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An Interesting Case of Recurrent Small Bowel Obstruction

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Small bowel · Neuroendocrine tumours · Gastrointestinal oncology

Abstract
Sclerosing mesenteritis is associated with a spectrum of diseases which include mesenteric lipodystrophy and mesenteric panniculitis. This inflammatory and fibrosing disorder can affect the small and large bowel wall and mesenteric vessels by exerting a mass effect. The following case highlights the difficulties with diagnosing and managing this unusual disease. A 64-year-old man presented with acute central abdominal pain, radiating to his back, and profuse vomiting. He was diagnosed clinically with small bowel obstruction. He had had an episode of small bowel obstruction 6 years earlier. At this time, he underwent an exploratory laparotomy, and a mass was identified in the small bowel mesentery. The features were thought to be in keeping with sclerosing mesenteritis. He had a dramatically favourable response to the initiation of prednisolone. He continued to be well and asymptomatic for a further 5 years on long-term maintenance low-dose steroids and 6-mercaptopurine. He re-presented in 2009 (six years after initial presentation) with very severe acute abdominal pain and vomiting. He had no recent change in weight or appetite, and had not had time off work. He underwent a second laparotomy and the tissue diagnosis was of metastatic carcinoid tumour involving the small bowel mesentery. This is the first case to our knowledge where sclerosing mesenteritis has been confirmed histologically on biopsy and then subsequently diagnosed with histologically proven carcinoid tumour. For this particular reason it must be always remembered that sclerosing mesenteritis is a 'pathological' and not a radiological diagnosis and that a large proportion of cases are associated with neoplasia.

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Introduction

Sclerosing mesenteritis is associated with a spectrum of diseases which include mesenteric lipodystrophy and mesenteric panniculitis. This inflammatory and fibrosing disorder can affect the small and large bowel wall and mesenteric vessels by exerting a mass effect. The first known series, which comprised 34 cases of 'retractile mesenteritis and mesenteric sclerosis', was published in 1924 [1]. This disorder is relatively uncommon and predominantly affects men aged between 50 and 70 years. There are a variety of symptoms associated with this disorder which include nausea, vomiting, weight loss and fever [2–4]. In 1997 after a review of 84 cases, Emory et al. [5] concluded that these histological variants are part of one disease process and that sclerosing mesenteritis would be an appropriate common term. Due to the low incidence of this condition it has been difficult to study the clinical features, natural history, and ultimate response to therapy and prognosis. Many of the therapeutic regimes to treat sclerosing mesenteritis have been guided by anecdotal evidence and small case series [6–9].

The pathophysiology of sclerosing mesenteritis remains obscure, although there have been several proposed mechanisms. These include abdominal surgery or trauma [5], autoimmunity [10–13], paraneoplastic syndrome [2, 3, 5, 14], ischaemic injury and infection [15]. The following case highlights the difficulties with diagnosing, managing and appropriate follow-up for this unusual disease.

Case Report

A 64-year-old man presented in 2009 with acute central abdominal pain, radiating to his back, and profuse vomiting. He had no recent change in weight or appetite and had not had time off work. In late 2008 he had also presented with two discrete acute episodes of small bowel obstruction. Both of these episodes responded rapidly to pulsed high-dose corticosteroids. His symptoms dated back to 2003 when he had presented with abdominal pain, weight loss and vomiting. At this time, he was diagnosed clinically with small bowel obstruction. At that stage he had a CT abdominal scan which diagnosed a 'small bowel mass'. The working diagnosis was small bowel lymphoma. He subsequently underwent an exploratory laparotomy. A mass was identified in the small bowel mesentery, this was biopsied and an adjacent lymph node was dissected. In 2003 the biopsy specimen was reported as follows: '… there is evidence of an infiltrative process, with entrapment of lymph nodes, nerves, blood vessels and fat. The process involves fibrosis, lymphocyte infiltration, focal fat necrosis and eosinophilic infiltration.' There was no evidence of lymphoma in the tissue or lymph node specimens. These pathological features were thought to be in keeping with sclerosing mesenteritis. This was confirmed by two experienced gastrointestinal pathologists. He had a dramatically favourable response to the initiation of prednisolone and was started on 6-mercaptopurine (as a steroid-sparing agent) and did not complain of any further abdominal pain or lethargy. He remained well and asymptomatic for a further 5 years on long-term maintenance low-dose steroids and 6-mercaptopurine.

