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CASE REPORT

Ultrasonography-Based Management of Sclerosing Mesenteritis: From Diagnosis to Follow-Up

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Abstract: Sclerosing mesenteritis (SM) is an idiopathic disorder affecting mesentery, characterized by fat necrosis, chronic inflammation and fibrosis. The clinical presentation varies from asymptomatic cases to acute abdomen. The diagnosis is suggested by imaging but can be definitely established only by biopsies. In this paper, we discuss ultrasonography-based management of SM.

Keywords: sclerosing mesenteritis, mesenteric lipodystrophy, mesenteric panniculitis, retractile mesenteritis, ultrasonography

Introduction

Sclerosing mesenteritis (SM) is a rare, idiopathic, benign disease affecting typically the small bowel mesentery, and is characterized by fat necrosis, non-specific chronic inflammation and fibrosis.[1] It was recognized for the first time by Jura in 1924 and defined as "retractile mesenteritis";[2] over the years, several terms have been used to describe this disease, such as "mesenteric lipodystrophy",[3] "mesenteric panniculitis",[4] "xanthogranulomatous mesenteritis"[5] and "mesenteric fibrosis".[6] Finally, in 1997, Emory et al concluded that these different conditions represented the histologic variants of a single entity, named "sclerosing mesenteritis", the preferred term for referring to the entire disease spectrum.[1]

SM is an uncommon disorder, typically diagnosed in the fifth to seventh decade of life with an estimated prevalence of <1% and male preponderance.[7,8] Although the etiopathogenesis is still unclear, previous abdominal trauma or surgery, cancer, autoimmunity, infection, and medications have been linked to the development of SM.[9]

Clinical manifestation of SM could vary from asymptomatic cases, diagnosed incidentally on imaging,[7] to bowel obstruction, mesenteric ischemia, chylous ascites and protein-losing enteropathy, caused by mass effect on neighboring structures.[10–12] Patients commonly show abdominal pain, diarrhea, weight loss, palpable abdominal masses, abdominal tenderness, nausea, vomiting, anorexia, and fever.[8] Taking into account its non-specific manifestations, radiologic imaging plays a key role in the diagnosis of SM;[13] nevertheless, a definite diagnosis can be established only by surgical or imaging-guided biopsies.[11]

In the absence of clinical trials, the treatment of SM is empirical and ranges, according to clinical presentation, from simple observation to corticosteroids, immunosuppressive drugs, hormonal therapy or surgery.[9] The progression is often benign and self-limiting with a good long-term prognosis, although some cases of severe/fatal SM have been reported in literature.[10,11]

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In this paper, we report an ultrasonography-based management model of SM, aimed at providing a possible diagnostic-therapeutic approach that might be useful for gastroenterology research field.

Case Report

In 2018, a 63-year-old male (height 1.81 m, weight 63.5 kg) was admitted to our hospital with a recent history of diffuse abdominal pain associated with change in bowel habits (4 bowel movements/day without mucus and blood) and severe weight loss (about 25 kg in the last two years). In anamnesis, he reported only classic infectious diseases of childhood. He did not refer previous surgery or familiar history of gastrointestinal diseases and cancer. Physical exam revealed a palpable abdominal mass in mesogastrium. Laboratory findings were normal in absence of anemia or leukocytosis. No serological inflammatory status was observed. Ileocolonoscopy was normal. Ultrasound (US) showed a well-defined hyperechoic mass (measuring 41 x 110 mm) in the root of the small bowel mesentery with reactive sub-centimetric node in the context of the thickened mesenteric fat (shown in Figure 1). On the basis of clinical and ultrasonographical signs, the diagnosis of "sclerosing mesenteritis" was made. After the exclusion of a possible associated immuno-mediated disorders by serology, a whole-body computed tomography (CT) scan was performed to rule out the occurrence of a concomitant neoplasia. The CT confirmed the US diagnosis of sclerosing mesenteritis by highlighting the presence of a "misty mesentery", characterized by the increase of mesenteric fat density associated with enlarged inflammatory nodes. Finally, the patient underwent a US-guided biopsy of mesentery with a histology indicative for SM (shown in Figure 2). After a brief course of steroids (prednisone 50 mg), we decided to maintain treatment with tamoxifen 20 mg/daily associated with aspirin 100 mg/daily, in order to prevent a potential vascular complication, such as mesenteric ischemia. At US-based 6-months and 12-months follow-up visits, we recorded an improvement of US picture associated with weight gain (about 10 kg) and a stable remission of diarrhea and abdominal pain. After 2 years of follow-up, the patient is healthy and continues to report for US-based follow-ups. No additional CT scans were needed.

