



A Case of Systemic Lupus Erythematosus Initially Presented with Acute Acalculous Cholecystitis

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SLE is an autoimmune disease with multiorgan involvement and a wide range of clinical manifestations, and inflammation of gallbladder also can be represented. There were a few cases of acute acalculous cholecystitis (AAC) in previous reports. Most of them tended to already know about underlying SLE when detected AAC at that time. It may be difficult to detect AAC caused by SLE not due to biliary stone if physician is not conscious of undiagnosed lupus. We introduce a 70-year old female pa-

tient, who is diagnosed with AAC. Her symptoms were satisfied the ACR classification criteria for SLE, and was diagnosed with SLE, simultaneously. After a high dose steroid pulse therapy, followed by cyclophosphamide, her symptoms have improved rapidly. In order to better diagnose and treat the disease, we need to be aware of AAC as a potential manifestation of SLE.

Key Words. Lupus, Cholecystitis

Introduction

Acute acalculous cholecystitis (AAC) is clinically identical to acute cholecystitis but is not associated with gallstones, and usually occurs in critically ill patients. It accounts for approximately 10 percent of cases of acute cholecystitis and is associated with high morbidity and mortality (1). There are multiple risk factors for developing AAC. Specific infections, trauma, septic condition, immunosuppression are major responsible for AAC, and a few study revealed that SLE rarely brings out AAC. The most of the cases of AAC developed during the course of SLE after diagnosis of this disease. And most of SLE patients with cholecystitis with or without cholelithiasis underwent surgical treatment. Only some of them were treated with steroid and immunosuppressive agents (2,3).

We described a case of a 70-year-old woman with SLE who initially presented with AAC and was successfully treated with high dose steroid and cyclophosphamide. In addition, we

suggest that AAC should be aware of as a very rare manifestation of SLE to better approach for early, adequate diagnose and treat disease.

Case Report

A 70-year-old woman visited emergency department with abrupt onset of abdominal pain on the right upper quadrant and fever. There was no specific past medical history noted including any immunologic or allergic disorders. On admission, the body temperature of 38.5°C, the pulse rate of 70/min, the respiration rate of 20/min and the blood pressure of 100/60 mm Hg were noted. She looked pale and severely dehydrated. On physical examination, there was tenderness on the right upper quadrant of her abdomen, the positive Murphy's sign. Abdominal CT (Figure 1) demonstrated gallbladder wall thickening with pericholecystic edema without any evidence of stone or biliary sludge.

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Figure 1. The abdominal CT shows gall bladder distension with mild wall thickening without visible stone.

Initial laboratory tests showed ESR 111 mm/hr, hs-CRP 137.8 mg/L, Procalcitonin 0.153 ng/mL, WBC 20,390/mm³, Hb 10.9 g/dL, platelet 76,000/mm³, total protein 5.8 g/dL, albumin 2.5 g/dL, AST 34 IU/L, ALT 39 IU/L, total bilirubin 1.04 mg/dL, direct bilirubin 0.64 mg/dL, Na/K 139/3.1 mEq/L, BUN/Creatinine 25/0.69 mg/dL. Acute cholecystitis was diagnosed and antibiotics were administered with percutaneous transhepatic gallbladder drainage (PTGBD). Unfortunately, her symptoms and laboratory tests aggravated with conventional treatment for acute cholecystitis. Follow up laboratory tests showed WBC 29,350/mm³, Hb 6.6 g/dL (corrected reticulocyte count 0.43%), platelet 3,000/mm³ and schistocytes were found on peripheral blood smear. Urinalysis showed proteinuria (2,344 mg in 24 hours) and pathologic casts. But, Procalcitonin was still the normal range. At the time, she revealed malar rash on the face (Figure 2), thus immunologic tests were done.

Immunologic tests showed positive FANA (cytoplasmic pattern 1 : 80), C3 25.9 mg/dL, C4 3.3 mg/dL but negative anti-dsDNA antibody (even on repeated tests). Direct/indirect Coombs' tests were positive. Anti-nucleosome and anti- β 2 glycoprotein antibodies were positive but lupus anticoagulant and anti-cardiolipin antibody were negative. Anti-Ro (SSA), anti-La (SSB), anti-RNP, anti-Sm antibodies, p-ANCA, and c-ANCA were all negative. Chest CT showed pleural thickening with some pleural effusion. A diagnosis of SLE was made by positive ANA, proteinuria, hemolytic anemia, malar rash and pleuritis, fulfilling 5 of ACR classification criteria for SLE. About proteinuria, we didn't perform renal biopsy. Intravenous steroid pulse therapy (1 g/day for 3 days) was done followed



Figure 2. The patient has revealed malar rash on her cheeks on the course of the disease.

by intravenous cyclophosphamide (400 mg/day). The symptoms markedly improved several days after the treatment. Follow-up laboratory results after 12 days revealed that elevated complement levels (C3 93.6 mg/dL, C4 17.4 mg/dL). Follow-up CT scan showed near complete recovery and PTGBD catheter was removed. Steroid was tapered by 5 mg per day and the patient was discharged for outpatient department.

