AMERICAN JOURNAL OF DIAGNOSTIC IMAGING, 2022
VOL 8, NO. 1, PAGE 6–12
10.5455/ajdi.20210504093726

CASE REPORT

Open Access

Multifocal sclerosing mesenteritis: A rare case report and review of the literature

Sumit Datta[1], Nilu Malpani Dhoot[1], Amit Dutta[2], Panchali Datta[3]

[1]Department of Clinical Imaging and Intervention Radiology, Apollo Gleneagles Hospital, Kolkata, India

[2]Department of Obstetrics & Gynaecology, Gouri Devi Institute of Medical Sciences & Hospital, Durgapur, India

[3]Department of Anatomy, Diamond Harbour Government Medical College, Diamond Harbour, India

ABSTRACT

Sclerosing mesenteritis (SM) is a rare benign disorder characterized by chronic nonspecific inflammation involving the adipose tissue of the bowel mesentery. It is distinguished histologically and radiologically by varying amounts of fibrosis, chronic inflammation, and fat necrosis. Numerous associated conditions have been variably described, including malignancy, recent abdominal surgery, and systemic inflammatory diseases. Since SM lacks specific manifestation and typical signs, the patients are often misdiagnosed.

We present a rare case of multifocal involvement of SM in an elderly patient. A male patient presented with recurrent abdominal pain and diarrhea over a few months. Computed tomography (CT) and magnetic resonance imaging showed an ill-defined mass in the region of root of mesentery and in the left perinephric region. The lesions were discrete. CT-guided biopsy was done and the diagnosis of SM was confirmed. Medical management with steroids was initiated. The patient recovered symptomatically. Follow-up imaging at 6 months and 9 years revealed resolution of the lesion.

To the authors' knowledge, there is no case reporting multifocal presentation of SM. In this paper, we provide an overview of the literature relevant to the diagnosis, etiology, and management of this unique condition.

ARTICLE HISTORY

Received May 04, 2021
Accepted December 18, 2021
Published January 06, 2022

KEYWORDS

Sclerosing mesenteritis
imaging findings

Introduction

Sclerosing mesenteritis (SM) is a rare benign disorder characterized by non-specific inflammation of mesenteric fat [1]. Although this protean process has various stages, namely mesenteric panniculitis, mesenteric lipodystrophy, and retractile mesenteritis, SM is the accepted term for this less understood entity [1,2]. It usually involves the small bowel mesentery at its root, at times the mesocolon and rarely, the peripancreatic region, omentum, retroperitoneum, or the pelvis [3,4]. Its origin remains unknown, and its best to call it idiopathic presently, even though several etiological possibilities have been suggested, including infection, autoimmunity, trauma, and malignancy. SM usually presents in the fifth and sixth decades and is twice as common in men as in women [5]. The imaging features are varied without a single hallmark, finding and providing an unmistakable radiological diagnosis of SM. Histopathology mostly remains the gold standard in ascertaining the disease.

Case Report

A 70-year-old man came to our tertiary care hospital with progressive, recurrent abdominal pain and multiple episodes of diarrhea for 2 months. There was no history of abdominal surgery, trauma, or malignancy. On examination, tenderness was elicited in the epigastric and left paraumbilical region with a lumpy feeling. Routine laboratory investigations were unremarkable except for borderline leukocytosis (total count: 11,200/cu.mm) and mildly raised erythrocyte sedimentation rate (ESR) (24 in the first hour).

Ultrasonography (USG) revealed an ill-defined hypoechoic soft tissue mass in the left perinephric area without any vascularity. The kidneys appeared

Contact Nilu Malpani Dhoot ✉ drniludhoot@gmail com Department of Clinical Imaging and Intervention Radiology, Apollo Gleneagles Hospital, Kolkata, India.

© 2022 The Authors. This is an open access article under the terms of the Creative Commons Attribution NonCommercial ShareAlike 4.0 (https://creativecommons.org/licenses/by-nc-sa/4.0/).

