



THE ROLE OF IMAGING IN DIFFERENTIATING SCLEROSING MESENTERITIS

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ABSTRACT

Sclerosing mesenteritis is a rare benign disorder characterized by nonspecific inflammation of the mesenteric fat. It is distinguished histologically by varying amounts of fibrosis, chronic inflammation, and fat necrosis. (1) Although this protean process has been known by a number of terms—including mesenteric panniculitis, mesenteric lipodystrophy, and retractile mesenteritis—*sclerosing mesenteritis* is the accepted term for this single clinicopathologic entity.(1,2) Definitive diagnosis of sclerosing mesenteritis requires histology, and surgical excisional biopsy is often needed for a complete analysis. Depending on the severity of symptoms, treatment may consist of simple observation or medical therapy with steroids, colchicine, orally administered progesterone, or immunosuppressive agents. Surgical resection may be indicated for patients with complications such as bowel obstruction or perforation.

Keywords: Mesentry, Histology, Computed Tomography(CT), Magnetic Resonance Imaging(MRI)

INTRODUCTION

This is a rare, slowly progressive condition of unknown origin, characterized by a chronic, nonspecific inflammation involving the adipose tissue of the small-bowel mesentery. When fibrosis is the dominant component, the disease is referred as retractile or fibrosing mesenteritis.(3) A well delineated, inhomogeneous fatty mass at the mesenteric root, envelopment of mesenteric vessels, absence of adjacent bowel loops involvement, which may or may not be displaced, and low-attenuation halo surrounding vessels (Fig. 1) are the characteristic CT features of mesenteric panniculitis.(4) Retractable mesenteritis appears on CT as an infiltrative soft-tissue mass with associated radiating linear strands of soft-tissue attenuation (Fig. 2). These features may mimic those of desmoid or carcinoid tumours. MRI may help in the differentiation by showing low signal intensity on both T1 and T2 images in fibrosing mesenteritis.(5) Calcification may be present in the necrotic central portion of the mass (Fig. 3).

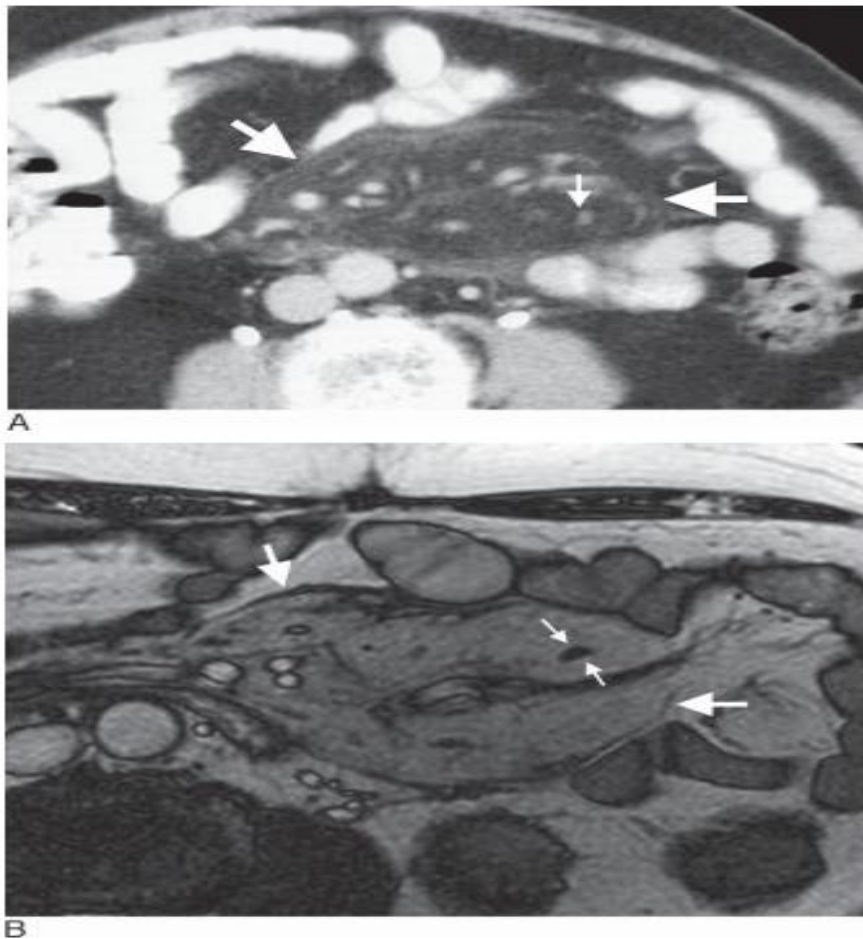


Figure 1: Mesenteric panniculitis. Enhanced CT (A) and true FISP MRI (B) in a patient who presented with abdominal pain show a well-delineated fatty mass (large arrows) extending from the root of the small-bowel mesentery toward the left abdomen, engulfing mesenteric vessels without distortion. Note the perivascular halo (small arrow).