She has been administered IV cyclophosphamide for four times, Follow up laboratory tests showed improved proteinuria (170 mg in 24 hours) and no pathologic casts after 4 month.

Discussion

Systemic lupus erythematosus (SLE) is a chronic inflammatory autoimmune disease of unknown etiology that can affect virtually every organ (4). Several gastrointestinal manifestations, including mesenteric arteritis, bowel perforation, gastric or duodenal ulcer, lupus enterocolitis, spontaneous peritonitis and pancreatitis have been reported in association with SLE (5-7). However, there were only a few reports of case with in patients with SLE and most of the patients had already been diagnosed with SLE before the occurrence of AAC (2-4, 8,9). Initial presentation with AAC in SLE is very unusual and it has difficulties in diagnosing. And most of previous reports showed that cholecystitis was developed during active stage of disease. In our case, the patient had no medical history and showed negative anti-dsDNA. Although ANA was positive, it showed very low titer (1 : 20 at the first time) to judge from her age. It made difficult to make appropriated diagnosis of SLE at the first time of disease presentation. If it was not for malar rash and procalcitonin in the normal range, it would be very difficult to make a decision. Although she needed a surgi-

cal treatment, surgery was not feasible since her laboratory test showed severe thrombocytopenia and hemolysis. Therefore, PTGBD was done first and broad spectrum antibiotics were administered as a conservative treatment. Of course, what is the leading factor in this case was difficult to distinguish that SLE flare first break, or otherwise cholecystitis would have occurred before. However, her condition was going worse even after the antibiotic treatment. When abdominal pain and other symptoms getting worse, procainin was within normal range. So, we were diagnosed with SLE flare up, steroid therapy was started.

Vasculitis and thrombosis are considered as two main causes of cholecystitis in SLE. Vasculitis of the gallbladder is characterized by the presence of acute arteritis with periarterial fibrosis (5-7). Mesenteric inflammatory veno-occlusive disease (MIVOD) is another rare cause of acalculous cholecystitis in SLE with unknown etiology (10). This type of vasculitis exclusively involves mesenteric vein and/or their branches sparing the arterial vasculature. Thrombotic cause is most frequently observed in SLE patients with antiphospholipid antibodies and characterized by thrombi in the gallbladder veins without evidence of vasculitis. Although our patient had positive anti- $\beta 2$ glycoprotein antibodies, there were no evidences of thrombosis in clinical and radiologic evaluations. So, she was treated with high dose steroid pulse therapy followed by cyclophosphamide with dramatic improvement of her condition. This findings favours an inflammatory etiology and not thrombotic, but MIVOD could not be ruled out in this case.

The adequate management of AAC in patients with SLE has been controversial. In general, the prognosis of AAC is worse than that of acute calculous cholecystitis and, therefore, the treatment of choice is surgical removal of the gallbladder. Operative treatment is considered the main treatment by many authors due to high risk of morbidity. Many previous cases were treated surgically by cholecystectomy (2). However, the present case suggests that, as far as SLE is in high activity, acalculous cholecystitis can be treated successfully by corticosteroid without surgical intervention, unless the risk of rupture is impending. Nonoperative management of AAC with immunosuppressive agents has been two previous case reports in the literature addressing successful treatment of SLE-induced AAC with high doses of corticosteroid (5,6). The decision on medical or surgical treatment should be based on the patient's general condition and risk factors. It has been sug-

gested that if the patient's general condition is good and there is no other risk factor for AAC or its serious complications, high-dose steroid therapy may be considered as the first line of treatment.

Summary

We herein reported a rare case of SLE patient with AAC as an initial presentation who was treated successfully with high dose steroid pulse therapy and immunosuppressive agent. Awareness of this finding would be valuable in the early diagnosis and adequate management of AAC in patients with SLE.

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