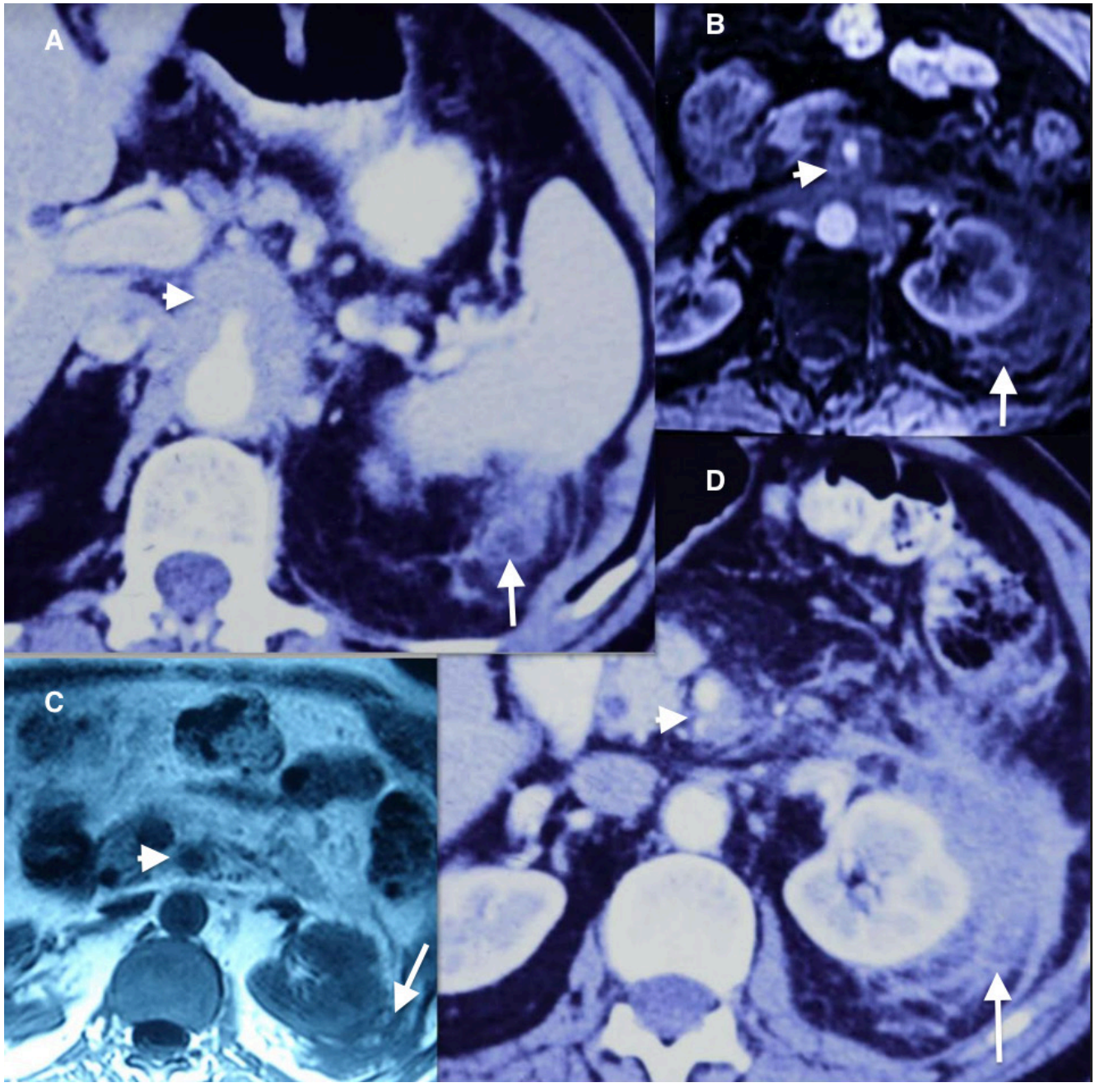


Figure 1. A and D. Axial sections contrast-enhanced CT abdomen depicts nonenhancing soft tissue mass encasing the SMA at root of mesentry (arrowhead). Another similar morphology mass in the left perinephric region (vertical arrow) with thickening of anterior and posterior perirenal fascia. B. Post gadolinium fat-saturated axial T1W MRI images reveals nonenhancing soft tissue mass encasing the SMA at root of mesentry (arrowhead). Another siilar morphology mass in left perinephric region (vertical arrow). C. T1W MRI axial sections showing soft tissue in the root of mesentry and in left perinephric region.

