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CASE REPORT

Acute lupus peritonitis associated with massive ascites

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Abstract

Systemic lupus erythematosus (SLE) is an autoimmune disease with multiple organ involvements and abdominal pain as the most common gastrointestinal symptom.
Herein, we describe a case of SLE presented with abdominal pain and massive ascites that had a good response to the high dose of prednisolone.

Keywords: Acute lupus peritonitis; Massive ascites

Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease characterized by involvement of various organs. Although gastrointestinal manifestations are frequently observed in these patients, their incidence varies widely. Among these, abdominal pain is the most common symptom.[1] Ascities in lupus is rarely massive and regardless of the etiology, it has a typical gradual onset and occurs after an established diagnosis.[2] The present report describes a 16 y/o girl with newly diagnosed lupus who presented with severe abdominal pain and massive ascites one month after diagnosis.

Case Report

A sixteen-years-old girl, with malar rash, photosensitivity, fever, arthritis, positive antinuclear antibody (ANA), anti double strand DNA antibody (Anti ds DNA Ab) and low complement levels was diagnosed as a definite case of lupus and treated with prednisolone (5 mg/day) and hydroxychloroquine (200 mg/day). She was well up to one month after starting medications, when developed a severe abdominal pain and protrusion. She was admitted to surgical emergency ward of Nemazee Hospital affiliated to Shiraz University of Medical Sciences in February 2005 with impression of acute abdomen but was found to have ascites. She was discharged when her symptoms improved spontaneously. On the following day, she again developed more severe abdominal pain associated with nausea and vomiting and was admitted to Internal Medicine Ward of the hospital. On physical examination, she was febrile with normal chest and cardiovascular condition. Examination of her abdomen showed hypomotility, guarding and tenderness.

Laboratory investigations showed white blood cell count of 10.2×10^9/L with 75% polymorphs, 15% lymphocytes, 8% monocytes and 2% basophils. Other findings included hemoglobin 10.3 g/dl, platelet 226000/L and sedimentation rate of 35 mm/h. Urine analysis revealed 3^+ albumins and was positive for Staphylococcus aureus. Liver and renal function tests were normal with amylase and lipase being within normal limits.

She had positive serum ANA, Anti-double strands DNA Ab, anti-cardiolipin antibody and low serum level of C_4 complement component. The abdominal paracentesis showed a low serum to ascites albumin gradient (3.5 and 2.9 g/dl albumin in serum and ascitic fluid, respectively) with negative results for routine and tuberculosis cultures, acid-fast bacilli staining and cytology as well as negative PPD skin test.

White blood cell count of ascitic fluid was 3100/mm^3 (98% Segmented, 2% lymphocyte). Ab-

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dominal ultrasonographies were all in favor of large amounts of fluid, normal bowel wall thickness and mild bilateral pleural effusion. Computed tomography of abdomen with oral and intravenous contrasts showed massive fluid, borderline spleen with homogenous pattern and normal bowel loops without any sign of mesenteric vasculitis. Echocardiography revealed minimal mitral regurgitation and mild pericardial effusion. Color Doppler sonography of abdominal vessels was normal. On admission, ceftriaxone with metronidazole and then amikacin were started and because of the persistent fever and severe abdominal pain, vancomycin was added to antibiotics after the urine culture results were obtained without any change in patient's condition. The surgeon's decision for diagnostic laparatomy was postponed when the rheumatologist suggested increasing the prednisolone dose to 1 mg/Kg/day, with a starting dose of 50 mg. The patient's condition improved and the fever subsided after 36 hrs. Some days later, she underwent kidney biopsy due to the presence of 594 mg albumin in the 24 hrs urine. Abdominal ultrasonography after 6 days showed a decrease in the amount of ascitic fluid.

Discussion

Gastrointestinal (GI) manifestations frequently occur in patients with systemic lupus erythematosus with peritonitis being the most common GI disorder.[1] Ascites is rarely massive and is either related to the typical manifestations of active disease or accompanies nephrotic syndrome, protein loosing enteropathy, constrictive pericarditis and conditions unrelated to lupus. Patients with marked ascites mostly attributable to chronic lupus peritonitis have been characterized by insidious onset, less abdominal pain, vomiting, diarrhea, fever, arthritis and central nervous system disorders, without any relation to the active disease.[2,3] These patients had also poor response to glucocorticosteriod therapy, because of persistent inflammation in the peritoneum and the presence of impaired vascular circulation in addition to immunological abnormalities.[4,5]

There are some reports of nonbacterial causes of acute peritonitis, presenting with abdominal pain, guarding rebound tenderness and large amounts of ascites in lupus patients. Abdominal sonography and computed scanning of abdomen showed massive ascites and bowel loop thickening with collection of intraluminal fluid (enteritis). However, most patients responded favorably to glucocorticosteroids.[1,6]

A study of pathologic specimens from two cases with acute abdominal peritonitis and ascitis, showed bowel loop thickening and thrombotic vasculitis in mesenteric artery and arteries of intestinal wall. Ultrasonography was therefore recommended in order to make an early diagnosis of intestinal vasculitis. It can be used safely or repeatedly without radiation exposure compared to other examinations such as computed tomography scans with contrast.[7] Acute peritonitis with massive ascites in the absence of enough evidences to rule out acute surgical conditions like appendicitis, bowel perforation or ischemia due to vasculitis, may lead to unnecessary laparatomies especially in patients whose clinical manifestations may be masked by glucocorticosteroid consumption.[8]

Our patients had no significant abnormalities in several ultrasonographies and computed tomography of abdomen to explain her massive ascites, without any sign of bowel loop thickening or edema (enteritis). Two specimens of peritoneal fluid were negative for routine and tuberculosis cultures. She was treated as a case of acute peritonitis with high doses of oral prednisolone (1 mg/Kg/day). ANA and anti-ds DNA antibodies were not measured in abdominal fluid, but the positive tests in serum, good response to high doses of prednisolone and renal involvement in the course of disease, were all in favor of peritoneal involvement due to underlying lupus disease activity without superimposed secondary causes.

The reports of lupus patients with large amounts of peritoneal fluids as the initial manifestation of the disease,[9,10] indicate the necessity of modifying the American College of Rheumatology (ACR) criteria for diagnosis of SLE and including the abdominal serositis in the current serositis criteria, manifested as either diffuse abdominal pain with rebound or guarding and ascites or bowel edema in the absence of other causes.[11]

In summary, massive peritonitis with abdominal pain and tenderness occurs rarely in patients with active lupus. Although surgical interventions must be considered in these patients, to avoid unnecessary evaluation laparotomy, more conservative managements and the use of such methods as ultrasonography and computed tomography scan may be more advisable to find other non-surgical causes of abdominal pain. Acute peritonitis and enteritis must also be considered in these patients.

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