Multiple Mesenteric Panniculitis as a Complication of Sjögren’s Syndrome Leading to Ileus

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Abstract

Mesenteric panniculitis (MP) is a benign fibroinflammatory process characterized by the presence of fat necrosis, chronic inflammation and fibrosis in the mesentery. Although various causal factors, such as malignancy, chronic inflammatory conditions and autoimmune processes, have been identified, the precise etiology remains unknown. We herein report a rare case of MP accompanying Sjögren’s syndrome in which a mass lesion and intestinal stenosis were observed simultaneously. This condition led to ileus, which was effectively treated using prednisolone.

Key words: mesenteric panniculitis, Sjögren’s syndrome, Weber-Christian disease, ileus

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Introduction

Mesenteric panniculitis (MP) is a relatively rare, chronic, nonspecific inflammatory disorder that can be difficult to diagnose (1). A careful assessment is important for accurately evaluating MP because there are several diseases with similar nonspecific symptoms. The differential diagnosis of MP includes colon cancer, metastatic colorectal cancer, lymphoma, peritoneal carcinomatosis, lipoma, liposarcoma, ischemic colitis, diverticulitis and inflammatory bowel disease (1, 2). The pathophysiology of this disease remains unclear despite its association with abdominal trauma or prior surgery, malignancy, vasculitis, ischemia, infection, allergic diseases and autoimmune conditions (3). The effectiveness of treatment with immunosuppressants, such as prednisone, azathioprine and cyclophosphamide, suggests the possible involvement of an immune mechanism in the pathogenesis of MP (4). In fact, MP has been reported in combination with autoimmune diseases, such as systemic lupus erythematosus (SLE), minimal change nephrotic syndrome and autoimmune hemolytic anemia (5-7).

In the present case, MP was accompanied by Sjögren’s syndrome (SS). Although rare, some cases of SS occurring in combination with panniculitis have been reported. However, the onset of SS in combination with MP is extremely rare; only one case report has been published involving a patient with human T-lymphotropic virus type-1 (HTLV-1) infection (8, 9). In this study, we report a case of MP associated with SS in which intestinal stenosis and a mass lesion were simultaneously detected.

Case Report

A 70-year-old woman underwent an examination in June 2007 due to the development of dryness in the oral cavity. A subsequent blood test revealed positivity for anti-SS-A antibodies, anti-SS-B antibodies and rheumatoid factor, leading to a diagnosis of SS.

The patient developed loss of appetite in July 2007, with a fever of over 38°C and severe abdominal pain in the left upper quadrant in early August 2007. Hence, she was urgently admitted to our hospital. Blood tests performed on admission showed a significant inflammatory response, with a white blood cell count of 16,860/mm³, C-reactive protein level of 14.8 mg/dL and decreased albumin level. Various

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Abdominal computed tomography (CT) revealed a mass lesion of 4 cm in the ileocecal area (Fig. 1A) and an increased concentration of mesenteric fat around the splenic flexure of the colon (Fig. 1B). Colonoscopy disclosed a large tumorous mass covered with normal mucosa in the ileocecal area, resembling a submucosal tumor (Fig. 2). In addition, circumferential mucosal redness, edema and luminal narrowing at the splenic flexure were observed. A tissue biopsy showed only nonspecific inflammatory findings. A barium enema performed three days after the colonoscopy did not visualize the oral side of the ileocecal region due to obstruction by the tumor. Progressive circumferential stenosis of the splenic flexure measuring 4 cm long, with an irregular border, was noted (Fig. 3).

No improvement was observed in the inflammatory response, despite treatment with fasting, transfusions and intravenous antibiotics. Two weeks after admission, abdominal distention developed and a high-pitched metallic sound was audible, indicating possible ileus due to stenosis of the splenic flexure. Therefore, bowel decompression was performed by inserting a transanal ileus tube up to the rostral portion of the splenic flexure. Abdominal surgery had not been previously attempted, and no definitive diagnosis was reached, even after a detailed examination. Consequently, on day 32 in the hospital, a laparoscopic biopsy of the mesentery with ileostomy was performed. Observation of the abdominal cavity indicated significant thickening of the mesentery in the ileocecal area and at the splenic flexure. The results of the mesenteric biopsy showed fibrosis of the adipose tissue, vascular proliferation and the infiltration of lymphocytes and neutrophils, leading to a diagnosis of MP (Fig. 4). Prednisolone (PSL; 30 mg/day) was administered from day 41, after which the MP and associated clinical symptoms improved dramatically. No relapse has occurred in the four years since the cessation of PSL therapy.

**Discussion**

Two findings in this case made the differential diagnosis difficult: the presence of discontinuous lesions in the ileocecal area and at the splenic flexure and the MP-like inflammatory findings in the splenic flexure on various tests, visualized as a mass lesion in the ileocecal area. Although MP was suspected as the most probable diagnosis based on the
CT findings and lack of improvement with antibiotic treatment and fasting, an intra-abdominal laparoscopic study was conducted and a mesenteric biopsy was performed to aid in confirming the definitive diagnosis.

No standard therapy for MP has yet been established. Conservative therapy, such as bowel rest, PSL, thalidomide, cyclophosphamide, progesterone, colchicine, azathioprine, tamoxifen, antibiotics, emetine and radiotherapy, is often used (10, 11). In addition, surgical resection is performed in cases in which conservative therapy results in no improvement, such as patients with severe obstructive impairment and organic lesions, including fibrosis (12). MP may be surgically treated when it is difficult to differentiate the lesion from malignancy, relapse is frequent or in cases of massive melena. Several reports have stated that bowel resection is effective in cases involving MP associated with intestinal stenosis; however, in most of these cases, the surgeries were performed before the diagnosis of MP. Although few reports are available on the use of PSL treatment for intestinal stenosis, this drug was particularly effective in our case, suggesting that PSL may be the first-choice treatment option in cases of intestinal stenosis secondary to MP.

Weber-Christian disease (WC), an idiopathic condition associated with frequent relapse characterized by the presence of systemic symptoms, such as fever and subcutaneous induration, was also considered in the differential diagnosis in this case (13). When mesenteric panniculitis develops as a consequence to WC, the condition is often resistant to steroid therapy and carries a poor prognosis. On the other hand, some cases of SS occurring in combination with WC have been reported (8, 14, 15); in these cases, steroid therapy was effective and the prognosis was good. In our case, WC was ruled out because no signs of subcutaneous panniculitis characteristic of WC were observed. The development of SS in combination with MP alone, without WC, is extremely rare.

We herein reported a case of MP associated with SS in which intestinal stenosis and a mass lesion were detected simultaneously. To our knowledge, this is the first report of an extremely rare condition, namely multiple MP as a complication of SS presenting with intestinal stenosis and ileus.

The authors state that they have no Conflict of Interest (COI).

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