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Edematous colitis of an unknown cause, diagnosis by pattern recognition.

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Dr. M. Bronswijk was responsible for the daily care of the patient, the initial clinical diagnosis and drafts of the initial manuscript. Prof. G. De Hertogh provided the final histopathological confirmation and revised the manuscript.

Prof. M. Ferrante supervised the patient care and was responsible for the critical revision of the final manuscript.

Conflict of interest statement

All three authors declare no competing interests regarding the publication of this paper.

Question: A 67-year old male, with a history of obesity, type 2 diabetes mellitus and hemochromatosis, was admitted at our tertiary center with a slowly progressive pain in the lower abdomen and fluctuating loose stools. Physical examination only showed progressive tenderness over the left hemi-abdomen. Laboratory evaluation revealed fluctuating C-reactive protein levels between 20 and 34 mg/l (upper limit of normal: 5.0 mg/l), without any signs of malabsorption. Stool samples did not reveal any bacterial or protozoal pathogens.

He was recently evaluated at another hospital where a diagnosis of rectocolitis of an unclear cause was made, after colonoscopy showed narrowing of the colon lumen and edematous mucosae in the descending colon and sigmoid. Following progressive abdominal pain despite a course of corticosteroids, the patient was admitted to our tertiary referral center for further investigation, during which also B-symptoms were noted. Abdominal CT (Computed Tomography) was repeated (Fig.A, right) and compared with the CT scan of the initial presentation at the referring hospital (Fig.A, left). Colonoscopy was also repeated, confirming the presence of colitis with prominent edema (Fig.B), limited to the descending colon, sigmoid and proximal rectum, with subsequent comparative biopsies showing edematous mucosa and capillary congestion.

What is the diagnosis and how should it be treated?

Answer: Idiopathic Myointimal Hyperplasia of the Mesenteric Veins, possibly due to underlying Factor V Leiden.

Revision of the CT showed a progressive wall thickening of the descending colon, sigmoid and proximal rectum, with increasing mesenteric inflammation, absence of the inferior mesenteric vein (IMV) and left sided tortuous mesenteric veins. The triad of a vascular distribution with inflammatory clinical findings and evidence of venous outflow tract problem, led to a strong suspicion of Idiopathic Myointimal Hyperplasia of the Mesenteric Veins (IMH MV).

IMH MV is a rare and poorly understood, non-thrombotic veno-occlusive disease, characterized by intima proliferation in small and medium sized mesenteric veins, leading to protracted rectosigmoiditis in predominantly male patients. Though most cases are diagnosed postoperatively after medical therapy has failed, IMH MV can be detected early on by means of several distinctive findings, preventing unnecessary treatment and diagnostics. Typical findings are progressive left sided colorectal wall thickening or rectosigmoiditis and absence of the IMV on radiological imaging, together with signs of venous stasis and a patent inferior mesenteric artery.¹ Segmental surgical resection remains the preferred treatment, which has been shown to be curative, without any signs of recurrence after 7 years of follow up.²

In our patient the decision for segmental resection was made after multidisciplinary discussion due to worsening abdominal complaints. During surgery a very rigid and fibrotic sigmoid mesocolon was identified (Fig.C), after which a left sided hemicolectomy with a transversorectal anastomosis was performed. In addition to subserosal fatty necrosis, microscopic evaluation of the resection specimen showed mucosal veins with severely reduced lumen due to extensive fibromyxoid intimal hyperplasia (Fig.D), confirming our diagnosis of IMH MV. No signs of vasculitis or thrombosis were seen. After 2 years of follow-up no signs of recurrence have been detected.

Though the exact pathogenesis remains unclear, several possible causes have been proposed, such as local trauma due to sigmoid hypermobility and thrombophilia caused by heterozygosity for Factor V Leiden.³ The latter was also confirmed in our patient, together with a c.665C>T polymorphism in the MTHFR gene, suggesting that thrombophilia and possible subsequent recurrent mesenteric thrombosis could increase risk of IMH MV.

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