normal. No definitive diagnosis could be made on USG. Contrast-enhanced computed tomography (CT) of the abdomen showed ill-defined soft tissue density mass involving the root of the mesentery. This mass was seen coursing along the superior mesenteric artery (SMA), encasing the origin of the SMA and the coeliac axis. This mass was further seen extending to the retroperitoneum encasing the abdominal aorta by more than 270° circumference. The encased vessels were filled well with contrast. Apart from this mass, a similar morphology mass of about 5 × 4 cm was seen in the left perinephric region. The left gerotas fascia was thickened with perinephric fat stranding. The kidneys appeared normal otherwise. No calcification or cystic change was noted in the masses. These two masses were

discrete without any continuity between the two (Fig. 1A and B). No significant enhancement was seen in a post-contrast study on CT. The differential diagnosis of lymphoma or SM was suggested in CT. An unlikely differential of tuberculosis was also mentioned.

Magnetic resonance imaging (MRI) of the abdomen was obtained to narrow the differentials. It showed two definite lesions similar to CT. Ill-defined T1 and T2 iso-intense soft tissue masses were observed in the left perinephric region (Fig. 1C) along with the thickening of the anterior and posterior perinephric fascia. No obvious hydronephrosis or hydroureter was noted. The left kidney was unremarkable. Another ill-defined similar morphology mass was seen encasing the abdominal aorta and its major branches, including the origins of the coeliac axis and SMA. This mass extended along the SMA, involving the root of the mesentery. The caliber of the encased vessels appeared normal. The bowel loops and the rest of the abdominal organs were unremarkable. Both these lesions were not contiguous with each other and did not show any enhancement on contrast administration. Therefore, MRI revealed multifocal masses of similar morphology involving the retroperitoneum and the root of the mesentery. The possibility of inflammatory mesenteritis with atypical bifocal presentation and lymphoma was raised. Since the treatment of these two diseases was different, histopathological diagnosis was contemplated.

Trucut biopsy of the left perinephric soft tissue was performed under CT guidance, and local anesthesia and immunohistochemistry (IHC) were also done. On histopathology, xanthogranulomatous inflammation was diagnosed (Fig. 2A and B). IHC was negative for CD20 and CD3 essentially, excluding lymphoma. It was positive for CD68 (histiocyte), confirming an inflammatory lesion. With the above clinical–radiological and histopathological–immunohistochemical findings, it was inferred as a rare case of SM with an atypical bifocal presentation involving the root of the mesentery and the retroperitoneum.

The patient was treated conservatively with a low dose of steroids (prednisolone 4 mg per day). He improved considerably, symptomatically over time. Follow-up MRI performed at 6 months (Fig. 3A) and 9 years (Fig. 3B–D) confirmed remission.

Discussion

First described in 1924, SM is a rare pathology with an estimated prevalence of less than 1% [6,7]. The diagnosis of SM could be based on imaging or pathology or both. A prospective study by Daskalogiannak et al. [6] on 7,000 patients reported an incidence of 0.6%. This study, along with many other studies, diagnosed SM based on CT imaging [1,6,8]. Another study conducted by Kuhrmeier [9] used 712 autopsies as a diagnostic criterion, suggesting an incidence of SM of approximately 1% [9].