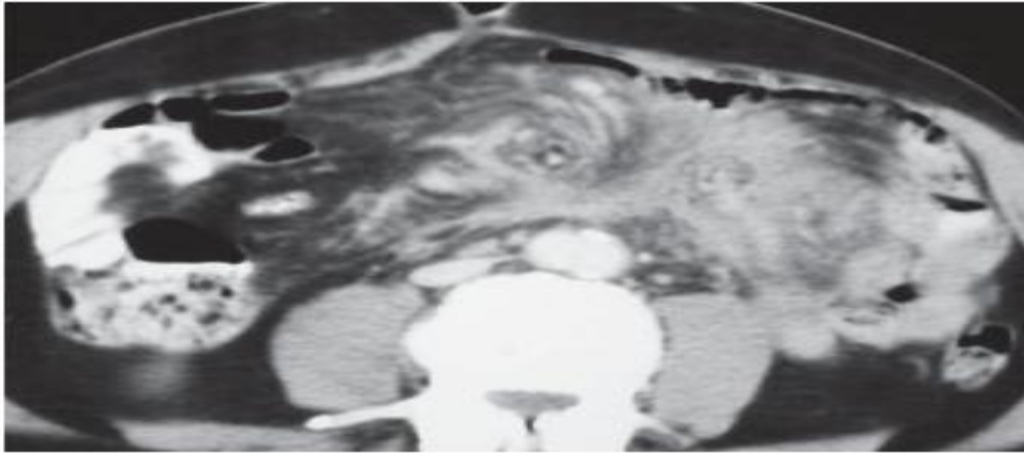


Figure 2: Fibrosing mesenteritis. Enhanced CT in a patient who presented with fever of unknown origin demonstrates a fibrofatty mesenteric mass with irregular borders surrounding mesenteric vessels. Strands of soft-tissue density are seen radiating from the mass to the adjacent mesenteric fat.



Figure 3: Fibrosing mesenteritis: CT appearances. Enhanced abdominal CT demonstrating a large, ill-defined, soft-tissue mesenteric mass with extensive calcification. Note retraction and thickening of the adjacent bowel loops.

Causes:

Sclerosing mesenteritis usually involves the small bowel mesentery at its root but can sometimes affect the mesocolon and, rarely, the peripancreatic region, omentum, retroperitoneum, or pelvis.(6,7) Its cause remains unknown, although several possibilities have been suggested, including infection, ischemia,

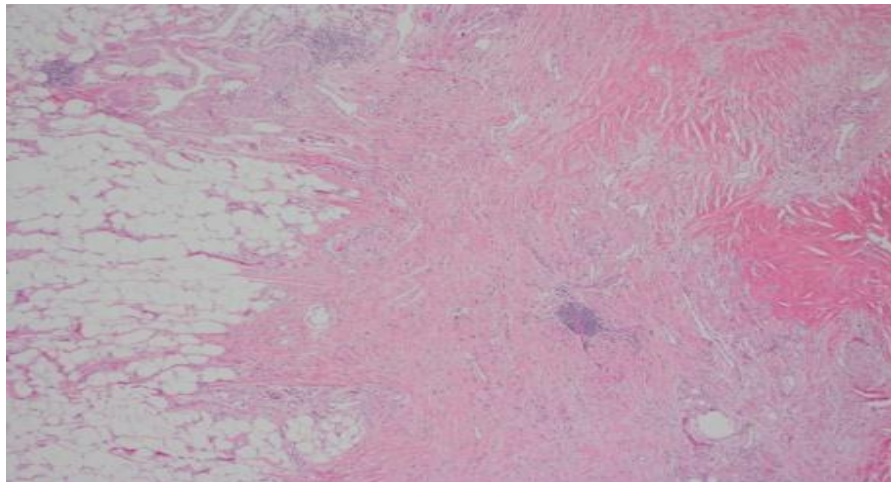
trauma, and vasculitis. Sclerosing mesenteritis is also associated with other idiopathic inflammatory disorders such as retroperitoneal fibrosis, Riedel's, thyroiditis, orbital pseudotumor, and sclerosing cholangitis. An association between prior abdominal surgery and sclerosing mesenteritis has been also reported. Furthermore, the disease is related to other factors, such as mesenteric thrombosis, mesenteric arteriopathy, drugs, thermal or chemical injuries, vasculitis, avitaminosis, autoimmune disease, retained suture material, pancreatitis, bile or urine leakage, hypersensitivity reactions, and even bacterial infection .(8,9) Other factors, such as gallstones, coronary disease, cirrhosis abdominal aortic aneurysm, peptic ulcer, or chylous ascitis, have also been linked to this disease .(10) More recent studies have shown a strong relationship between tobacco consumption and panniculitis.(9)

Clinical Presentation:

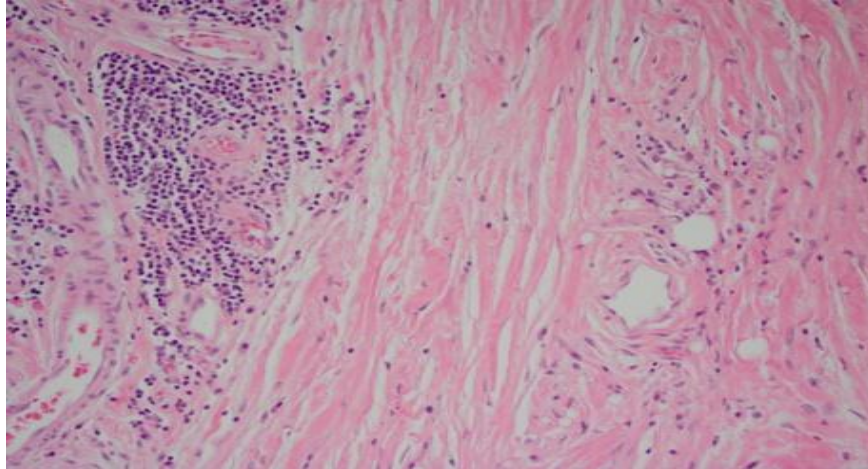
The average age of patients at presentation is 60 years, and the condition is more common in males than females. Patients may present with an abdominal mass, pain, bowel obstruction, bowel ischemia, or diarrhoea. Not uncommonly, sclerosing mesenteritis is an incidental finding in an asymptomatic patient.

