A RARE INITIAL MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS - ACUTE PANCREATITIS WITH MULTIPLE COMPLICATIONS

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Abstract

Acute pancreatitis as an initial presentation of systemic lupus erythematosus is rare, but its outcome is often fatal. We report a 23-year-old female patient presenting with abdominal pain and ascites followed by acute lung injury. Increased serum amylase (717 IU/l) and lipase (5579 IU/l) levels suggested acute pancreatitis with complications, which was confirmed by computerized tomography. Multisystemic involvement including seizure, pancytopenia and serositis suggested SLE, which was confirmed later by increased ANA (1:160) and anti-dsDNA (1:1280) levels. Pulse steroids (1 g of IV methylprednisolone per day for three doses) were administered promptly, followed by a 2-month treatment with oral steroids and azathioprine, and the patient recovered. We should keep a high index of suspicion of the diagnosis, when we meet patient with acute idiopathic pancreatitis. Aggressive treatment with immunosuppressant may save the patient’s life.

Key words: Acute pancreatitis, Systemic lupus erythematosus, Abdominal pain, Complications

INTRODUCTION

Systemic lupus erythematous (SLE) is a multisystemic autoimmune disorder that mostly affects young women, especially during their childbearing years. The most common clinical symptoms are polyarthritis and dermatitis, however any symptom or sign of the disease may be its initial manifestation which frequently makes diagnosis difficult.¹ A number of lupus patients develop episode of acute pancreatitis (AP), but diagnosis of acute pancreatitis as the initial manifestation of SLE is rare.²

We report a young female with abdominal pain, elevated levels of pancreatic enzyme, hypocalcemia, hypoxemia, and other laboratory abnormalities. She was first diagnosed with acute severe pancreatitis with complications and then with SLE after further investigations.

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CASE PRESENTATION

A 23-year-old woman arrived at the emergency department with acute ill-looking appearance, complaining of abdominal pain, poor intake, and nausea for 3 days. She was afebrile, pulse rate 93 beats per minute, blood pressure of 99/69 mmHg. Physical examination revealed clear breath sounds with no cardiac murmurs, and the abdomen was diffusely tender without signs of peritoneal irritation. No pitting edema was noted in the lower extremities. Neither joint swelling nor skin rash was found. According to self-report, the patient denied having any history of abdominal disease, alcohol abuse, or symptom of arthritis, oral ulcer or dermatitis.

A complete blood count showed anemia of 9.1 g/dL hemoglobin, leukopenia of 3690/mm^3 (normal 5000 to 10,000), and thrombocytopenia of 116,000/mm^3 (normal 150,000 to 400,000). Serum amylase level was elevated at 717 IU/L (normal < 110) as was the patient’s lipase level, 5579 IU/L (normal < 60). Liver enzymes were elevated (GOT/GPT: 750/117, normal < 35), albumin was 1.5 g/dL (normal 3.5 to 5), calcium was 7.0 mg/dL, triglyceride level was 446 mg/dL (normal < 150), and cholesterol level was normal. Creatinine was 1.9 mg/dL (normal 0.3 to 1.4), and urinalysis showed proteinuria, hematuria and hyaline, and granular casts. Abdominal computerized tomography (CT) (Fig. 1) revealed pancreatic swelling without pancreatic or biliary obstruction, and intraperitoneal fluid accumulation within the abdomen and pelvis.

AP was diagnosed and treated with hydration and intravenous Gabexate mesilate (300 mg/day) infusion. Laparotomy performed after the patient developed acute abdomen with peritonitis on the second day of hospitalization and showed pancreatitis with free ascites, diffuse fat necrosis and calcium-soaponification plaques. Unfortunately, acute respiratory failure occurred, with low PO2 and PCO2. Chest X-ray (Fig. 2) showed diffused mottling in the lung fields consistent with acute lung injury (PaO2/FiO2= 287) caused by AP.

The patient’s condition continued to deteriorate, with development of generalized seizure, loss of consciousness and hemodynamic instability. Echocardiography showed normal left ventricular functions with small amounts of pericardial and pleural effusion. Multiorgan involvement in this young female and progressing idiopathic AP led to suspicion of autoimmune disease-associated
pancreatitis. Further laboratory studies disclosed the following values: positive speckled antinuclear antibodies (ANA) at 1:160, positive anti-dsDNA at 1:1280, positive anti-cardiolipin IgG antibody at 40 (normal < 15), positive anti-platelet antibody screening, C3 at 22.9 mg/dL (normal > 86), and C4 at 1.72 mg/dL (normal > 16). RF, anti Ro Ab, anti La Ab, anti RNP Ab, and anti Sm Ab were all negative.