From his most recent admission in April 2009 a CT abdomen was performed which demonstrated 'a soft tissue mass measuring 22 × 30 mm, "spiculated" in appearance and anterior to the third part of the duodenum. Small bowel loops appeared to encircle and were adherent to the soft tissue density and there were dilated proximal small bowel loops. There was a 9 mm low density lesion in segment 3 of the liver' (fig. 1). The differential diagnosis was sclerosing mesenteritis with adhesions or carcinoid tumour (with a desmoplastic reaction). He had a second laparotomy which confirmed the soft tissue mass adherent to the mid small bowel and a mesenteric mass. A nodule was identified and biopsied from the left lobe of the liver. The biopsy results revealed cells in a 'nested appearance' that stained positively for AE1/AE3, chromogranin and synaptophysin, consistent with a diagnosis of carcinoid. There was also carcinoid tumour present in the resected node and liver nodule. Therefore the diagnosis was metastatic carcinoid tumour. He subsequently had normal urinary 5-HIAA and neuroendocrine markers and did not report any symptoms of the carcinoid syndrome. Subsequent pathological review and repeat staining of the biopsy taken in 2003 did not reveal any evidence of carcinoid disease.

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Discussion

Sclerosing mesenteritis has been previously associated with other co-existent conditions including lymphoma, breast cancer, melanoma, bronchial carcinoma, renal carcinoma, multiple myeloma, hepatocellular carcinoma, prostate carcinoma, ovarian carcinoma, and gastrointestinal carcinomas [3, 14, 16]. In the literature there has been confusion when radiological imaging 'mistakenly' clinically and radiologically diagnosed sclerosing mesenteritis, when in fact the underlying diagnosis was carcinoid disease after histological confirmation.

This is the first case to our knowledge where sclerosing mesenteritis has been confirmed histologically on biopsy and subsequently diagnosed as histologically proven carcinoid tumour. What makes this case particularly interesting is that this patient had a dramatic response to steroids. Steroid responsiveness does not tend to be a feature of carcinoid tumours where the mainstay of therapy is octeotride [17, 18] and occasionally interferon-alpha [19–22] for refractory symptoms, but steroid responsiveness is consistent with sclerosing mesenteritis. Steroids have been shown to be beneficial in the treatment of sclerosing mesenteritis [6, 7, 9]. This patient had no symptoms to suggest features of the carcinoid syndrome, which is often the case in metastatic carcinoid disease – in fact more than 90% of cases of carcinoid syndrome will have the disease.

For this particular reason it must be always remembered that sclerosing mesenteritis is a 'pathological' and not a radiological diagnosis. Therefore when diagnosing sclerosing mesenteritis a clinician must have a high index of suspicion for other serious co-existent conditions, especially neoplasia.

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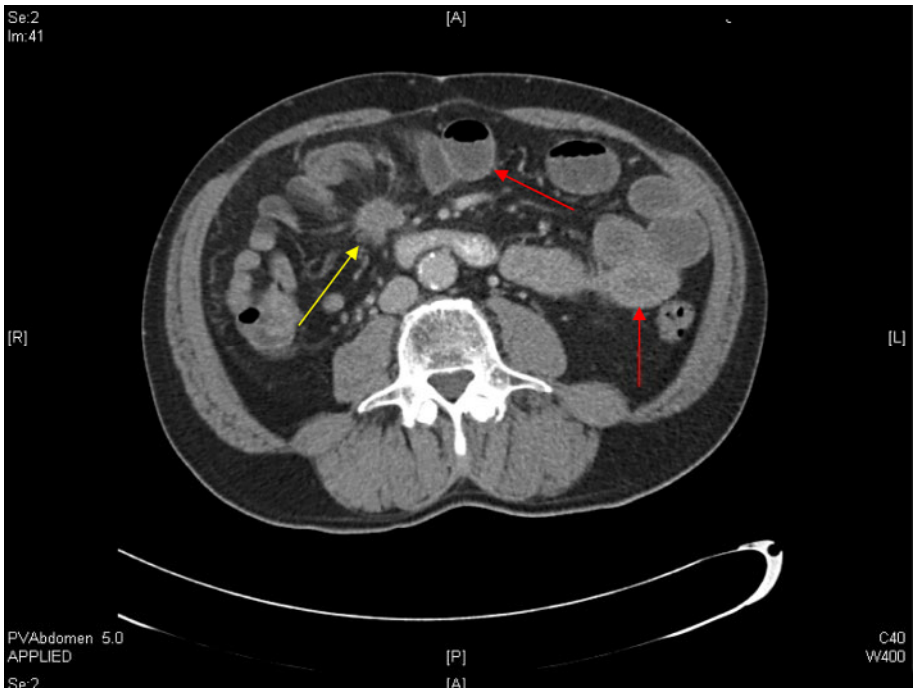
Fig. 1. Abdominal CT demonstrating the 'spiculated' soft tissue mass in the upper small bowel anterior to the third part of the duodenum (yellow arrow). The dilated proximal small bowel segment is indicated by the red arrows. Note that the colon is collapsed due to a proximal obstruction.



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