SM presenting in a 3-year-old patient is published in the literature, but generally it occurs in the fifth to seventh decade of life [8,10]. The age range of presentation of SM is 3–88 years with a mean age of 55 ± 19.2 years. The male population is more

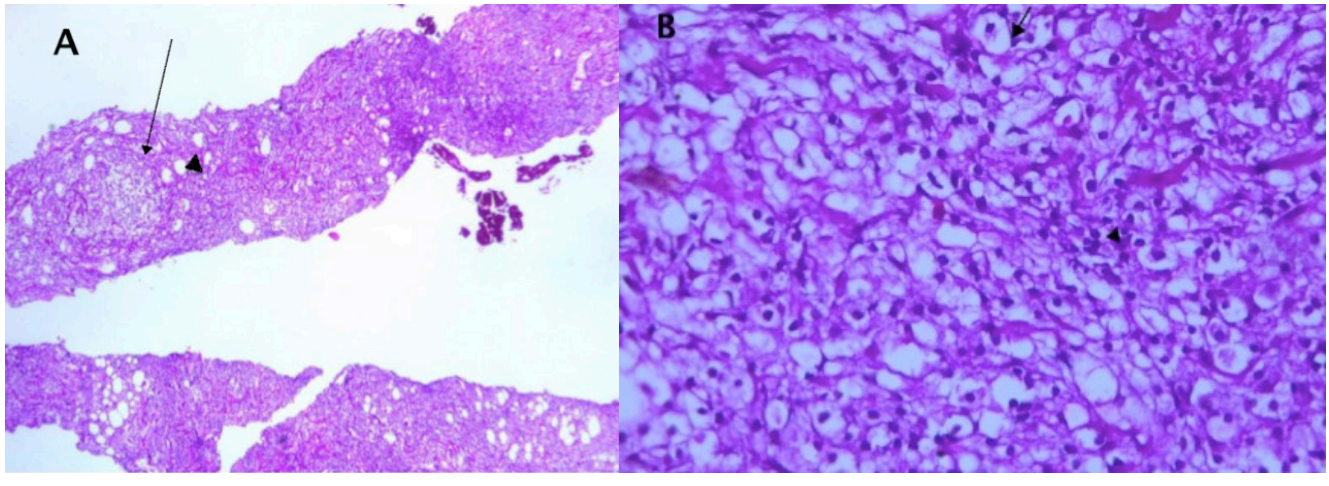


Figure 2. A. Scanner view (10×) shows cores of fibroadipose tissue (arrow head) infiltrated by sheets of foamy histiocytes, occasional multinucleated giant cells, ill-formed granulomata (arrow), and mixed inflammatory cells. There is no epitheloid cell granuloma or malignancy. B. High-power (40×) magnification shows ill-formed granuloma, sheets of histiocyste (arrow), and mixed inflammatory cells and giant cell (arrowhead).

