

Painless massive ascites and hypoalbuminemia as the major manifestations of systemic lupus erythematosus

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Systemic lupus erythematosus (SLE) is frequently associated with ascites, but rarely without proteinuria. We report a 10-year-old girl with distended, non-tender abdomen with shifting dullness and no pitting edema in the lower legs before admission. Facial rash had appeared 1-2 weeks before admission and became more prominent 3 days prior to admission. Hypoalbuminemia with hypertriglycemia (but no proteinuria or diarrhea) was noticed. The antinuclear antibody titer was 1:2560 (speckle type) and the anti-double-stranded DNA was 1:160. Abdominal echo revealed no cirrhosis change or venous obstruction. Chest X-ray and electrocardiogram revealed no cardiomegaly or pericardial effusion. The serum prealbumin was low on admission day 5, but the liver function tests were within normal range. We deduced that the hypoalbuminemia in SLE without nephritis may be secondary to mesenteric vascular leakage. SLE may present with initial manifestation of painless massive ascites. Careful utilization of history taking, chest X-ray, electrocardiogram, cardiac and abdominal echo, urinary analysis and serum prealbumin is helpful in decision-making while assessing such patients.

Key words: Ascites, hypoalbuminemia, mesentery, systemic lupus erythematosus, vasculitis

Introduction

Ascites in systemic lupus erythematosus (SLE) may be a consequence of nephrotic syndrome, protein-losing enteropathy, constrictive pericarditis or Budd Chiari syndrome. We report a case of SLE with initial presentation of massive ascites. There were no heart or liver diseases, no obstruction of the inferior vena cava, nor was proteinuria found in this patient. The ascites responded well to pulse steroid treatment.

Case Report

A 10-year 6-month old girl had suffered from frequent hair loss, painless oral ulcers and poor appetite for 2 months prior to admission. The presence of facial rash over bilateral cheeks was noticed 1-2 weeks prior to admission (Fig. 1). She had edematous eyelids without



Fig. 1. Facial rash over bilateral cheeks noticed 1-2 weeks prior to admission.

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itching sensation, and her appetite grew poorer over the next week. The abdomen was distended with shifting dullness but there was no abdominal pain or diarrhea. The liver was not enlarged. There was no pitting edema in the lower legs, livedo reticularis or petechiae.

The patient was admitted for suspicious SLE, but nephrotic syndrome with hypoalbuminemia was ruled out. On admission, complete blood cell counts revealed a leukocyte level of $4.48 \times 10^3/\text{mm}^3$, hemoglobin 9.2 g/dL, mean corpuscular volume 86.5, hematocrit 28.8% and platelet $170 \times 10^6/\text{mm}^3$. The C-reactive protein was <5 mg/dL (normal range, <5 mg/dL). Serum albumin was 2.5 g/dL (normal range, >3.5 g/dL). The serum triglyceride was high (339 mg/dL; normal range, 35–160 mg/dL) and the cholesterol level was 241 mg/dL (140–200 mg/dL). Routine urine tests revealed no proteinuria, however. The daily protein loss was 37.25 mg/day (<500 mg/day). The corrected clearance of creatinine was normal ($113 \text{ mL}/\text{min}/1.73\text{m}^2$).

The antinuclear antibody titer was 1:2560, speckle type (normal range, $<1:80$), and the anti-double-stranded DNA was 1:160 (normal: negative) and the anti-extractable nuclear antigens, anti-Ro (Sjögren's syndrome A), and antinuclear ribonucleoprotein (nRNP) antibody were positive. The anti-smooth muscle (Sm) antibody was negative. The lupus anticoagulant was $<1:1.2$ (normal range, $<1:1.2$). Complement factors C3 (20.8 mg/dL) and C4 (1.74 mg/dL) and serum hemolytic complement activity (CH50; 5.18 IU) were depressed (normal range, 83–125.5 mg/dL, 17.2–32.8 mg/dL and 27.3–33.7 IU, respectively). The erythrocyte sedimentation rate was 130 mm in the first hour.

The prealbumin level on day 5 of admission was 10.3 mg/dL (normal range, 17–42 mg/dL). The coagulation profile was normal [activated partial thromboplastin time (aPTT): patient/control = 23.2/29.7 sec, prothrombin time (PT) patient/control = 10.2/11.5 sec, international normalized ratio = 0.8]. The serum aspartate transaminase (AST) and alanine transaminase (ALT) was 19 IU/L and 8 IU/L, respectively (normal range, 34 and 33 IU/L, respectively).

The abdominal echo revealed marked ascites in the cul-de-sac (Fig. 2). No liver cirrhosis or obstructions of the inferior vena cava were noticed. The pancreas was normal. The electrocardiogram (ECG) was normal, and the chest X-ray revealed neither pleural effusion nor cardiomegaly.

After admission, albumin (1 g/kg/day) was administered with intravenous furosemide 1 mg/kg body



Fig. 2. Abdominal echo showing marked ascites in the cul-de-sac.

weight/day. In addition, intravenous hydrocortisone 2 mg/kg/dose 6-hourly was given. The abdominal girth reduced dramatically, from 63 cm to 59 cm within 1 day. The albumin level returned to 4 mg/dL on day 3 and then dropped to 3.4 mg/dL on day 6. Body weight decreased on day 3 (28 kg to 26.5 kg) but rebounded on day 6–7 (28 kg) while an attempt was made to shift treatment to oral steroids. The abdominal girth on day 7 increased slightly to 61.5 cm. Pulse methylprednisolone therapy was given (30 mg/kg/day for 3 days), and then shifted to oral prednisolone (2 mg/kg/day). The patient did not regain appetite until day 11 (immediately after 3 days of steroid pulse therapy).

