congestion, while strictures are rarely seen (4). Early stages of disease result in edematous and

erythematous mucosa. Ulcers and inflammatory exudates develop with disease progression (9, 10).

Contrast enhanced computed tomography of the abdomen demonstrates the range of intestinal involvement and typically showes features of ischemic colitis: a segment of diffuse circumferential colonic wall thickening, poor mural enhancement, submucosal edema, and pericolic fat stranding. IMHMV is often misdiagnosed as inflammatory bowel disease (11).

Treatment

Surgical resection of the affected bowel is considered the treatment of choice. According to the available literature data, in follow-up duration up to 7 years after resection all patients appear to be cured, with no recurrence of disease-related activity or symptoms. Of the reported cases of IMHMV, Hui Li and his collegues observed complications in 20 out of 70 patients. They were mainly attributed to inflammatory bowel disease-related medication, delay of operation and need for emergent surgery. The most common complications were intestinal perforation and massive hematochezia (4).

CONCLUSION

Idiopatic myointimal hyperplasia of the mesenteric veins should be considered in middle-aged patients with subacute segmental colitis and symptoms characteristic of inflammatory bowel disease or chronic ischemic bowel disease, especially in those who do not improve with therapy. The preoperative diagnosis remains a challenge. Surgical resection of the ischemic portion of the bowel is curative. There is no known evidence of disease recurrence.

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