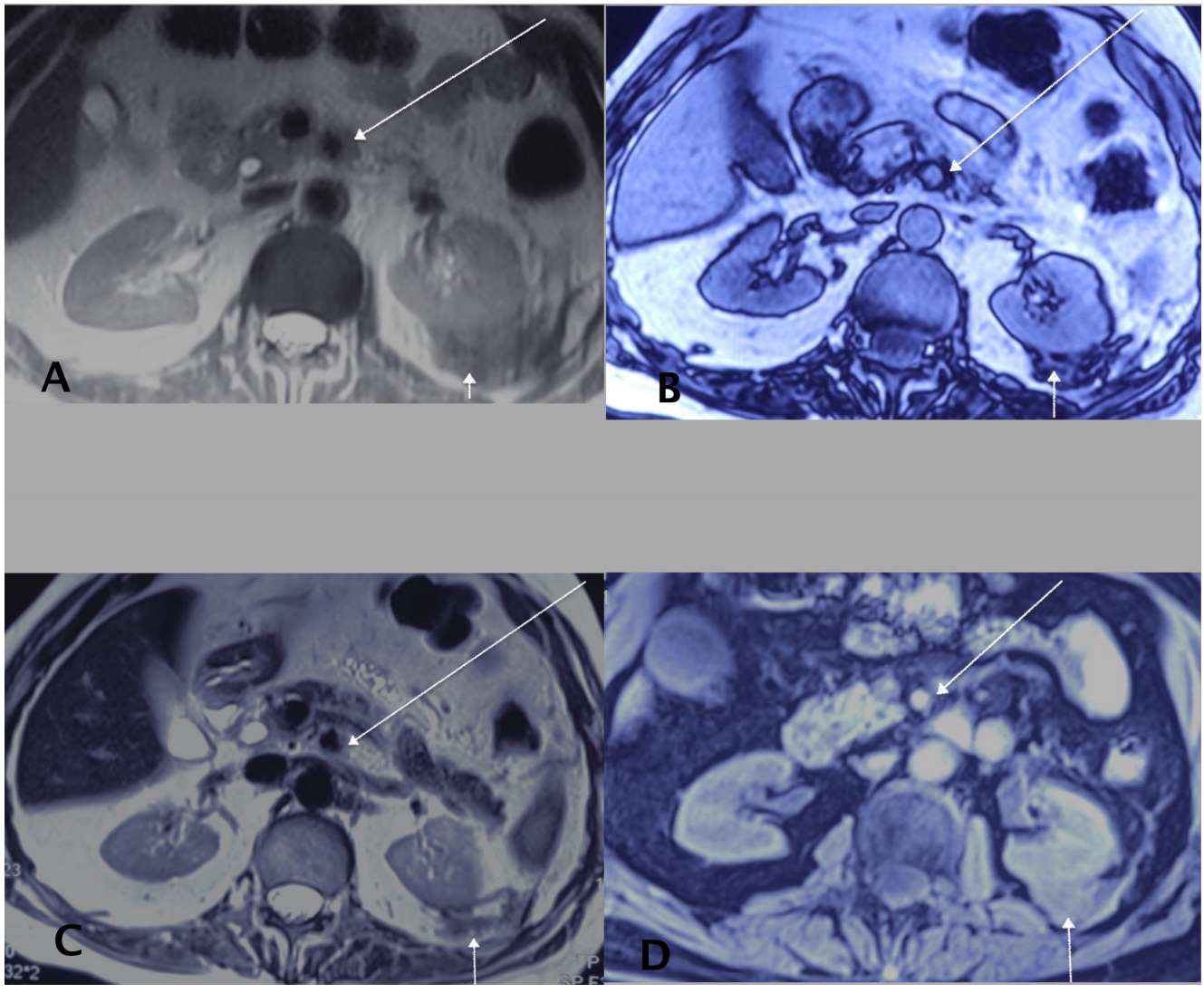


Figure 3. A. Follow-up at 6 months. T2WI MRI axial scan shows significant reduction in the soft tissue around SMA (oblique arrow), root of mesentery and perinephric region (vertical arrow). B. Follow-up at 9 years. Out-phase MRI section shows further reduction in the soft tissue near SMA and perinephric region. C. Follow-up at 9 years. T2W MRI section shows further reduction in the soft tissue near SMA and perinephric region. D. Follow-up at 9 years. Postcontrast fat-saturated MRI axial section shows further reduction in the soft tissue near SMA and perinephric region confirming remission.

affected than the female population suggested by most studies [8,11]. However, prospective studies by Daskalogiannak et al. [6] and Coulier [12] showed a slightly higher female predominance.

A clear understanding of the etiopathogenesis of SM is ideal. Multiple possible etiologies have been proposed for SM. They include abdominal surgery/trauma, autoimmune phenomenon, paraneoplastic process, and ischemia/infection [7]. The iatrogenic etiology of abdominal surgery/trauma has been implicated as one of the commonest causes of SM in the literature [13,14]. The association of SM with the prior abdominal operation was reported ranging from 24% to 53% [2,8,15]. Several authors have suggested autoimmunity as an etiology of SM. Case reports have been published, which indicate a possible association of SM to retroperitoneal fibrosis, Riedel's thyroiditis, and primary sclerosing cholangitis [8]. Since the clinical nature of SM is variable, a response to immunomodulatory medications possibly suggests an autoimmune process. Our patient also responded well to corticosteroids. Some studies have proposed SM to be an IgG4-mediated sclerosing disorder [15,16]. Whether SM is associated with paraneoplastic syndrome is debatable. Ogden et al. [14] raised the initial question, where he found a relationship between SM with lymphoma. This concern was continued in the literature with

contradictory results. Multiple studies showed a statistically significant association of SM to malignancies with a reported range of 8.9%–56% [16–19], whereas Gogebakan et al. [11] revealed no statistical difference between SM patients with underlying malignancy and controls. Infection has also been put forward as one of the etiologies of SM; however, very little evidence is published. Case reports have been presented with a history of chronic infections including tuberculosis, histoplasmosis, Whipple's disease, typhoid fever, and syphilis that could have possibly lead to SM [20]. None of these studies undoubtedly confirm an etiology consistent with SM. In the present scenario, it is best to assume SM as idiopathic.