Microscopic (histologic) description:

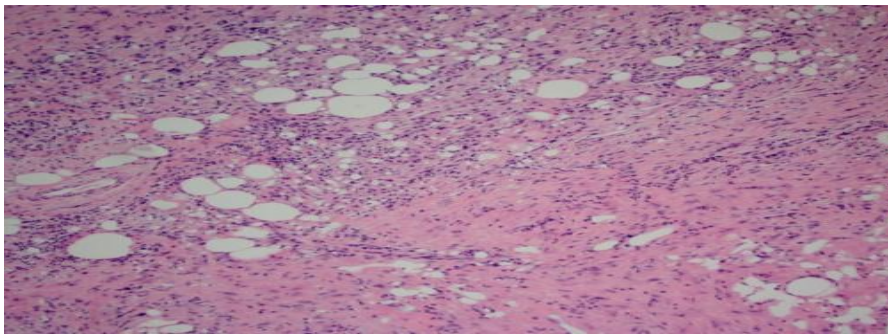
- ❖ Fibrosis with dense collagen, fat necrosis, chronic inflammation (especially around vessels) and variable focal calcification
- ❖ Minimal atypia, no or few mitoses
- ❖ IgG4 related cases lack fat necrosis and show obliterative phlebitis and increased inflammation



Fibrosis and chronic inflammation



Fibrosis and chronic inflammation, high power



Fibrosis, chronic inflammation and fat necrosis

Imaging finding

Thickness of the involved mesentery
Increased fat density
Fibrosis and enlarged lymph nodes
Fat ring sign
Pseudocapsule
Dilated or engorged mesenteric vessels
Well-defined or poorly defined mesenteric mass
Strand-like densities around the mesenteric vessels
Well-defined soft tissue nodules (usually less than 5 mm)
Bowel obstruction

Table 1: Radiographic Features of Sclerosing Mesenteritis

Computed Tomography:

The CT appearance of sclerosing mesenteritis reflects the underlying histology of the lesion (Fig. 5). Most commonly, it presents as a soft tissue mass with variable enhancement. The mass may show well-circumscribed or infiltrating margins , and lesions may contain central calcification, possibly related to fat

necrosis. Larger masses may demonstrate cystic features, suggesting necrosis. Enlarged mesenteric or retroperitoneal nodes may be found in association with sclerosing mesenteritis. Linear bands of fibrosis may radiate from the mass, affecting the small bowel by retraction and shortening of the mesentery rather than by direct invasion. The small bowel may then become kinked or fixed, resulting in obstruction; it may also be edematous and thickened owing to obstruction of lymphatics and veins. (7,11,12)

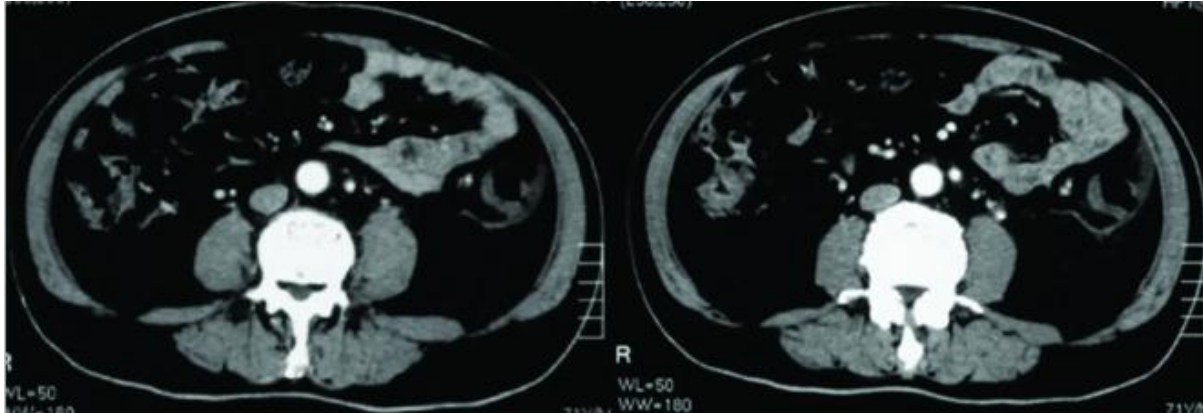


Figure 4: CT imaging of the abdomen showed thickening of the sigmoid colon wall which corresponded to a palpable firm cord-like mass (20 × 5 cm) in the left lower quadrant with tenderness on physical examination.

