



An unusual presentation of sclerosing mesenteritis mimicking inflammatory bowel disease in a teenager

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UP collected the clinical data, and wrote the initial and final drafts of the paper; SB wrote the initial draft and supervised data collection; SRD collected the clinical data; IS achieved the histopathological diagnosis; DES supervised the data collection and the manuscript

This paper presents a rare condition as a differential diagnosis in a teenager with right iliac fossa pain.

Case report

A 16-year-old male patient who was previously fit and healthy presented initially with sudden onset of lower abdominal pain. He had not suffered from similar pain in the past. He had not suffered from any significant medical diseases. He smoked up to 16 cigarettes a day, his bowel and bladder habits were regular. There was no family history of inflammatory bowel disease. Physical examination revealed his heart rate was 111 beats per minute and he was afebrile. On abdominal examination he was tender over both iliac fossae, more on the right side. Blood tests showed white cell count was 12 and C-reactive protein was <6. Urinalysis was positive for nitrates and protein. Based on his clinical presentation and elevated white cell count he was diagnosed to have appendicitis. He went on to have open appendectomy; later we found the histology did not reveal any inflammation.

Three months later the teenager presented again with right iliac fossa pain and signs of local peritonitis. Laboratory tests revealed white cell count was 18.1 and C-reactive protein was 51. A laparoscopy was performed which showed postoperative adhesions which were released. This was followed by recovery and discharge from the hospital. Following his discharge he re-presented within a period of two months with right iliac fossa pain and an elevated white cell count of 13 and C-reactive

protein of 248. The clinical presentation was investigated with a computed tomography (CT) scan of the abdomen and pelvis which did not reveal any abnormality in the right iliac fossa; the rest of the abdomen was unremarkable.

Persisting pain and escalating inflammatory markers prompted a laparotomy which revealed thickened ileocaecal area suggestive of inflammatory bowel disease. Following recovery, the provisional diagnosis of inflammatory bowel disease was investigated with colonoscopy with terminal ileal biopsies, oesophagogastroduodenoscopy and barium-follow-through all of which were in the normal range. However the barium follow-through, all of which were normal. An isotope-scan (white cell tagged) showed inflammation of the distal small bowel. Following a gastroenterology consultation, the patient was commenced on systemic steroids for suspected Crohn's disease. Ten days later, a further admission (fifth) with similar symptoms and signs warranted a CT scan, which demonstrated multiple mildly dilated fluid filled loops of small bowel with no other significant pathology. He underwent a subsequent diagnostic laparoscopy where further division of adhesions was performed. There was evidence of scarring in the terminal ileum (Figure 1) and a full thickness biopsy of the abnormal ileum was taken. The biopsy revealed evidence of serositis without any histological features of Crohn's disease.

Compounding the diagnostic conundrum, the teenage patient was admitted for the sixth episode with a similar presentation. Prompted by recurrent admissions, impairment of the patient's

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Figure 1

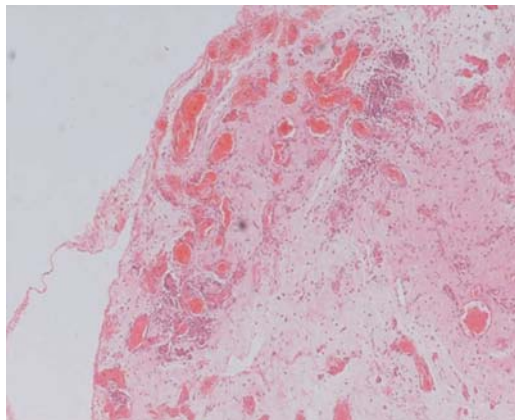
White patchy lesion at ileocaecal junction on a background of scarring and abnormal vasculature seen during laparoscopy 133 × 91 mm (300 × 300 DPI)



quality of life and after extensive discussion with the patient it was decided to proceed to diagnostic laparoscopy and limited right hemicolectomy as the visible disease process was confined to that region. The surgery was undertaken on an urgent basis. Recovery was uneventful and cessation of symptoms was observed at the 10-month follow-up. Histology of the resected specimen revealed sclerosing mesenteritis (Figure 2), which is relatively rare pathology in a teenage patient.