Symptoms of SM are nonspecific. Patients may be asymptomatic in up to 15% of the disease [6]. The most frequent symptom is abdominal pain. A published systemic review described the following symptomatology percentage-wise. It revealed abdominal pain in 78.1%, fever in 26.0%, weight loss in 22.9%, diarrhea in 19.3%, vomiting in 18.2%, anorexia in 13.5%, constipation in 10.9%, bloating in 9.4%, malaise in 5.7%, nausea in 5.7%, pain with eating in 4.7%, and fatigue in 2.1% of SM patients [8]. The duration of symptoms varied from 24 hours to 10 years. Our patient presented with recurrent abdominal pain and diarrhea for 2 months. Elevation of inflammatory markers like ESR and C-reactive protein has been reported in 80% of the cases [4]. Our patient also had mildly raised ESR. No specific laboratory marker confirms the diagnosis of SM, making imaging the prime investigating modality.

On cross-sectional imaging, SM most commonly presents as a soft tissue mass with variable enhancement. The mass may show well-circumscribed or infiltrating margins. The lesion may contain central calcification; possibly, related to fat necrosis. Larger masses may demonstrate cystic features suggesting necrosis. Narrow bands of fibrosis may radiate from the mass, affecting the small bowel by retraction and shortening of the mesentery rather than direct invasion. The small intestine may become kinked or fixed, resulting in obstruction [4,21,22]. On CT, there are multiple diagnostic signs reported. The fat ring sign and the pseudocapsule sign have been widely used and reported [2,6]. The misty mesentery sign has been used for increased attenuation in the mesentery [23]. Coulier published five-sign CT imaging criteria specifically for mesenteric panniculitis [12]. These criteria were appreciated and used by many authors, subsequently in establishing SM [11,12]. These diagnostic findings were not histologically verified, although they do represent the nearest general standards available [24]. On MR, SM in the retractile fibrotic phase has a signal intensity pattern consistent with fibrosis—low to intermediate signal intensity on T1-weighted images and low signal intensity on T2-weighted images [25,26]. It may also have high signal intensity on T2-weighted images [27,28] likely dominated by myxomatous change or active proliferation of fibrosis accompanied by hypervascularity [22]. Our case had a infiltrative, non-enhancing mass in two different locations.

Despite imaging providing a confident suggestion of SM, definitive diagnosis is generally more acceptable after histopathology. In our case, CT-guided biopsy was done. The biopsy was done from the perinephric region as it was a more accessible site. It was a simple image-guided biopsy and yielded good material. Before the image-guided biopsy, the complications and the risk of renal/bowel injury, bleeding, and infection were explained to the patient. Our procedure did not have any complications. The histopathology and immunohistochemistry, coupled with imaging features, gave us a definite diagnosis of SM. The histopathological sample showed cores of fibroadipose tissue infiltrated by sheets of foamy histiocytes, occasional multinucleated giant cells, ill-formed granulomata (arrow), and mixed inflammatory cells. There was no epitheloid cell granuloma or malignancy.

Depending on the severity of the symptoms, treatment may be simple observation, as the disease is known to be self-limiting at times. Medical therapy with steroids, colchicine, tamoxifen, 6-mercaptopurine, antibiotics, azathioprine, methotrexate, progesterone, or immunosuppressive agents can be offered [5,6]. Surgical resection may be indicated for patients with complications such as bowel obstruction or perforation [29]. The overall prognosis of SM is usually very good with a benign and self-limiting course in most cases [15,30,31]. The pain disappears in 75% of the patients, and mass regresses by 66%, usually within 2 years [32]. In our patient, there was good symptomatic relief, and imaging remission on follow-up at 6 months and then 9 years.

