CASE REPORTS

Mesenteric vasculitis associated with Sézary syndrome

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Abstract

A 71 year old lady with Sézary syndrome presented with chronic diarrhoea and cramping abdominal pains. A small bowel meal x ray examination showed two mid-ileal strictures. At laparotomy the small intestine was found to be sub-acutely obstructed and resection of a 15 cm ileal stricture and stricturoplasty of a second, 10 cm stricture were performed. Histological examination of the stricture revealed a mesenteric vasculitis with secondary ischaemic changes in the small bowel wall. Mesenteric vasculitis causing small bowel stricture may be associated with Sézary syndrome.

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Keywords: Sézary syndrome, mesenteric vasculitis, small bowel stricture.

Small bowel strictures are often due to Crohn's disease, and less commonly due to malignancy, non-steroidal anti-inflammatory drugs (NSAIDs) and ischaemia, but primary mesenteric vasculitis should be considered and may be difficult to diagnose.

Case report

A 71 year old lady presented with Sézary syndrome, namely erythroderma, generalised lymphadenopathy, and the presence of circulating Sézary cells with T cell receptor gene rearrangements. Previous treatments with emollients, topical and systemic corticosteroids, etretinate and psoralens with long wave ultraviolet irradiation (PUVA) had been unsuccessful, but she had shown significant symptomatic benefit following four cycles of extracorporeal photopheresis. Photopheresis entails extracorporeal exposure of blood lymphocytes to photoactivated 8-methoxypsoralen, followed by return of the treated cells, and is thought to act by enlisting the immune system to control aberrant clones of T cells.¹

She then developed persistent watery diarrhoea, with occasional cramping abdominal pains and considerable weight loss. An intermittent, low grade fever was recorded. There was no evidence of pathogenic organisms, including *Clostridium difficile*, in multiple stool specimens. A plain abdominal x ray showed

dilated loops of small bowel but no evidence of intestinal obstruction. At sigmoidoscopy a mildly inflamed rectal mucosa was observed and histological examination of a biopsy specimen showed mild non-specific inflammatory changes.

Further investigation revealed anaemia (haemoglobin 10.1 g/dl (normal 12.0-15.0), mean cell volume 93 fl (normal 80-100), and normal serum values of haematinics), raised white cell ($12.2 \times 10^6/\text{ml}$ (normal 4.0-11.0) with 7% eosinophilia and 19% Sézary cells), and platelet counts ($776 \times 10^6/\text{ml}$ (normal 150-400)), erythrocyte sedimentation rate of 26 mm in the first hour and C reactive protein of 139 mg/l (normal <10). The serum albumin concentration fell steadily, to a nadir of 14 g/l (normal 41-51) over seven weeks, and oedema developed. The patient was fed an elemental diet with reduction in diarrhoea and increase in the albumin value to 28 g/l. She then developed a swollen left leg and an ultrasound Doppler examination confirmed an iliofemoral deep venous thrombosis and anticoagulation was started.

A small bowel meal x ray examination was abnormal with two localised mid-ileal strictures (Fig 1) but computerised tomography of the abdomen was normal. The aetiology of the stricture was unclear and so a laparotomy was performed when the small intestine was found to be sub-acutely obstructed and resection of a 15 cm ileal stricture and stricturoplasty of a second, 10 cm stricture were performed.

Histological examination of the stricture showed acute and chronic inflammation affecting all small arteries and veins in the mesentery. There was necrosis and ulceration of the mucosa, marked active chronic inflammation, and fibrosis of the submucosa and muscularis mucosa. There were no fissures, granulomata or other changes suggestive of Crohn's disease. The changes were consistent with a primary mesenteric vasculitis with secondary ischaemic changes in the bowel wall (Fig 2). Treatment with prednisolone 30 mg once daily was started and within two weeks the diarrhoea had resolved, the patient gained weight, and the C reactive protein fell to 24 mg/l. There has been no evidence of relapse for one year and the dose of prednisolone has been slowly reduced to 10 mg once daily.

No conventional cause for a vasculitis was found: there were no cutaneous stigmata of

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Figure 1: A small bowel meal x ray examination showing an ileal stricture (marked with arrows).

vasculitis or of connective tissue disease; anti-nuclear and rheumatoid factors and antineutrophil cytoplasmic antibody were negative. Tests for hepatitis B and C and for human T cell lymphotropic virus I were negative and there was no evidence of myeloma. There was no family history of thrombosis and anticardiolipin antibody, protein C, protein S, and antithrombin III were all normal. NSAIDs had not been taken.

Discussion

This is a case of mesenteric vasculitis causing small bowel strictures in a patient with Sézary syndrome. Sézary syndrome is a cutaneous form of leukaemia lymphoma characterised by erythroderma, pruritis, adenopathy, and circulating atypical cells with cerebriform nuclei.² Mesenteric vasculitis has been described as a histological feature of Crohn's disease³⁻⁶ but primary mesenteric vasculitis is a rare cause of small bowel strictures.⁷ Rheumatoid arthritis,⁸ scleroderma,⁹ polyarteritis nodosa,^{10 11} giant cell arteritis,^{12 13} Henoch-Schoenlein purpura,¹⁴ Wegener's granulomatosis, and Churg-Strauss syndrome^{15 16} are a group of inflammatory disorders that may result in mesenteric vasculitis, but this usually presents with acute mesenteric ischaemia with or without perforation or gastrointestinal haemorrhage, rather than strictures.

There are no previous reports of this association, although there is a single report of a cutaneous necrotising vasculitis associated with Sézary syndrome.¹⁷ Thrombosis of small vessels without vasculitis has been reported in the antiphospholipid syndrome in association with mycosis fungoides, another T cell lymphoma.¹⁸ Mycosis fungoides has also been associated with cutaneous vasculitides, but not with intestinal involvement.¹⁹

Figure 2: Section of mesenteric vessels from small bowel stricture showing vasculitis (haematoxylin and eosin X100).

Mesenteric vasculitis causing small bowel strictures may be associated with Sézary syndrome.

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