Magnetic Resonance Imaging:

On MRI, sclerosing mesenteritis has signal intensity patterns consistent with fibrosis—low to intermediate signal intensity on T1-weighted images, and low signal intensity on T2-weighted images.(25,26) Sclerosing mesenteritis may also have high signal intensity on T2-weighted images,(13,14) likely corresponding to the phase of the disease dominated by myxomatous change or active proliferation of fibrosis accompanied by hypervascularity.(12)

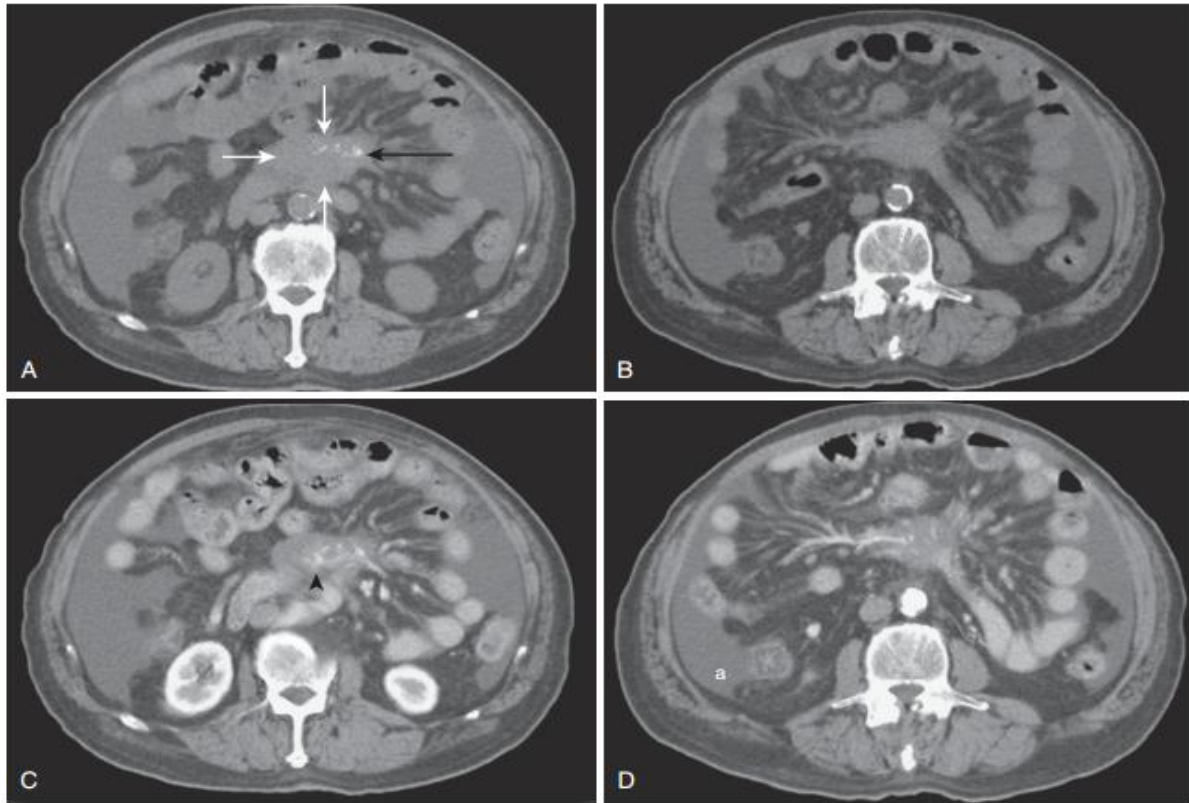


Figure 5: Sclerosing mesenteritis in a 70-year-old man. **A** and **B**, Noncontrast axial CT images show a well-circumscribed soft tissue mass (*white arrows*) in the small bowel mesentery, with coarse calcification (*black arrow*). **C** and **D**, On postcontrast images at the corresponding levels of **A** and **B**, the mass encases the mesenteric vessels, causing vessel engorgement in the leaves of the mesentery and ascites (*a* in **D**). The superior mesenteric artery (*black arrowhead* in **C**) is encased by the soft tissue mass. (Courtesy of Dr. Kwon Hyuan Ha, Asan Medical Center, Seoul, Republic of Korea.)

Diagnostic criteria:

The medical records were reviewed and if the diagnosis could be confirmed with histology or radiology, the patients were included. The patients with typical radiological appearance on CT were labelled MP and the histologically confirmed cases with atypical radiology were labelled SM. Sclerosing mesenteritis was considered histologically confirmed if the pathologist suggested SM or if the clinician concluded the inflammatory changes consistent with SM. A group consisting of two radiologists and two clinicians reviewed the CT examinations. The Coulier CT criteria were used for radiologic inclusion. [15,16] Mesenteric panniculitis was considered confirmed if three out of five criteria were present: (A) Fatty mass lesion in the small intestinal mesentery, (B) hyper attenuation of the fat, (C) lymph nodes in the fatty mass, (D) halo surrounding lymph nodes or vessels and (E) pseudo capsule. The images were graded using a scoring system

based on the five diagnostic criteria (A-E). Scores 0–3 were given for each criterion. Zero corresponded to no pathological findings and 3 to extensive findings. A total score of 3–4 represented mild, 5–10 moderate and 11–20 extensive radiological changes. Examples can be seen in Figs. Figs.6 and 7.