Body weight steadily decreased to 25 kg, and the abdominal girth to 56 cm by day 13. The serum albumin level was 4.02 mg/dL after albumin administration, 3.4 mg/dL on the day of rebound, and returned to 3.7 mg/dL without intravenous albumin on day 13. The urine output was 2 mL/kg/h on day 1–7 and returned to 4 mL/kg/h on day 13. The prealbumin level returned to normal (26 mg/dL) 3 days after pulse therapy.

The patient was discharged on day 15 with body weight of 25 kg and abdominal girth of 55.5 cm. In clinical follow-up, urinalysis revealed proteinuria 3+, with transient mild hematuria (occult blood 2+). After adding cyclophosphamide (1.5 mg/kg/day), the hematuria ceased. The daily protein loss was 554 mg/day, with the serum albumin 3.5 mg/dL. She is now receiving oral prednisolone (1.7 mg/kg/day) and cyclophosphamide (1.5 mg/kg/day).

Discussion

Ascites is usually overlooked in SLE patients unless accompanied by pain. Schousboe et al compared clinical manifestations in acute and chronic lupus peritonitis [1].

In acute lupus peritonitis, ascites develops quickly and is usually accompanied by abdominal pain, which may mimic surgical abdomen. In contrast, chronic lupus peritonitis is more likely to be painless over the course of several months. In SLE patients with painful ascites, redness and edematous intestine wall with plasma cell infiltration were usually found at exploratory laparotomy [2-5].

In contrast, a review by al-Hakeem and McMillen reported that 9 out of 13 patients undergoing laparotomy turned out to have cholecystitis, perforated ulcer, colonic perforation, diverticulitis, and adhesions in the evaluation of abdominal pain in SLE patients [6]. Careful evaluations are necessary in treating SLE patients with acute onset of painful ascites.

SLE with ascites is usually found to be accompanied by nephritis in most patient diagnoses [3,4,7], but our patient had a normal urinalysis and daily urinary protein excretion. Protein-losing enteropathy was excluded because no diarrhea occurred. Liver function tests were normal (normal serum AST, ALT; normal PT and aPTT; and compensated elevation of triglyceride/cholesterol) and the abdomen echo revealed no cirrhosis. No pleural effusion, cardiomegaly or pericardial effusion was found in the chest X-ray and ECG; pericardial effusion itself may be a contributing factor for ascites. The cause of hypoalbuminemia was taken as leakage from mesenteric vessels as a manifestation of vasculitis in SLE, rather than gastrointestinal or renal loss.

The decreased synthesis of prealbumin may be the result of poor appetite for 1 week, since the serum half-life of prealbumin is 2 days. After pulse steroid therapy, appetite was regained within 3 days, and body weight, abdominal girth, and the edematous state of eyelids declined (Fig. 1). The serum level of albumin remained steady at 3.5 mg/dL and then slowly elevated. The prealbumin level returned to normal after pulse therapy. This suggests that pulse steroid therapy may slow the inflammation of mesenteric vessels, decrease vascular leakage, and thus stabilize the serum albumin level, with the synthesis of prealbumin returning to normal when the liver receives adequate amounts of peptides.

This girl satisfied 6 of 11 criteria of SLE on the basis of: 1) malar rash; 2) painless oral ulcers; 3) relative anemia, leucopenia and thrombocytopenia; 4) antinuclear antibodies 1:2560; 5) anti-double-stranded DNA 1:160; and 6) proteinuria when followed up in clinic. However, the ascites may relate to vasculitis as

one of the manifestations of SLE [2,5,7]. Although the anticardiolipin antibody and the lupus anticoagulant were all negative in this patient, there was 1 report of chronic ascites (3 months) in a 72-year-old woman with antiphospholipid antibody and low C3 level [8]. The initial response to steroid therapy was good, but on day 7, she had severe abdominal pain with complications and finally died. The postmortem examination revealed extensive infarction of intestine and thrombotic occlusion of superior mesenteric artery. Therefore, following the antiphospholipid antibody in SLE patients with ascites may be important. Our patient did not have antiphospholipid antibody, and presented as simple non-tender ascites.

In conclusion, we believe that painless ascites due to vascular leakage of mesenteric vessels might have been one of the first manifestations of SLE without nephritis on this subject. History taking and serial examination such as chest X-ray, ECG, cardiac and abdomen echo (for pericardial effusion, liver cirrhosis, and inferior vena cava patency), urinary routine, 24-hour creatinine clearance and protein loss, PT/aPTT test and serum prealbumin are recommended to identify the pathogenesis of ascites.

Thrombotic occlusion of intestines could be detected by tracing antiphospholipid antibody, anticardiolipin antibody, lupus anticoagulant and PT/aPTT test during the treatment course. Paracentesis might be cautiously considered unnecessary if the origin of ascites is auto-immune-mediated and responsive to treatment [9,10]. When painful ascites is the main complaint, possible surgical conditions such as cholecystitis, perforated ulcer, diverticulitis, perforation and adhesions should not be ruled out.

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