Netherlands
The Journal of Medicine

CASE REPORT

Intestinal ischaemia caused by mesenteric inflammatory veno-occlusive disease

E. Eryigit[1*], F. Hoentjen[1], E. Barbe[2], J.J.M. van Meyel[1]

Departments of [1]Internal Medicine and Gastroenterology, and [2]Pathology, St. Lucas Andreas Hospital, Amsterdam, the Netherlands, [*]corresponding author: E_eryigit@hotmail.com

ABSTRACT

Mesenteric inflammatory veno-occlusive disease (MIVOD) is a rare cause of intestinal ischaemia. Previously described cases of MIVOD demonstrate vasculitis in mesenteric veins with thrombotic occlusion. It is important to distinguish MIVOD from other diseases, such as mesenteric venous thrombosis and systemic diseases. We present a case of a 39-year-old Turkish male in whom MIVOD was diagnosed after exclusion of other causes of ischaemic enteritis.

KEYWORDS

Intestinal ischaemia, thrombosis, vasculitis

INTRODUCTION

Mesenteric inflammatory veno-occlusive disease (MIVOD) as a cause of intestinal ischaemia was first described by Flaherty *et al*. in 1994.[1] They described seven patients who presented with signs of intestinal ischaemia requiring surgical intervention. In each case, the resected colon, small bowel, or both showed venulitis affecting veins of the bowel and mesentery, resulting in thrombotic occlusion of these veins. Vasculitis occurred without involvement of the mesenteric arteries and in the absence of systemic vasculitis or primary intestinal disease. The incidence and aetiology of MIVOD remain unclear because only a few cases have been reported so far. We describe a case of a Turkish male who presented with signs of intestinal ischaemia and required surgical intervention; the diagnosis of MIVOD was made after histopathological investigation of the resected bowel.

CASE REPORT

A 39-year-old Turkish male presented to the emergency room with a six-day history of constipation and progressive epigastric pain, nausea and anorexia for the last four days. Besides urinary stones, his medical history was unremarkable; he was not on any medication. Physical examination revealed a blood pressure of 126/79 mmHg, a pulse rate of 88 beats/min and a body temperature of 38.2 °C. The abdomen demonstrated epigastric tenderness without peritoneal signs. Laboratory findings showed an increased erythrocyte sedimentation rate (40 mm/h), a leucocyte count of 10.2 x 10^9/l and elevated C-reactive protein (231 mg/l). Liver and pancreas enzymes were within normal ranges. An abdominal ultrasound showed ascites, predominantly around the liver. A chest X-ray showed no signs of perforation and an abdominal X-ray excluded bowel distension. The patient developed increasing abdominal pain and subsequently peritoneal signs. An exploratory laparotomy followed, which demonstrated a segment of oedematous, ischaemic ileum; 52 cm was resected. Macroscopic examination of the resected necrotic ileum showed areas of haemorrhage. Microscopic examination revealed haemorrhagic mesenterial fat and necrosis without ulceration. No evidence for Behçet's or Crohn's disease was found. The associated small and medium-sized mesenteric veins showed necrotising vasculitis with occlusive thrombi with a focal marginalisation of granulocytes and deposition of fibrin within the vessel wall (*figures 1* and *2*). Neither arterial involvement nor granulomas were seen. These histopathological findings fit the diagnosis of ischaemic enteritis based on a veno-occlusive mesenteric vasculitis. Blood tests for hypercoagulability and systemic vasculitis were negative. The patient recovered completely with no recurrence in a follow-up period of more than 15 months.

Figure 1. *Veno-occlusive example: focal thrombosis of a middle sized mesenterial vein (20x)*

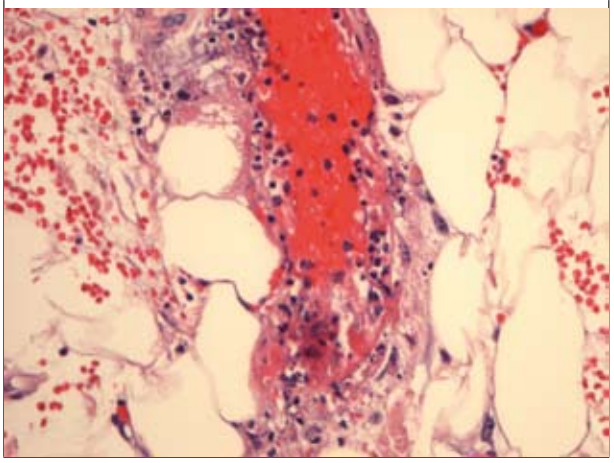
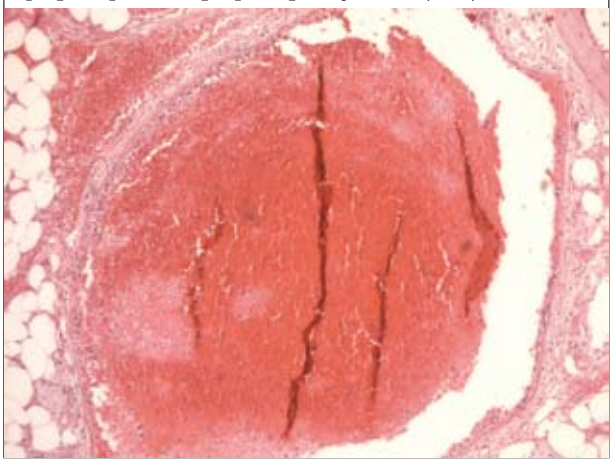