To conclude, despite extensive research on SM, the definite etiology of the disease remains unclear. Multiple case reports on various imaging features and CT signs are available. There is no report presenting multifocal involvement of SM to our knowledge. Our case is rare and unique, where we

identified multifocal locations of SM involving the root of the mesentery and retroperitoneum. We followed-up with our patient for 9 years and demonstrated the resolution of the pathology on imaging.

Conflicts of interest

The authors have no conflicts of interest.

Consent for publication

Informed written consent was obtained from the patient for publication of this report and any accompanying images.

REFERENCES

[1] Emory TS, Monihan JM, Carr NJ, Sobin LH. Sclerosing mesenteritis, mesenteric panniculitis, and mesenteric lipodystrophy: a single entity. Am J Surg Pathol 1997; 21:392–8; doi:10.1097/00000478-199704000-00004

[2] Sabate JM, Torrubia S, Maideu J, Franquet T, Monill JM, Pérez C. Sclerosing mesenteritis: imaging findings in 17 patients. AJR Am J Roentgenol 1999; 172:625–9; doi:10.2214/ajr.172.3.10063848

[3] Ghanem N, Pacha G, Bley T, Kotter E, Langer M. MR findings in a rare case of sclerosing mesenteritis of the mesocolon. J Magn Reson Imaging 2005; 21:632–6.

[4] Horton KM, Lawler LP, Fishman EK. CT findings in sclerosing mesenteritis (panniculitis): spectrum of disease. Radiographics 2003; 23:1561–7; doi:10.1148/rg.1103035010

[5] Chawla S, Yalamarthi S, Shaikh IA, Tagore V, Skiafe P. An unusual presentation of sclerosing mesenteritis as pneumoperitoneum: case report with review of literature. World J Gastroenterol 2009; 15:117–20.

[6] Daskalogiannaki M, Voloudaki A, Prassopoulos P, Magkanas E, Stefanaki K, Apostolaki E, et al. CT evaluation of mesenteric panniculitis: prevalence and associated diseases. AJR Am J Roentgenol 2000; 174:427–31; doi:10.2214/ajr.174.2.1740427

[7] Green MS, Chhabra R, Goyal H. Sclerosing mesenteritis: a comprehensive clinical review. Ann Transl Med 2018; 6(17):336; doi:10.21037/atm.2018.07.01

[8] Sharma P, Yadav S, Needham CM, Feuerstadt P. Sclerosing mesenteritis: a systematic review of 192 cases. Clin J Gastroenterol 2017; 10:103–11; doi:10.1007/s12328-017-0716-5

[9] Kuhrmeier A. Mesenteric lipodystrophy. Schweiz Med Wochenschr 1985; 115:1218–24.

[10] Adachi Y, Mori M, Enjoji M, Ueo H, Sugimachi K. Mesenteric panniculitis of the colon. Review of the literature and report of two cases. Dis Colon Rectum 1987; 30:962–6; doi:10.1007/BF02554286

[11] Gogebakan O, Albrecht T, Osterhoff MA, Reimann A. Is mesenteric panniculitis truely a paraneoplastic phenomenon? A matched pair analysis. Eur J Radiol 2013; 82:1853–9; doi:10.1016/j.ejrad.2013.06.023

[12] Coulier B. Mesenteric panniculitis. Part 2: prevalence and natural course: MDCT prospective study. JBR-BTR 2011; 94:241–6.

[13] Jura V. Sulla mesenterite retratile e sclerosante. Policlinico (Sez Chir) 1924; 31:575.

