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Pathology Page

Sclerosing mesenteritis

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A 55-year-old woman presented with a 1-week history of vomiting, watery diarrhea, and intermittent abdominal pain. She had occasional similar episodes of bowel habit changes without signs of gastrointestinal bleeding in the 3 months preceding presentation. No fever, night sweats, or remarkable body weight loss was present. The physical examination revealed no abnormalities. Laboratory investigations revealed leukocytosis (14790/μL, normal range: 3500~9900/μL) and mildly increased high-sensitive C-reactive protein (0.985 mg/dL, normal range: 0–0.3 mg/dL).

Abdominal computed tomography (CT) with contrast material revealed an ill-defined mesenteric mass-like lesion with surrounding fat attenuation [Figure 1a and b, yellow arrow] and multiple lymph nodes around the para-aortic region and mesenteric root. On suspicion of lymphoma or tuberculosis-associated mesenteric panniculitis, laparoscopic exploration was performed, which revealed diffuse mesenteric thickening and indurations forming an ill-defined bulging mass around the mesenteric root [Figure 1c] with an indurated texture and mild enlargement of the lymph nodes. Histopathological examination revealed fibrosis, disruption of fat, and infiltration of lipid-laden macrophages and lymphocytes [Figure 2], while lymph nodes were reactive. In general, the biopsy specimens showed mesenteric lipodystrophy and panniculitis with proliferative stromal cells, xanthomatous cells, and patchy lymphoid infiltrates. No evidence of vasculitis or prominent plasma cell infiltrates was found. Immunohistochemically, the stromal cells were positive for CD68 and vimentin, while negative for leukocyte common antigen (CD45), cytokeratin (AE1/AE3), cytokeratin (OSCAR), ALK-1, S-100, CD34, CD117, and β-catenin. IgG4-positive plasma cells were scanty. Acid-fast and Gomori methenamine silver stains revealed negative results. Taken together with the clinical presentation, sclerosing mesenteritis was considered. She received conservative treatment postoperatively and has had no recurrent symptoms after 20 months.

Sclerosing mesenteritis is a rare idiopathic inflammatory disorder with a nonspecific clinical presentation and no consensus on treatment. It was first described in 1924 and is characterized by various processes including nonspecific inflammation, fibrosis, adipose necrosis, and degeneration [1]. Based on the variable degrees of fibrosis, chronic inflammation, and fat necrosis, the entity might be called sclerosing (retractile) mesenteritis, mesenteric panniculitis, and mesenteric lipodystrophy [2], which are synonymous with Pfeifer–Weber–Christian disease, xanthogranulomatous mesenteritis, liposclerotic mesenteritis, inflammatory pseudotumor, mesenteric lipogranuloma, systemic nodular panniculitis, and misty mesentery. The pathological differential diagnosis includes fibrosis and chronic inflammation due to foreign material, desmoplastic metastatic carcinoma, mesenteric fibromatosis, sarcoma, retroperitoneal fibrosis, lymphoma, inflammatory pseudotumor, reaction to an adjacent cancer or chronic abscess, and Whipple disease [2].

It most commonly affects the small-bowel mesentery and is diagnosed primarily during the sixth to seventh decades of life [3]. Most symptoms are caused by the mesenteric mass encasing the bowel, blood vessels, and lymphatics, resulting in abdominal pain, bowel obstruction, ischemia, and diarrhea. Blood tests are usually normal, although elevation of the erythrocyte sedimentation rate, neutrophilia, or anemia can occasionally be seen [4]. The pathogenesis of sclerosing mesenteritis is unclear, although several mechanisms have been postulated, including previous abdominal surgery or trauma, autoimmunity, paraneoplastic syndrome, ischemia, and infection [5]. Some authors have shown that a subset of patients have abundant infiltrates of IgG4-positive plasma cells and have proposed that this disease could be an IgG4-related sclerosing disorder [3,6]. Our patient, however, exhibited scanty IgG4-positive plasma cells in the biopsy specimen and had no personal or family history of autoimmune disorders. Postoperative tests for antinuclear antibody,