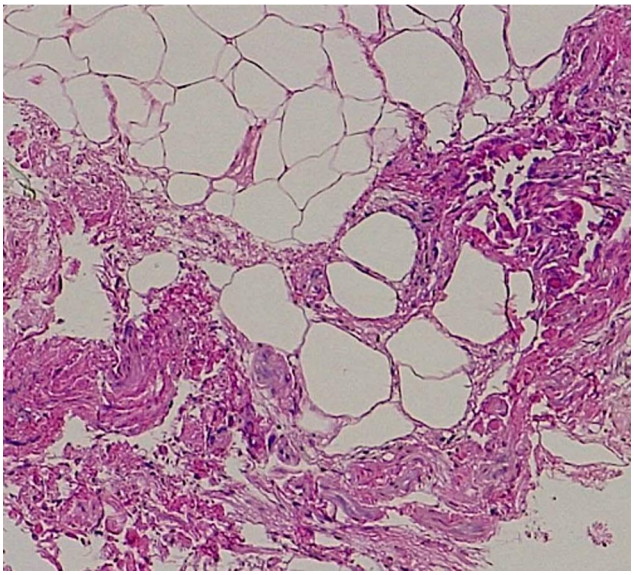

Figure 2 Sclerosing mesenteritis at histology. Fibrosis with dense collagen, fat necrosis, and chronic inflammation are evident.

Discussion and Conclusion

SM is a rare inflammatory disorder of unknown etiopathogenesis, characterized by fat necrosis, non-specific chronic inflammation and fibrosis of the small bowel mesentery.[1] On the basis of predominant histology, it is possible to

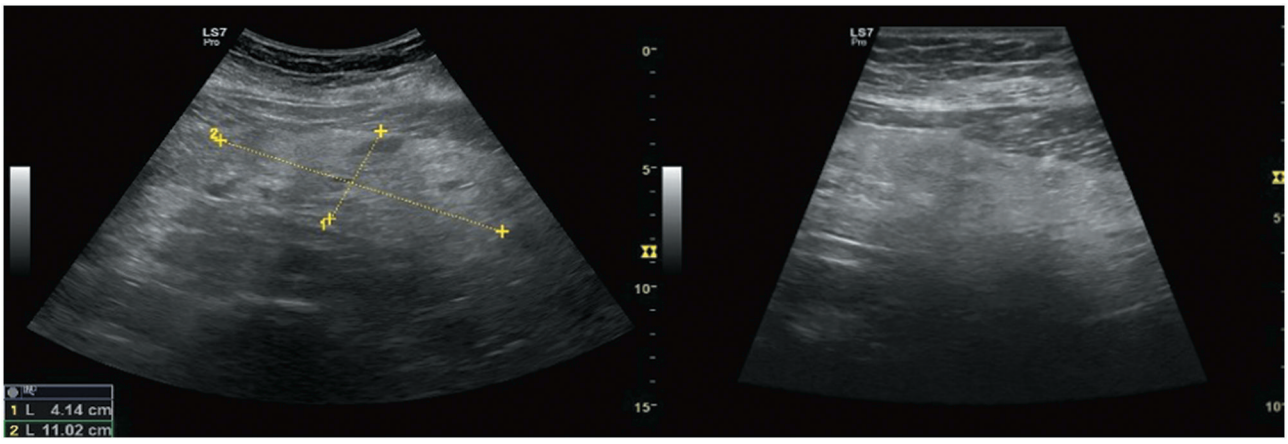

Figure 1 Sclerosing mesenteritis at ultrasonography. The thickened and hyperechoic mesentery is well evident.