Figure 6: Moderate radiological SM with a well-defined fatty mass in the jejunal mesentery without mass effect (1p), hyperattenuation of the fat (3p), lymphnodes (2p), halo (2p) and a pseudocapsule (1p)

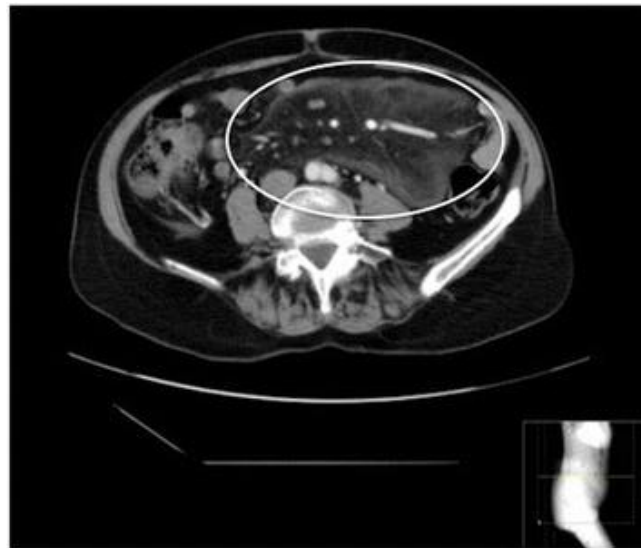


Figure 7: Extensive radiological SM with a large well defined fatty mass in the small intestine mesenteria (3p), marked hyperattenuation of the fat (3p), multiple lymphnodes (3p) with halo (3p) and a clear pseudocapsule (3p)

If the observed changes could be explained by adjacent pathology such as a neoplasm or other defined inflammation in the area (secondary mesenteritis) the patient was excluded.

Since the criteria for establishing the diagnosis based on findings on Magnetic Resonance Imaging (MRI) alone are not yet defined, patients exclusively examined with MRI were not included in this study.

Clinical scores:

Medical records were used to grade the severity of the symptoms. The patients were divided into four different categories: Asymptomatic patients (score 1), symptomatic but without systemic signs of inflammation (normal CRP and no history of fever) (score 2), symptomatic with systemic signs of inflammation (elevated CRP and/or fever due to SM were no other apparent explanation could be found) (score 3) and severe disease (chronic disease, complications, multiple hospitalisations or therapy resistant disease) (score 4).

Imaging Findings in Mesenteric Panniculitis:

Mesenteric panniculitis is a subtype of sclerosing mesenteritis consisting primarily of chronic inflammation. On CT this process may appear as subtle increased attenuation within the mesenteric fat, often at the root of the mesentery, with accompanying small nodes (Fig. 8). Mesenteric panniculitis may also exhibit a tumoral pseudocapsule consisting of a peripheral band of soft tissue attenuation that separates the normal mesentery from the inflammatory process, (2) and there may be spared fat around the vessels and lymph nodes, exhibiting the so-called fat-ring sign (17); this has been described as *segmental misty mesentery*. (18)

Although this CT appearance has been closely tied in the literature to the histologic diagnosis of mesenteric panniculitis, it must be emphasized that this radiologic appearance is nonspecific, and in clinical practice, a segmental misty mesentery is much more likely to result from more common causes, including edema, fibrosis, hemorrhage, or neoplasm (Fig. 9). (19,20,18) Therefore, the finding of a segmental misty mesentery should prompt a search for an adjacent inflammatory process (e.g., pancreatitis, appendicitis), vascular disorder (e.g., portal hypertension), neoplasm (e.g., primary malignancy, active or treated lymphoma), or relevant clinical history (e.g., prior abdominal surgery). Often the cause of a misty mesentery is unknown, and radiologic findings are stable over time and of no clinical consequence (Figs. 10 and 11). (18) In this clinical setting, a follow-up CT may be warranted to clarify the primary cause or exclude an underlying malignancy. In particular, when enlarged mesenteric nodes are evident, further clinical evaluation is prudent to exclude an early presentation of lymphoma or low-grade lymphoma.

