Abstract. – Sclerosing mesenteritis (SM) is a rare, idiopathic disorder of unknown aetiology that involves the adipose tissue of the mesentery, being characterized by chronic and non-specific fibrous inflammation. Patients usually present with non-specific clinical manifestations, such as abdominal pain and diarrhoea. The diagnosis of SM is difficult and it can be definitely established only by means of surgical or imaging-guided biopsy. Different therapeutic strategies have been used in case series with different rate of success. The disease is generally self-limiting, and the long-term prognosis is good, even if some cases of severe SM are reported in literature. Here, we report a fatal case of sclerosing mesenteritis associated to protein-losing enteropathy.

Key Words: Sclerosing mesenteritis, Panniculitis, Protein-losing enteropathy.

Introduction

Sclerosing mesenteritis (SM) is a rare, idiopathic, benign disorder of unknown aetiology that involves the adipose tissue of the mesentery, being characterized by chronic and non-specific fibrous inflammation\(^1\). First described by Jura in 1924\(^3\), numerous terms have been used to describe this disease, especially on the basis of the predominant histology, retractile mesenteritis or mesenteric fibrosis (predominant fibrosis)\(^4\), mesenteric panniculitis (marked chronic inflammation)\(^5\) and mesenteric lipodystrophy (predominantly fatty degeneration and necrosis)\(^6\). This varied terminology has caused considerable confusion until 1997 when, after a review of 84 cases, Emory et al\(^1\) concluded that these histologic variants are part of the spectrum of one disease, named sclerosing mesenteritis (SM), in which a fibrous component is constantly present.

The rarity of this condition has generated many troubles and discussions on epidemiological studies, clinical features and therapeutic strategies. Patients usually present with abdominal pain and distension, diarrhoea, weight loss and palpable abdominal masses\(^1\). Being its clinical manifestations non-specific, the diagnosis of SM remains difficult. Clinically, definitive diagnosis is established by means of a surgical or imaging-guided biopsy. The disease is generally self-limiting, and the long-term prognosis is good, even if some cases of severe/fatal SM are reported in literature\(^7\).

Treatment decisions are based on personal experience and case series. Different therapeutic strategies, including corticosteroids, colchicine, immunosuppressive drugs, and hormonal therapy have been used with different rate of success\(^7\).

Here, we report an unusual and fatal case of sclerosing mesenteritis associated to a protein-losing enteropathy (PLE).

Case Report

A 59-year-old male (height 1.65 m, weight 60.5 kg) was admitted to our hospital with a 3 months history of recurrent upper quadrants abdominal pain, associated with postprandial vomiting, diarrhoea (5 bowel movements/day without blood), weight loss (12 kg after symptom onset) and a clinical picture consistent with PLE. His past medical history included hypertension and a previous laparoscopic cholecystectomy after acute cholecystitis two years before. He had no familiar history of gastrointestinal disease or any form of malignancy.

Physical examination revealed slight fever, hypotension (95/60 mmHg), tachycardia, bilateral pitting oedema of the lower extremities. No palpable abdominal mass was observed.

Laboratory findings indicated the presence of chronic inflammatory status with significant ele-
vation of both ESR and C reactive protein, mild iron deficiency anaemia, severe hypoalbuminemia (2.3 g/dL), hypokalemia and pseudo-hypocalcemia.

CT scan showed mild segmental stricture of some proximal jejunal loops with the presence of significant hypertrophy of the mesenteric fat; the extensive fibro-fatty proliferation of the mesentery was associated with multiple enlarged lymphnodes (Figure 1a); flap bilateral pleural effusions and ascites were also evident. No (arterial/venous) vascular sign of mesenteric thrombosis was evident at angio-CT reconstructions. This complex picture appeared to be suspected for SM (in differential diagnosis with Crohn’s disease). Upper endoscopy with push enteroscopy showed the presence of superficial ulcers with a major (extrinsic) stricture sited at the fourth portion of duodenum/first loop of the jejunum. Histology did not show specific inflammatory signs (immunohistochemistry negative for CMV, absence of granulomas, absence of neoplastic changes).

Despite the continuous administration of methylprednisolone 60 mg/day, albumin, potassium, calcium and supportive parenteral nutrition, we did not observe significant improvement in laboratory parameters and clinical conditions so that the patient was referred to diagnostic/therapeutic surgery.

The surgical procedure evidenced a significant thickening of the mesenteric fat with the presence of a multiple visceral kinking of the bowel loops. Biopsies were performed on the fibrotic stretch of the mesentery. The histological examination showed fibrous and fatty tissue with focal edema and moderate lymphoid infiltrate without significant atypia, a picture consistent with the diagnosis of SM (Figure 1b).