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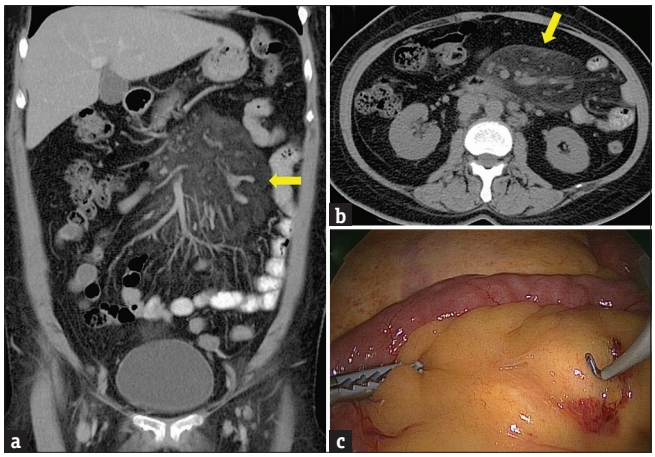

Figure 1: Different views from abdominal computed tomography with intravenous contrast showing increased density of the central mesenteric fat, forming an ill-defined mesenteric mass-like lesion, with mild lymphadenopathy around the mesenteric root (a and b, yellow arrow); laparoscopic exploration reveals diffuse mesenteric thickening and indurations (c)

serum antineutrophil cytoplasmic antibodies, anti-SSA/Ro, anti-SSB/La, and rheumatic factor also failed to show any abnormality.

Dual-phase abdominal CT scan is the most sensitive imaging modality for sclerosing mesenteritis, and its appearance varies from increased attenuation to solid soft-tissue masses. Two radiological findings, a "fat ring sign" and a "tumor pseudocapsule," are considered specific for sclerosing mesenteritis [3,7]. Magnetic resonance imaging findings appear to be similar to CT, but a combination of the two has been suggested as having a higher sensitivity in the detection of sclerosing mesenteritis [8]. The differential diagnosis includes lymphoma, carcinomatosis, fibromatosis, amyloidosis, mesenteric edema, pancreatitis, inflammatory myofibroblastic tumor, and liposarcoma. Although radiological characteristics are helpful, histological proof is essential for definitive diagnosis, especially with an atypical clinical and radiological appearance. Beyond diagnostic sampling, surgery is usually not necessary. Treatment is usually empiric and depends on the severity and type of individual symptoms. The lesion generally has a good prognosis. However, significant morbidity or a chronic course occurs in about 20% of patients [3]. Long-term follow-up is recommended.

Declaration of patient consent

The authors certify that they have obtained an appropriate patient consent form. In the form, the patient has given consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

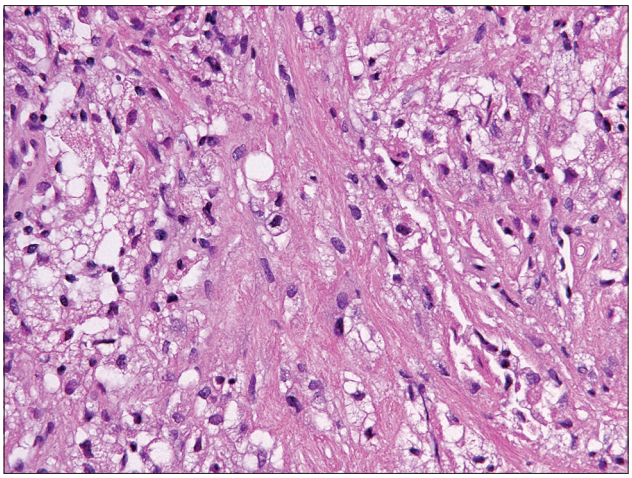

Figure 2: Microscopically, proliferative stromal cells, foamy macrophages, and lymphocytes are present in the mesenteric fat with mild fibrosis (H and E, ×400)

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Conflicts of interest

There are no conflicts of interest.

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