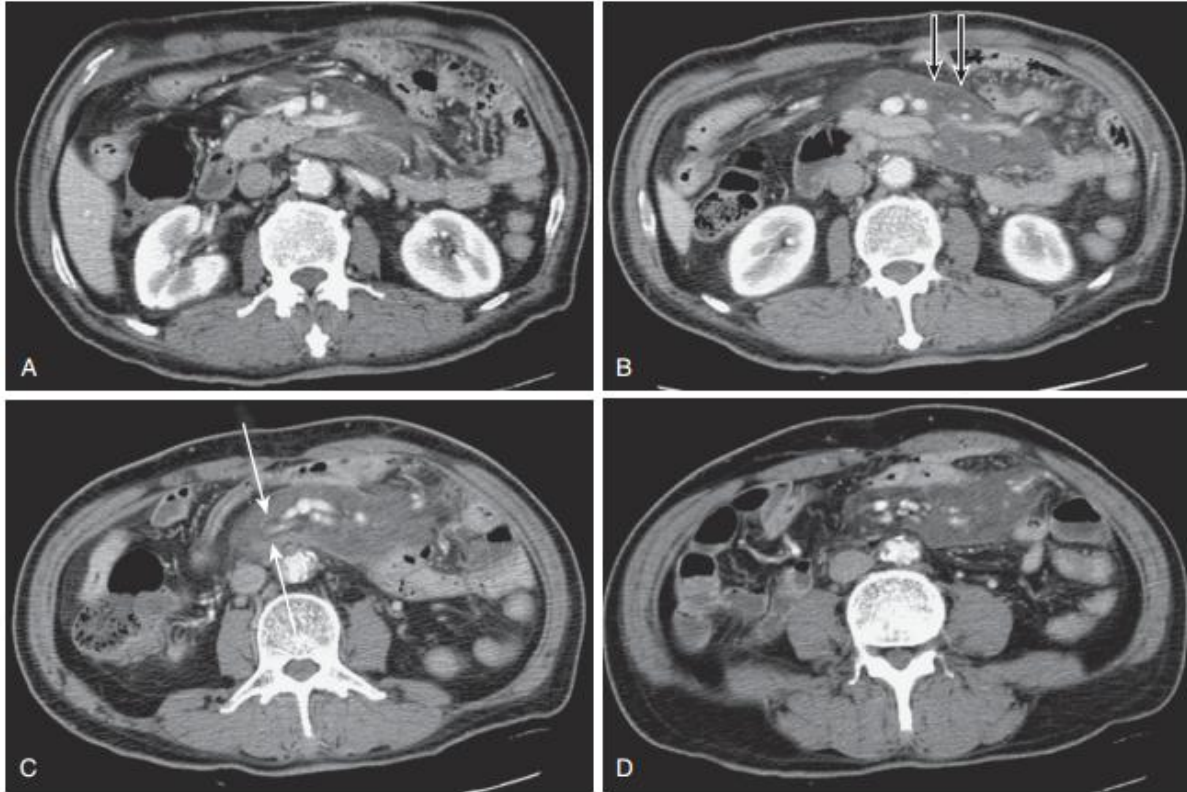


Figure 8: Mesenteric panniculitis in a 70-year-old man. A to D, Serial axial CT images show segmental increased attenuation in the small bowel mesentery, separated from the adjacent fat by a tumoral pseudocapsule (*arrows* in B). There is preservation of the fat around the vessels (“fat-ring” sign) that are surrounded by the increased density (*arrows* in C). (Courtesy of Dr. Kwon Hyuan Ha, Asan Medical Center, Seoul, Republic of Korea.)

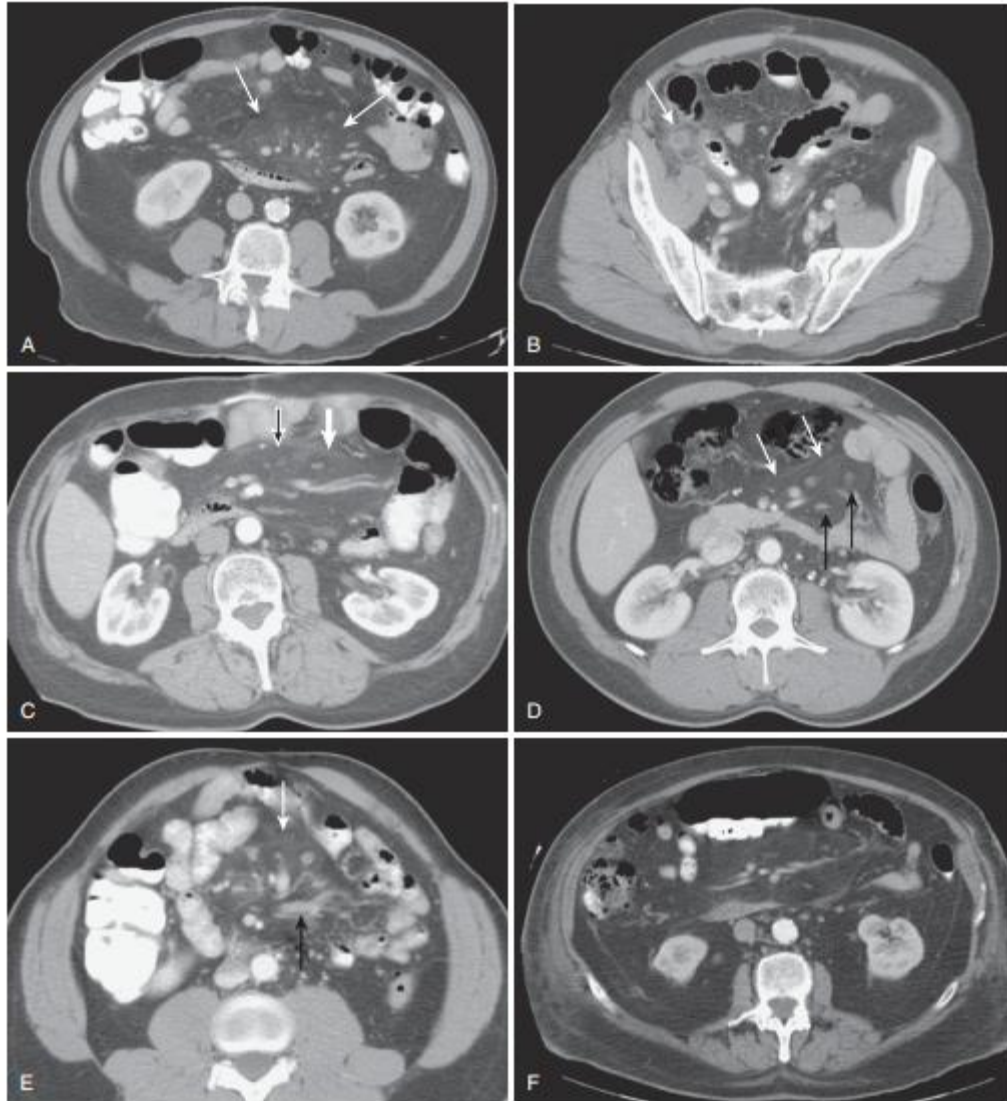


Figure 9: Mimics of mesenteric panniculitis. **A** and **B**, Axial postcontrast CT images show hazy infiltration of the small bowel mesentery (*arrows* in **A**) due to appendicitis (*arrow* in **B**). **C**, Band of soft tissue—the tumoral pseudocapsule (*arrows*)—demarcating a “misty mesentery” in a patient with Crohn’s disease. **D**, Hazy central small bowel mesentery (*white arrows*) with prominent lymph nodes (*black arrows*) in a patient who had been treated for testicular cancer. **E**, Misty mesentery (*white arrow*) with mesenteric vein engorgement (*black arrow*) in a patient previously treated for lymphoma. **F**, Hazy mesentery in a patient with pancreatitis. The fat-ring sign is well seen around the vessels and lymph nodes in **C** to **F**.