We decided to start therapy with tamoxifen (20 mg/day) which showed no efficacy.

After 1 month from the surgical procedure our patient died because of rapidly progressive ischemic ileo-colic itis.

**Discussion**

SM is a rare, idiopathic disorder characterized by non-specific chronic inflammatory infiltration and fibrosis in the mesenteric fat. This process also may lead to abdominal masses, the involvement of vessels and mesenteric calcification. Some possible pathogenetic factors have already been reported: trauma, infection, autoimmune, surgery, para-neoplastic response, and IgG4-related disease. However, because of its rarity, the pathogenesis of SM remains uncertain. SM is more frequent in males, especially during the 6th and 7th decades of life. The mechanical effect of the mesenteric mass encasing the intestines, blood vessels and lymphatics is at the basis of the most frequent symptoms: ab-

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*Figure 1. A, SM at CT scan: Increased attenuation (haziness) of the mesenteric root is depicted with associated “mass effect” and displacement of the small bowel loops. Small mesenteric adenopathies (arrows) can also be appreciated along with engorgement of the mesenteric veins (arrow-head) in the right flank. B, SM at histology: mesenteric inflammation with fibrosis and lipodystrophy is well evident (staining: hematoxylin-eosin; magnification: 100×).*
dominal pain, distension, diarrhoea, weight loss, and a palpable abdominal mass. These clinical features are caused by the mechanical effect of the mesenteric mass encasing the intestines, blood vessels, and lymphatics. Therefore, SM-related complications include small bowel obstruction and ischemia, and superior mesenteric vein thrombosis. Also, there is a number of reports which present PLE, as in the present case, in association with intestinal ischemia with perforation. As reported in this patient, anaemia, elevation of the erythrocyte sedimentation rate and C-reactive protein level, such as other non-specific abnormalities, may be found in SM. Because its clinical manifestations and laboratory findings are non-specific, the diagnosis of SM is difficult to perform and it can be definitively established only by surgical or imaging-guided biopsy. The most consistent histological findings are the presence of fibrous tissue and a chronic inflammatory infiltrate. According to literature we got the diagnosis by histology on mesenteric biopsies performed during surgery. The most common finding at CT scan is the presence of soft tissue mass in the small bowel mesentery (especially at the root of the small bowel mesentery). Sabate et al. then, indicated in two studies the CT sings that can be considered somewhat specific for SM: “fat ring sign” which is based on preservation of the densitometric values of fat nearest the mesenteric vessels a “tumoral pseudocapsule” which represents a band of soft tissue that separates the uninvolved mesentery from the inflamed fat, “calcifications” which are often contained in the lesion.

Most cases have been reported as having a self-limiting benign course. Nevertheless, in about 20% of patients, it is associated with significant morbidity and a chronic debilitating course, until few fatal cases. About treatment of SM there is no clear consensus. Most patients are managed conservatively and about a half of them may not require therapy. In effect, some authors recommend that asymptomatic or mild symptomat SM may be left untreated and observed. Some symptomatic cases improved after treatment with corticosteroids, azathioprine, cyclophosphamide, thalidomide and tamoxifen, though no randomized controlled trial has been performed on this specific issue. Surgical resection of the lesion is of little benefit, except in the case of bowel obstruction or perforation for which surgical resection is recommended. Others authors recommend aggressive immunosuppressive therapy with prednisolone and cytostatic agents to prevent progression of the lesion once the diagnosis is established.

In this patient, we used corticosteroids without obtaining a significant improvement of clinical and serological outcomes. Moreover, we had no enough time to try different therapeutic strategies due to the rapid worsening of general condition, which was greatly affected by PLE-induced malnutrition not responding to a supportive parenteral nutrition regimen. The pathogenesis of such PLE was probably a direct consequence of the SM. Stasis of the mesenteric veins most likely caused intestinal ischemia and reduction of portal blood flow. Most probably, inflammation and thickening of the mesentery could play a role in the pathogenesis of PLE and the sudden-onset abdominal pain was caused by the intestinal ischemia correlated to mesenteric veins involved in SM.

**Conclusions**

SM should be taken into account for the differential diagnosis in patients with abdominal symptoms and PLE. PLE in patients affected by SM should be considered a negative prognostic factor and it could be of importance in making a rapid decision for aggressive medical therapy. Many other cases and studies are needed to assess diagnostic criteria and the better therapeutic strategy for patients affected by SM.

**Conflict of Interest**

The Authors declare that there are no conflicts of interest.

**References**


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