Lupus mesenteric enteritis is an uncommon condition in systemic lupus erythematosus (SLE) patients with acute abdominal pain. We describe a 28-year-old woman with a 3-day history of watery diarrhea and intermittent vomiting. Abdominal ultrasonography revealed thickening of the intestinal wall and edema with the lumen filled with fluid in this patient with nonspecific symptoms. These ultrasonographic findings led the physician to perform an abdominal computed tomography (CT) scan, which revealed typical findings of lupus mesenteric enteritis: intestinal wall thickening with target signs and prominent engorgement of mesenteric vessels with a palisade pattern. After high dose methylprednisolone (1 g/day intravenously) was administered for 3 days, the patient’s symptoms markedly improved. Additionally, resolution of the mural thickening was observed within 1 week by abdominal ultrasound examinations. Our experience indicated that abdominal ultrasonography can be important in the correct diagnosis of lupus mesenteric vasculitis (LMV).

Keywords: abdominal computer tomography; abdominal sonography; acute abdomen; lupus mesenteric vasculitis; systemic lupus erythematosus

1. Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease. It is characterized by immune dysregulation that results in the production of autoantibodies, generation of circulating immune complexes, and activation of the complement system. Lupus mesenteric vasculitis (LMV) results from immune complex deposition and thrombosis of the intestinal vessels in SLE. LMV is an uncommon condition in SLE patients with acute abdominal pain. Symptoms of LMV are usually nonspecific and abdomen ultrasonography is very useful for early and correct diagnosis of this uncommon cause of abdominal pain in patients with SLE. We describe a case of LMV to emphasize the importance of abdominal ultrasonography in diagnosis of this uncommon condition in SLE patients with abdominal pain.

2. Case Report

A 28-year-old woman presented to our outpatient department with a 3-day history of watery diarrhea and intermittent vomiting. When questioned further, she also complained of diffused abdominal pain. Her medical history was notable for SLE, pleuritis, nephritis, and left middle cerebral artery infarction 7 years earlier, with no evidence of recurrence on follow-up. Her current medications included per os azathioprine 50 mg qd, prednisolone 15 mg qd, and warfarin 2.5 mg qd. Her most recent monthly serum test showed anti-double stranded DNA antibody 368,420, and 622 IU/mL (positive: >139 IU/mL), C3
(normal: 90–180 mg/dL)/C4 (normal: 10–40 mg/dL): 51/11, 55/11, and 46/9.3 mg/dL. The daily urine protein was 0.65 mg. She was asymptomatic until 3 days prior to this presentation. Symptoms of SLE flare-up, such as molar rash, photosensitivity, oral ulcers, arthritis, serositis, seizure, and psychosis were not observed when she presented to our hospital. A physical examination demonstrated tenderness over the periumbilical area, and a soft abdomen without rebounding pain. Laboratory findings at presentation revealed a white blood cell count of 8700/mm³ (neutrophils/lymphocytes: 94/3), hemoglobin 13.8 mg/dL, platelets 13,800/mm³, and C-reactive protein 2.53 mg/dL. Levels of amylase (116 IU/L), Gamma-glutamyl transpeptidase (γ-GT) (12 U/L), alkaline phosphate (23 U/L), alanine aminotransferase (18 IU/L), and aspartate aminotransferase (18 U/L) were normal. Her SLE activity index score was 8 (proteinuria: 0.645 g/24 hours, low complement, increased DNA binding). An abdominal X-ray demonstrated a gasless ileus (Fig. 1), and ultrasonography showed thickening of the small intestinal wall and edema with the fluid-filled lumen (Figs. 2 and 3). The abdominal computed tomography (CT) scan is shown in Fig. 4, which revealed the typical manifestations of LMV: intestinal wall thickening with target signs (an enhancing inner and outer rim with hypoattenuation in the center) in several segments of the small intestine. The inner layer and outer layer of the small intestine are indicated by arrowheads and arrows, respectively. Edema change of the small intestine wall is noted. The oval structure seen on abdominal sonography is a cross-section of the small intestine. The inner layer and outer layer of the small intestine are indicated by arrowheads and arrows, respectively. A marked thickening and hypoechoic change of the small intestinal wall were also found, which implied edema of the intestinal wall. Abdominal X-ray shows little normal bowel gas.
the bowel and prominent engorgement of mesenteric vessels with a palisade pattern (the comb sign). After high dose methylprednisolone (1 g/day intravenously) was administered for 3 days, the patient's symptoms markedly improved. Thereafter, resolution of the mural thickening was observed within 1 week by abdominal ultrasound examinations.

3. Discussion

Gastrointestinal symptoms are common in SLE patients, and are usually caused by reactions to medications, and viral or bacterial infections. Although SLE-related gastrointestinal involvement such as LMV is not a common cause of gastrointestinal manifestation, a delayed diagnosis can be life-threatening. The symptoms of LMV vary from mild, nonspecific abdominal pain, bloating, nausea/vomiting or diarrhea, to necrosis and intestinal perforation which manifests as severe extensive gastrointestinal bleeding or acute surgical abdomen. LMV has a good therapeutic response to corticosteroids and immunosuppressive agents. However, it is difficult to differentiate uncommon and life-threatening LMV from common causes of gastrointestinal manifestation in SLE patients by nonspecific clinical symptoms and laboratory parameters. A pathological diagnosis is the “gold standard”; however, it is invasive and not always available. Therefore, accurate diagnosis of LMV usually depends on an abdominal CT scan. The “target sign” (abnormal bowel wall enhancement) and “comb sign” (stenosis or engorgement of mesenteric vessels) are typical manifestations of LMV on CT scans. Wall edema and thickening of the small intestinal wall observed in abdominal ultrasonography resembled an accordion. Although not specific to LMV, this usually implies serious abdominal problems and an abdominal CT scan is necessary for the differential diagnosis. LMV is not a common cause of gastrointestinal symptoms in SLE patients. Disease activity is an important clue to diagnosis. Careful evaluation of sonographic findings, as quickly as possible, is critical to allow for an early and correct diagnosis in SLE patients with gastrointestinal manifestations, especially those with nonspecific symptoms. However, an abdominal CT scan should be performed prior to final diagnosis in cases where LMV is suspected. Ultrasonography can be used repeatedly without the risks of being exposed to radiation, and is also helpful in patient follow-up after the initiation of treatment.

References