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A Case Report of Mesenteric Panniculitis and Primary Sjögren's Syndrome

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ABSTRACT

Mesenteric Panniculitis is a benign fibro-inflammatory process involving adipose tissue of the mesentery. It is characterised by fat necrosis, chronic inflammation and fibrosis, causing thickening and shortening of the mesentery. Patients may present with localised abdominal pain, abdominal mass, intestinal obstruction and ischaemic colitis. We report a case of mesenteric panniculitis causing abdominal pain in a patient with active Primary Sjögren's Syndrome. The rarity of this case makes it of interest. We review the current literature on mesenteric panniculitis and its association with connective tissue disease and inflammatory conditions. A 64-year-old Caucasian male presented in 1994 with dry mouth. A diagnosis of Primary Sjögren's Syndrome (PSS) was made on salivary gland biopsy. In 2010 he presented with an exacerbation of his eye symptoms, muscle pain and fatigue. He complained of abdominal pain and night sweating, but denied any weight loss or change in bowel habit. There was no significant past medical history other than PSS. On examination he had a small right submandibular node and mild synovitis at the right proximal interphalangeal joint and carpometacarpal joint. Examination of the abdomen showed marked umbilical tenderness but no organomegally. Blood tests at this time showed an active inflammation: CRP of 61 (NR < 5 mg/L), ESR 39 (NR < 20 mm/s), strongly positive ENA Ro and La. IgG was elevated at 18.6 (NR 5.8 - 15.4), Complement was low at 0.17 (NR 0.18 - 0.6). An abdominal ultrasound scan demonstrated a 6 × 3 × 3 cm area of diffuse homogenous fat encasing some mesenteric vessels in the area of focal tenderness. CT abdomen and pelvis showed oedematous mesenteric fat and lymph nodes in the jejunal small bowel mesentery, consistent with mesenteric panniculitis. Laparoscopic biopsy was discussed with the surgical team, but was felt not indicated as risk outweighed potential benefit. The patient was treated with a 9-week reducing course of oral steroids. His abdominal symptoms resolved although CT abdomen showed little improvement in mesenteric panniculitis. A review of the literature suggests that currently there is no standard treatment and management should be guided by patient symptoms. Mesenteric Panniculitis is rare; as a result evidence for treatment is limited to individual case reports. There is no clear link between symptom improvement and radiological resolution of mesenteric panniculitis. It has, therefore been suggested that follow-up imaging should be limited to those with persistent symptoms. Overall the prognosis for mesenteric panniculitis is good, up to half of patients do not require treatment, and recurrence of symptoms is uncommon.

Keywords: Sjögren's Syndrome; Autoimmune Disease; Mesenteric Panniculitis

1. Introduction

Mesenteric Panniculitis is a benign fibro-inflammatory process involving adipose tissue of the mesentery [1]. It is characterised by fat necrosis, chronic inflammation and fibrosis, causing thickening and shortening of the mesentery [2]. The pathophysiology remains unclear, although it has been linked with malignancy—in particular lymphoma, gastrointestinal and genitourinary [3]. Patients with mesenteric panniculitis may present with localised abdominal pain, abdominal mass, intestinal obstruction and ischaemic colitis, however between 30% - 50% remain asymptomatic [2].

We report a case of mesenteric panniculitis causing abdominal pain in a patient with active Primary Sjögren's Syndrome. The rarity of this case makes it of interest: We review the current literature on mesenteric panniculitis and its association with connective tissue disease and

inflammatory conditions.

2. Case Report

2.1. Background

EM is a 64-year-old Caucasian male. His initial symptoms began in 1994, when he required multiple dental fillings and complained of dry mouth. A diagnosis of Primary Sjögren's Syndrome (PSS) was made on Salivary gland biopsy. His symptoms became more troublesome in 2003. He complained of dry eyes and mouth; wrist, back and neck pain; dry skin and profound fatigue. There were no significant symptoms of the respiratory, gastrointestinal, genitourinary or neurological systems at this time. His symptoms were well controlled with lubricating agents and NSAIDs as required.

2.2. Progression of Disease

In 2010, he presented with an exacerbation of his eye symptoms, increasing muscle pain and fatigue. He complained of abdominal pain and night sweating, but denied any weight loss or change in bowel habit. He also complained of increasing pain in his legs and lower back. He denied paraesthesia or loss of power. There was no significant past medical history other than PSS, in particular, no history of vascular disease. He holidayed in China in September 2009 but was well throughout his stay and on his return.

2.3. Examination Findings

On examination he had a small right submandibular node and mild synovitis at the right proximal interphalangeal joint and carpometacarpal joint. Examination of the abdomen showed marked umbilical tenderness but no organomegally. Straight leg raising was significantly reduced bilaterally, and there was altered sensation in the L5/S1 distribution bilaterally. He also had some tenderness at L5.

2.4. Investigations

Blood tests at this time showed an active inflammation with an elevated CRP of 61 (normal range < 5 mg/L), ESR 39 (normal range < 20 mm/s), and strongly positive ENA Ro and La. IgG was elevated at 18.6 (normal range 5.8 - 15.4), Complement was low at 0.17 (normal range 0.18 - 0.6) Liver function tests and renal function were normal. Quantiferon gold test was positive, this was not pursued further. There was no evidence of malarial parasites on blood film. An abdominal ultrasound scan demonstrated a 6 × 3 × 3 cm area of diffuse homogenous fat encasing some mesenteric vessels in the area of focal tenderness. At this stage a CT was advised to further characterise this area.

CT abdomen and pelvis showed oedematous mesenteric fat and lymph nodes in the jejunal small bowel mesentery, consistent with mesenteric panniculitis.