[14] Ogden WW 2nd, Bradburn DM, Rives JD. Mesenteric panniculitis: review of 27 Cases. Ann Surg 1965; 161:864–75; doi:10.1097/00000658-196506000-00006

[15] Akram S, Pardi DS, Schaffner JA, Smyrk TC. Sclerosing mesenteritis: clinical features, treatment, and outcome in ninety-two patients. Clin Gastroenterol Hepatol 2007; 5:589–96; quiz 23–4; doi:10.1016/j.cgh.2007.02.032

[16] Van Putte-Katier N, van Bommel EF, Elgersma OE, Hendriksz TR. Mesenteric panniculitis: prevalence, clinicoradiological presentation and 5-year follow-up. Br J Radiol 2014; 87:20140451; doi:10.1259/bjr.20140451

[17] Badet N, Sailley N, Briquez C, Paquette B, Vuitton L, Delabrousse É. Mesenteric panniculitis: still an ambiguous condition. Diagn Interv Imaging 2015; 96:251–7; doi:10.1016/j.diii.2014.12.002

[18] Canyigit M, Koksal A, Akgoz A, Kara T, Sarisahin M, Akhan O. Multidetector-row computed tomography findings of sclerosing mesenteritis with associated diseases and its prevalence. Jpn J Radiol 2011; 29:495–502; doi:10.1007/s11604-011-0587-5

[19] Wilkes A, Griffin N, Dixon L, Dobbs B, Frizelle FA. Mesenteric panniculitis: a paraneoplastic phenomenon? Dis Colon Rectum 2012; 55:806–9; doi:10.1097/DCR.0b013e318252e286

[20] Vlachos K, Archontovasilis F, Falidas E, Mathioulakis S, Konstandoudakis S, Villias C. Sclerosing mesenteritis: diverse clinical presentations and dissimilar treatment options. A case series and review of the literature. Int Arch Med 2011; 4:17; doi:10.1186/1755-7682-4-17

[21] Lawler LP, McCarthy DM, Fishman EK, Hruban R. Sclerosing mesenteritis. Depiction by multodetectar CT and three dimensional volume rendering. AJR Am J Roentgenol 2002; 178:97–9.

[22] Levy AD, Rimola J, Mehrotra AK, Sobin LH. From the archives of the AFIP: benign fibrous tumours and tumour like lesions of the mesentery: radiologic-pathologic correlation. Radiographics 2006; 26:245–64.

[23] Mindelzun RE, Jeffrey RB Jr, Lane MJ, Silverman PM. The misty mesentery on CT: differential diagnosis. AJR Am J Roentgenol 1996; 167:61–5; doi:10.2214/ajr.167.1.8659422

[24] Coulier B. Mesenteric panniculitis. Part 1: MDCT—pictorial review. JBR-BTR 2011; 94:229–40.

[25] Badiola-Varele CM, Sussman SK, Glickserin MF. Mesenteric panniculitis. Findings on CT, MRI and angiography—case report. Clin Imaging 1991; 15:265–7.

[26] Kronthal A, Kang Y, Fishman E, Jones B, Kuhlman JE, Tempany CM. MR imaging in sclerosing mesenteritis. AJR Am J Roentgenol 1991; 156:517–9.

[27] Kakitsubata Y, Umemura Y, Kakitsubata S, Tamura S, Watanabe K, Abe Y, et al. CT and MRI manifestation of intra-abdominal panniculitis. Clin Imaging 1993; 17:186–8.
[28] Kobayashi S, Takeda K, Tanaka N, Hirano T, Nakagawa T, Matsumoto K. Mesenteric panniculitis: MR findings. J Comput Assist Tomogr 1993; 17:500–2.
[29] Yong HA, Sophia K, George S. Mesentery. In: Haaga JR (ed.). CT and MRI of the whole body, 5th edition, Mosby Elsevier, Philadelphia, PA, pp 1748, 2009.
[30] White B, Kong A, Chang AL. Sclerosing mesenteritis. Australas Radiol 2005; 49(2):185–8.
[31] Gonzalez Rodrigez CI, Cires M, Rubro T, Jimenez FJ, Garcia de Eulate J, Artondo MT. A case of mesenteric panniculitis. An Sist Sanit Navar 2008; 31(1):81–5.
[32] Reske M, Namiki H. Sclerosing mesenteritis. Report of two cases. Am J Clin Pathol 1975; 64(5):661–7.