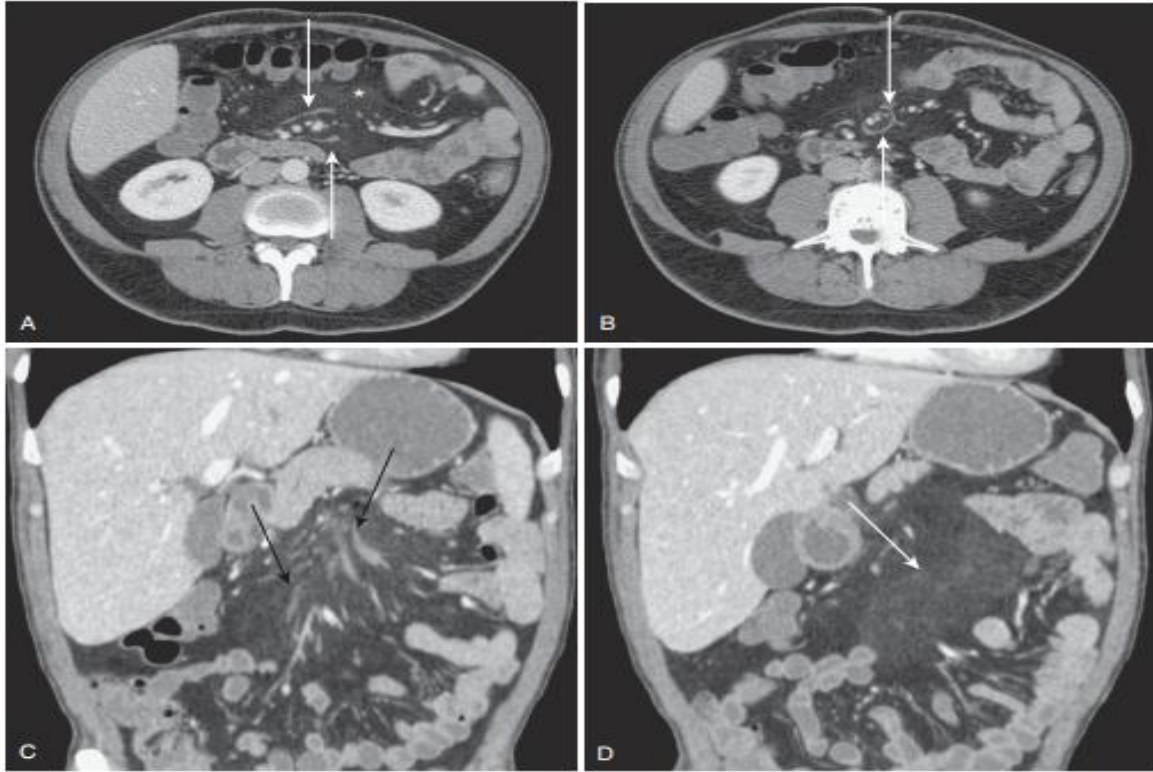


Figure 10: Idiopathic “misty mesentery.” **A** and **B**, Serial contrast-enhanced axial CT images show mild increased attenuation in the small bowel mesenteric fat (*asterisk*). Additional linear, branching soft tissue structures (*arrows*) are present within the mesentery, which are not identifiable as veins or arteries. **C**, These soft tissue structures (*arrows*) are better appreciated on a coronal reformatted CT image and presumably represent dilated lymphatics. **D**, A more anterior coronal reformatted CT image demonstrates the misty mesentery (*arrow*).

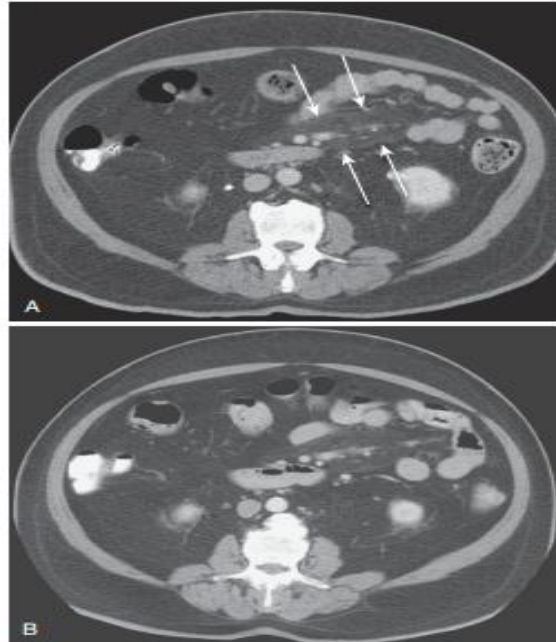


Figure 11: Idiopathic “misty mesentery” in a patient with chronic abdominal pain. **A**, Axial enhanced CT image shows increased attenuation in the small bowel mesentery with a tumoral pseudocapsule (*arrows*). **B**, Findings were relatively stable after 2 years.