Figure 2. *Small mesenterial vein with necrotising venulitis characterised by destruction of the vessel wall with fibrinoid necrosis and an infiltrate of lymphocytes and polymorph infiltrate (20x)*



DISCUSSION

We present a patient who developed peritoneal signs leading to laparotomy. After small bowel resection, an ischaemic ileum due to MIVOD was diagnosed. Histopathological findings of the resected material revealed an isolated vasculitis of the small mesenteric veins and their intramural tributaries with thrombosis as a secondary manifestation. No arterial involvement was seen. Our histopathological findings are in agreement with previous reports on MIVOD. For example, Tempia-Caliera *et al.* described thrombophlebitis of small veins in the proximal ascending colon with fibroblastic organisation without arterial involvement.[2] Furthermore, Hu *et al.* demonstrated mesenteric oedema and haemorrhage, haemorrhagic infarctions and necrotising venulitis in the submucosa and mesentery with thrombotic occlusions of recent onset.[3] The diagnosis of MIVOD in our case was based on histopathological findings, in combination with the exclusion of other diseases. Intestinal vasculitis mostly occurs secondary to systemic vasculitis, as in Buerger's disease, Behçet's disease, rheumatoid arthritis and systemic lupus erythematosus (SLE) or in association with primary intestinal diseases such as Crohn's disease (*table 1*). However, unlike MIVOD, the systemic vasculitis will predominantly affect the arteries. Furthermore, our patient had no extra-intestinal signs of a systemic disease and no history of bowel disease. Finally, associations with mesenteric venous thrombosis such as a hypercoagulable state, trauma or sepsis were not present in our patient.

A few prior cases with similar presentation and histopathology findings have been reported in literature.[3,4] The disease clinically presents as an ischaemic colitis with abdominal pain for days or weeks accompanied with nausea and bloody stools. Characteristic features of MIVOD are lymphocytic, necrotising, granulomatous or mixed inflammatory infiltrates of the mesenteric veins and its intramural tributaries with secondary development of thrombosis as an important and immediate cause of ischaemic intestinal damage. Some cases describe myointimal hyperplasia associated with chronic MIVOD. No involvement of arterial inflammation or occlusion has been described. The aetiology of MIVOD remains unclear but associations with the antiphospholipid syndrome and the drug rutoside, an antioxidant drug used to treat varicose veins, have been described.[5,6] MIVOD seems to predominantly affect the colon, although it has also been reported to affect the small bowel, omentum and gallbladder.

Treatment of MIVOD involves surgical intervention, as described in previously published case reports. MIVOD is difficult to diagnose in an early stage due to the nonspecific symptoms and the histopathological requirements. Therefore, excluding other diseases using laboratory and radiology findings, such as mesenteric venous thrombosis

Table 1. *Major causes leading to arterial and venous ischemia of the bowel*

Arterial ischemia
Superior mesenteric artery embolism
Superior mesenteric artery thrombosis
Vasculitis (Behcet's disease, SLE, in association with Crohn's disease)
Shock
Venous ischemia
Mesenteric venous thrombosis (hypercoagulable state, trauma, sepsis)
Mesenteric inflammatory veno-occlusive disease

(MVT) and systemic diseases such as Behcet's disease and SLE, becomes of crucial importance. The latter diseases would require therapies such as corticosteroids, immunosuppressive therapy, or anticoagulant therapy. All of these therapies come with a significant risk of side effects.

Follow-up of MIVOD has been reported for up to 15 years in literature. Although recurrence of MIVOD is unusual after surgical resection, it has been described in one case.[2] Maintenance therapy is not required since the prognosis of MIVOD is excellent and the disease is unlikely to reoccur. Our patient recovered completely with no recurrence in a follow-up period of more than 15 months. In conclusion, we present a case of mesenteric inflammatory veno-occlusive disease leading to intestinal ischaemia. This diagnosis should be considered after exclusion of other causes of ischaemic enteritis.

REFERENCES

1. Flaherty MJ, Lie JT, Haggitt RC. Mesenteric inflammatory veno-occlusive disease. A seldom recognized cause of intestinal ischemia. Am J Surg Pathol. 1994;18:779-84.
2. Tempia-Caliera AA, Renzulli P, Z'graggen K, Lehmann T, Ruchti C, Buchler MW. Mesenteric inflammatory veno-occlusive disease : A rare cause of intestinal ischemia. The first description of recurrent disease. Digestion. 2002;66:262-4.
3. Hu JCC, Forshaw MJ, Thebe P, Stewart M. Mesenteric inflammatory veno-occlusive disease as a cause of acute abdomen: report of five cases. Surg Today. 2005;35:961-4.
4. Lie JT. Mesenteric inflammatory veno-occlusive disease (MIVOD): An emerging and unsuspected cause of digestive tract ischemia. Vasa. 1997;26:91-6.
5. Gul A, Inanc M, Ocal L, Konice M, Aral O, Lie JT. Primary antiphospholipid syndrome associated with mesenteric inflammatory veno-occlusive disease. Clin Rheumatol. 1996;15:207-10.
6. Saraga EP, Costa J. Idiopathic entero-colic lymphocytic phlebitis: cause of ischaemic intestinal necrosis. Am J Surg Pathol. 1989;13:303-8.