CT PET confirmed a mild mesenteric panniculitis, but otherwise no significant abnormality.

The patient was reviewed by a neurologist who felt that his lower limb weakness and patchy sensory disturbance were most likely due to a radiculopathy, however a small fibre neuropathy could not be ruled out. Nerve conduction studies, CK and MRI lumbar spine were normal.

It was noted that IgA Tissue Transglutaminase 92.6U (normal range 0 - 15U) and IgA endomysial antibody was positive. A GI Endoscopy was performed to rule out coeliac disease (associated with peripheral neuropathy). Duodenal biopsy was normal, with no increase in epithelial lymphocytes or abnormality of the villous architecture. Coeliac disease and TB were excluded.

Laparoscopic biopsy was discussed with the surgical team, but was felt not indicated as risk outweighed potential benefit.

2.5. Management

The patient was treated with a 9-week-reducing course of oral steroids. His abdominal symptoms resolved although CT abdomen showed little improvement in mesenteric panniculitis. The decision not to pursue further investigation was discussed and agreed by the patient as his symptoms improved and he had undergone a number of investigations already.

3. Discussion

Mesenteric Panniculitis is a non-specific inflammatory process, most commonly affecting the fatty tissue at the root of the small bowel mesentery, [4] resulting in thickening and shortening of the mesentery. The bowel, adjacent lymph nodes and vessels are usually not affected [2].

Little is known about the pathophysiology of mesenteric panniculitis. Characteristic histopathological findings include non-focal lymphoplasmocytic infiltration, fat cell necrosis, foamy macrophages and focal fibrosis. Schaffler *et al.* argue that adipocytes and adipocytokines are involved in primary inflammatory processes, and that mesenteric panniculitis is just one example of this [5].

There is a variety of terminology relating to the above process, which is based on the varied histological findings. These include mesenteric lipodystrophy (predominantly fatty necrosis), mesenteric panniculitis (Chronic inflammation), and retractile mesenteritis or mesenteric fibrosis (predominantly fibrosis). Emory *et al.* suggested the overarching terminology of sclerosing mesenteritis, under which the headings mesenteric panniculitis (predominant inflammation and fat necrosis) and retractile me-

senteritis (predominant fibrosis and retraction) are recognised [6].

Such varied histology is likely to account for the varied clinical presentation. Documented presenting symptoms include abdominal pain, palpable mass, weight loss, small bowel obstruction, diarrhoea and chylous ascites [7, 8]. Occasionally it is asymptomatic, identified as an incidental finding on CT [3].

A recent systematic review (between 1980 and 2012) of 262 patients who were admitted with mesenteric panniculitis demonstrated that 69% were male, with a mean age of 63 years [8]. Similarly, Akram *et al.* reviewed 92 cases between 1982-2005, and found that 70% were male, the mean age of onset was 65 years (55 - 72 years) [9]. The prevalence of mesenteric panniculitis was documented as 0.6% in one study: Daskalogianniki *et al.* prospectively reviewed over 7000 abdominal CTs, looking for features typical of mesenteric panniculitis. Of these, 49 demonstrated mesenteric panniculitis. 34 cases were associated with malignant disease, 11 cases associated with benign disease, and in the remaining 4 cases no association was identified [3].

Etiological associations of mesenteric panniculitis include malignancy, infectious disease, autoimmune disease, vasculitis, cirrhosis, peptic ulcer disease, pancreatitis, abdominal aortic aneurysm, previous abdominal surgery and elevated IgG4 related disease [3] (a fibroinflammatory condition initially associated with autoimmune pancreatitis, but now recognised in a number of conditions including atopic dermatitis, vasculitis, pancreatic cancer, biliary tract disease, and also present in 5% of the normal population) [10].

In terms of autoimmune disease, mesenteric panniculitis has been recognised in patients with autoimmune haemolytic anaemia [11], Coeliac disease, thyroiditis, primary sclerosing cholangitis, rheumatoid arthritis, lupus and polychondritis [12,13]. However to our knowledge, there is only one other case documenting an association with Sjogren's syndrome [14]. Like our case report, this patient presented with abdominal pain. She initially underwent surgery for acute appendicitis and ileocaecal abscess. She had a recurrence of her symptoms associated with annular rash and fever. Review of the histopathology suggested mesenteric panniculitis. She was treated with 20 mg prednisolone daily and her symptoms resolved. CT also demonstrated complete resolution of the soft tissue density after 3 weeks of prednisolone treatment.

It is also worth noting that although our patient did not have clinical or histological evidence of coeliac disease, he did have raised IgA TTG and IgA endomysial antibody. There is a documented association between mesenteric panniculitis and coelic disease, supporting the link between mesenteric panniculitis and autoimmune disease.

Daskalogiannaki *et al.* found that of the 15 patients with non-malignant disease, only three had follow-up imaging. CT findings showed stable disease, that is, there was no progression to retractile mesenteritis or malignancy. Of the four biopsy-proven symptomatic patients, three were treated with prednisolone. Two showed a clinical improvement, and one had on-going intermittent abdominal pain. On follow-up CT, none of the patients had resolution or change in the mass [3]. The evidence for radiological improvement from other case reports following treatment is mixed [9,13,15]. This suggests that reimaging should be guided by patient symptoms in clinical practice.

There is no consensus on treatment of mesenteric panniculitis. Most would agree that treatment should be symptom-directed. Potential treatment options include surgical resection, immunomodulatory (steroids, azathioprine, cyclophosphamide), antifibrotic agents (thalidomide, [16] tamoxifen, [9] pentoxyfylline [17] NSAIDS (colchicine) [18] and conservative management. In this case report our patient showed a moderate response to prednisolone and no further intervention was required.

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