DIFFERENTIAL DIAGNOSIS

Lymphoma and small bowel carcinoid are the main differential diagnostic considerations for a soft tissue mass within the mesentery. Lymphoma, however, does not usually calcify unless previously treated and does not often cause bowel ischemia. Lymphoma is also more likely to demonstrate discrete enlarged nodes. Serotonin production from small bowel carcinoids can produce a fibrotic retraction that may cause desmoplastic kinking of the small bowel, similar to the retraction observed on CT in cases of sclerosing mesenteritis. However, focal small bowel mural thickening or mass favors the diagnosis of carcinoid. Additionally, most small bowel carcinoid tumors are positive with somatostatin-receptor scintigraphy, and imaging with indium-111 pentetretotide can differentiate between the two diseases. Benign entities in the differential diagnosis include atypical infectious causes such as Whipple’s disease, which may affect the mesentery and retroperitoneum with lipogranulomatous inflammation. A diagnosis of Whipple’s disease can be made by using polymerase chain reaction to verify the causative bacillus. Actinomycosis can also cause an aggressive, infiltrative soft tissue process on CT and may affect the mesentery (see Fig. 12).

Additionally, Weber-Christian disease, a rare, systemic inflammatory disorder of fat, may have multifocal areas of fat necrosis and inflammation in the mesentery, identical to sclerosing mesenteritis.

Differentiation of Weber-Christian disease from sclerosing mesenteritis relies on the identification of other features of the former. Typically, these patients also have lower extremity skin nodules, fever, myalgia, arthritis, and arthralgia. Other benign fibrous lesions such as mesenteric fibromatosis, inflammatory pseudotumor, and extrapleural solitary fibrous tumor may also be considered in the differential diagnosis.

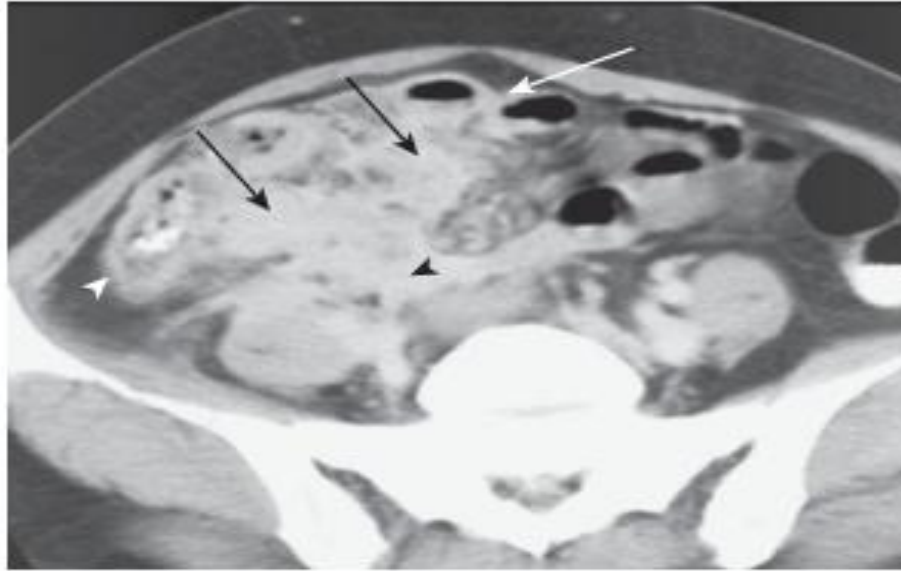


Figure 12: Actinomycosis. Axial CT scan of the pelvis shows heterogeneously and irregularly enhanced density (*black arrows*) within the mesentery and pericolic fat extending to the pelvic retroperitoneum (*black arrowhead*). Involved small bowel loops (*white arrow*) and ascending colon (*white arrowhead*) show wall thickening

TREATMENT

Mesenteric panniculitis resolves spontaneously in most cases, however, palpable masses may often be found between 2 and 11 years after diagnosis, especially in patients with associated comorbidity.(8) In such cases, several types of treatment have been proposed but no consensus has been established. In general, treatment has been reserved for symptomatic cases, incidental masses may be observed and left untreated. Therapy is individualized on a case basis. Treatment may be attempted with a variety of drugs including steroids, thalidomide, cyclophosphamide, progesterone, colchicine, azathioprine, tamoxifen, antibiotics and emetine, or radiotherapy, with different degrees of success.(21-23) Surgery may be attempted if medical therapy fails or in the presence of life threatening complications such as bowel obstruction or perforation.(24)

In conclusion, SM, a rare disease, should be considered in the differential diagnosis in malignancy and inflammatory disease of the intestine. Imaging studies, preferably CT, of the abdomen, including the bowel

and mesentery, should be performed. Finally, surgery, intraoperative findings and pathologic examination of the resected bowel segment and mesentery can confirm the diagnosis of SM. Although several drugs (steroids, colchicine, immunosuppressive agents, or orally administered progesterone) may be used to treat SM, surgery is preferred for those patients who are at the stage of fibrosis (retractile mesenteritis), particularly in those patients complicated by bowel obstruction.

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