Figure 2

Histopathology specimen from right hemicolectomy showing chronic inflammation of the mesentery with fibrosis 149 × 121 mm (150 × 150 DPI)



Discussion

Sclerosing mesenteritis is a rare, benign inflammatory disorder of unknown aetiology, affecting the membranes of the digestive tract. The commonest site affected is the mesentery of the small bowel and more rarely the disease affects the mesocolon,¹ peripancreatic region, omentum and pelvis. Characteristic features include fat necrosis, fibrosis and chronic inflammation. Sclerosing mesenteritis usually presents in the fifth and sixth decades and is twice as common in men than in women.² Sclerosing mesenteritis is synonymous with mesenteric hypodystrophy, retractile mesenteritis, mesenteric panniculitis and mesenteric sclerosis.

The aetiology of sclerosing mesenteritis is idiopathic in most cases but the disease is known to have association with previous abdominal surgery; retroperitoneal fibrosis, Riedel's thyroiditis, sclerosing cholangitis, vasculitis and granulomatous disorders. Clinical features are variable; abdominal pain is the most common symptom but sclerosing mesenteritis also presents with distension/bloating, diarrhoea, weight loss, fever, abdominal mass and bowel obstruction. Clinical examination does not contribute much to the diagnosis of sclerosing mesenteritis apart from non-localizing tenderness and the presence of an abdominal mass in rare cases.

Diagnosis of sclerosing mesenteritis is radiological, histopathological or incidental. CT scan findings may include the appearance of a heterogeneous soft tissue mass in the mesentery of the small bowel, entrapment of the superior mesenteric artery with no vascular involvement, and no invasion of adjacent small bowel loops. Two CT findings are considered more specific for the diagnosis of sclerosing mesenteritis³:

- The presence of a tumour pseudo-capsule, which is a hyperattenuated stripe surrounding the mass in the mesentery of the small bowel. This is seen in 60% of cases;
- The 'fat ring' sign of hypodense fatty halo surrounding mesenteric nodules and vessels. This is seen in up to 75% of cases;
- The definitive diagnosis of sclerosing mesenteritis is made through histological analysis of specimens obtained laparoscopically or through open laparotomy.

In the early stage when fat necrosis predominates with little or no fibrosis, the diagnosis of mesenteric lipodystrophy has been used. Later, when fat necrosis subsides and chronic inflammation becomes predominant, it may be designated mesenteric panniculitis. Finally, when fibrosis becomes the main feature, sclerosing or retractile mesenteritis has been used.² On immunohistochemical staining, sclerosing mesenteritis specimens stain positive for vimentin, desmin and b-cathepsin suggesting increased myofibroblastic activity⁴ and negative for desmin, keratin, and S-100 protein.⁵

Treatment is tailored to the patient's individual symptoms, accompanying conditions and complications. In one study conducted by Akram *et al.* of ninety two patients, 52% of patients did not receive any treatment, 26% were treated with medical therapy alone, 13% with surgery alone and 9% with both surgical and medical therapy.⁶ Corticosteroids, colchicine, azathioprine, thalidomide, cyclophosphamide, tamoxifen, progesterone are some of the therapeutic agents used with varied results. Our patient was commenced on steroids and the literature does suggest better efficacy with combination treatment which may explain that lack of resolution. Surgical treatment options may range from excision of the mass to resection of small/large bowel.

Differential diagnoses of sclerosing mesenteritis are inflammatory bowel disease, extra-abdominal fat necrosis (Weber-Christian disease), pancreatitis, retroperitoneal fibrosis, desmoids, carcinomatosis, carcinoid, primary mesenteric mesothelioma and B-cell lymphoma.⁷ Sclerosing mesenteritis is a benign disease but left untreated runs a rather debilitating course. Complications include bowel obstruction, gastrointestinal bleeding and arterial/venous thrombosis. Fatal outcomes have been reported due to these complications.

In summary there is no agreed management of sclerosing mesenteritis, however literature suggests watchful waiting could be done in cases with fat necrosis alone,¹ once chronic inflammation sets in various immunosuppressive therapies are suggested^{6,8,9} and surgery is reserved for failed medical therapy and complications.¹⁰

Conclusion

Sclerosing mesenteritis is a rare non-neoplastic idiopathic inflammatory disorder commonly affecting adults who are middle-aged or older. The take home message from our report is that this condition can be seen in a young adult which was initially mimicking appendicitis, and later inflammatory bowel disease. However, the diagnostic conundrum was resolved by surgery and the histology of the specimen.

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