divide SM into three categories: mesenteric lipodystrophy (fat necrosis), mesenteric panniculitis (chronic inflammation) and retractile mesenteritis (fibrosis).[1] SM has been hypothesized to be a progressive inflammatory process, moving from mesenteric lipodystrophy to retractile mesenteritis, triggered by a wide variety of stimuli. In this context, similarly to which occurs in atherosclerosis, the change of macrophages resident in mesenteric fat in foam-cells related to the upregulation of PPAR-γ and scavenger receptor expression might be the starting point in the development of mesenteric panniculitis.[14] The most common conditions associated with SM are previous abdominal trauma or surgery;[8] in this context, SM might be the result of an abnormal response to healing and repair of connective tissue after trauma or surgery in genetically predisposed subjects.[9] Moreover, SM has also been considered a paraneoplastic syndrome, since it is associated with different malignancies, such as lymphomas, chronic lymphocytic leukemia, myeloma, carcinoid, gastrointestinal cancers, renal cancer, and lung cancer.[15]

The rarity of this disease and the lack of clinical trials and practical guidelines have generated many difficulties regarding its definition, the correct establishment of the diagnosis and the possible therapeutic strategies. In 2007, Akram et al reported findings on a sample of 92 patients affected by SM.[10] After ten years, a systematic review of case reports by Sharma et al identified 192 cases of SM.[8] Considering these data, SM patients commonly show abdominal pain (75%), diarrhea/constipation (35%), palpable abdominal mass (33.3%), abdominal tenderness (25%), weight loss (25%), nausea and vomiting (20%), anorexia (15%), and fever (12.5%).[8,10] Laboratory findings are non-specific and consist of the increase in inflammatory markers, such as erythrocyte sedimentation rate and C-reactive protein, anemia, leukocytosis, and hypoalbuminemia.[8,10,11] Regarding the imaging features, the detection at CT scan of an increase in the mesenteric fat density, named "misty mesentery", should be considered the first indication, but non-specific mark of SM.[16] Coulier et al described five typical CT scan-signs for radiological diagnosis of SM: 1) hyperattenuating mesenteric mass; 2) mass-effect on neighboring mesenteric structures; 3) sub-centimetric nodes in the context of mesenteric fat; 4) "fat halo sign" with the hypoattenuation of fat surrounding the mesenteric nodes or vessels; 5) hyperattenuating pseudo-capsule.[17] Furthermore, US and magnetic resonance imaging (MRI) have also been performed for the diagnosis of SM, but there are fewer studies for these procedures than CT and lack of standardized criteria.[18–20] In this context, Roson et al retrospectively evaluated clinical, CT, and sonographic findings in 26 patients with mesenteric panniculitis, estimating that sonographic features of MS, such as a well-defined homogeneous and hyperechogenic mass, non-deviated vessels within the mass, and displaced bowel loops, well correlated with CT in 92% of cases.[18] However, at the moment, although US has been widely used in diagnostic work-up of intestinal diseases characterized by a secondary inflammatory involvement of mesentery (eg, Crohn's disease, colic diverticulitis, peritoneal carcinosis),[21–24] its use in SM is underestimated and under-investigated. Given this background, the clinical case we have reported could be a starting point for a wide and standardized use of US in a new clinical scenario.

There is no clear consensus regarding therapeutic strategies, thus the treatment for SM is empirical and varies according to clinical presentation and personal experience. Danford et al proposed a therapeutic algorithm, in which patients are divided into asymptomatic and symptomatic; while the first group are only observed, the second group (the symptomatic group) receive treatment. The first line therapy consists of the use of steroids (prednisone 40 mg/daily tapered slowly) in combination with tamoxifen 10 mg twice daily.[9] In case of poor improvement of symptoms, the second line therapy to be adopted includes the association between prednisone and azathioprine 2–2.25 mg/kg daily or simply thalidomide 200 mg daily.[9] Other medical drugs used successfully, but less frequently, in different case reports are colchicine,[25] 6-mercaptopurine,[26] methotrexate,[27] cyclophosphamide[28] and infliximab.[29] Surgery is reserved only in patients with complications, such as bowel obstruction.[9,12] In our case, the patient was effectively treated using steroids plus tamoxifen.

In conclusion, US could be a sensitive, reliable and non-invasive procedure for the diagnosis and the follow-up of SM. Multicenter diagnostic trials, including a wide SM population, are needed to define the actual diagnostic accuracy of US in SM.

Ethics Approval and Informed Consent

This case report has been prepared after obtaining an informed consent from the patient to publish the case details and the accompanying images published. No institutional approval was required to publish the case details.

Disclosure

The authors declare that they have no conflicts of interest to disclose.

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