It was assumed that pancreatitis was the initial manifestation of SLE exacerbation, together with serositis, renal disorder, neurological disorder, hematological disorder, immunological disorder on serological testing, and antinuclear antibodies, which fit 6 of the 11 revised criteria of the American Rheumatism Association for diagnosis of SLE.3

The patient was treated with pulse steroid therapy (1 g of IV methylprednisolone per day for three doses), followed by prednisolone 50 mg/day, and azathioprine at 50 mg/day. Anticoagulation with aspirin was also given. After 7 days of immunosuppressant therapy, there was gradual improvement, with normocalcemia achieved. Liver function test and amylase level normalized after 10 days. Assisted ventilation was required for 15 days. The patient was finally discharged 48 days after admission. She received surgical debridement for a reason secondary to necrosis with peritonitis, although infection was not proven. Her Ranson score at 48 hours after admission was 7, meaning more than 50% mortality and associated with more systemic complications, including acute respiratory failure, ascites and pleural effusion.

Pascual-Ramos et al.8 state that the SLE disease activity index is significantly increased in those patients with idiopathic pancreatitis. Laboratory findings and clinical symptoms of our patient were compatible with active SLE, including pancytopenia, proteinuria, hypocomplementemia, serositis and elevated titer of autoantibodies. This patient had multiorgan involvement of SLE, namely the circulatory, renal and immune systems, as well as neurological involvement. However, the most striking feature of our patient was the idiopathic necrotic AP. Widespread SLE including active AP was diagnosed.

Lupus activity is significantly associated with increased mortality of AP patients (40%
among those with SLE manifestations). There is 100% mortality rate for those who develop circulatory shock or acute renal failure, with an 87% rate in patients with respiratory insufficiency. In other cases, the severity and complications of some episodes of AP in SLE patients contribute to the final development of chronic pancreatitis. Tsianos et al. reported that about 30.5% of asymptomatic SLE patients have hyperamylasemia; therefore subclinical pancreatic damage might occur very frequently in SLE.

The diagnosis of lupus-associated pancreatitis is based on clinical findings such as abdominal pain and peritonitis as well as laboratory findings like pancreatic enzymes, immunological data and suggestive image findings. Etiology diagnosis is fairly important for treatment decision. Corticosteroids and azathioprine were prescribed for our patient after SLE diagnosis, with her condition gradually stabilizing. Even though steroids themselves may cause pancreatitis, there is a significantly decreased mortality rate and improved prognosis among patients who are treated with steroids or azathioprine during pancreatitis.

**SUMMARY**

Although lupus-associated pancreatitis is uncommon, due to its potentially fatal outcome, it should be suspected in acute idiopathic pancreatitis and any SLE patient with abdominal pain. The mortality of patients with lupus-associated pancreatitis can be reduced with earlier diagnosis and earlier steroid therapy, in addition to other supportive management.

**REFERENCES**

紅斑性狼瘡少見的初期表現—
急性胰臟炎合併多種併發症

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摘要

以急性胰臟炎作爲紅斑性狼瘡的初期表現是非常少見的，卻常常會致命。我們報告一位23歲女性病患因腹痛和腹水隨後發生急性肺損傷，同時發現胰臟酵素值上升，在經由電顱斷層確診後，初期診斷爲急性胰臟炎。然而接著同時發生了多種系統性疾病，包括抽搐、全血球缺乏、纖維炎。經由更進一步的檢查如 ANA（1:160）和 anti-dsDNA（1:1280）指數上升，才確認是紅斑性狼瘡。於是馬上使用高劑量類固醇治療，接著使用口服類固醇和 azathio-prine治療2個月之後，病患逐漸恢復正常。當我們遇到不明原因的急性胰臟炎時就必須要把紅斑性狼瘡納入鑑別診斷，因爲積極的以免疫抑制劑治療可以拯救病患的生命。

關鍵詞：急性胰臟炎，系統性紅斑性狼瘡，腹痛，併發症

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