CASE REPORTS

A report of systemic lupus erythematosus accompanying acute pancreatitis

Hui Shi[1], Riga Wu[2], Dantong Wang*[1]

[1]*Department of Rheumatology and Immunology, Baogang Hospital, Baotou, Inner Mongolia, China*
[2]*Imaging Center, Baogang Hospital, Baotou, Inner Mongolia, China*

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Abstract

Objective: Systemic lupus erythematosus (SLE) is a multi-system autoimmune disease, in which multiple organs are involved. Lupus with pancreatitis (LP) is a rare disease. It is an effective strategy for clinical work to comprehensively study the pathogenesis, diagnosis and treatment of LP.
Methods: A report of 42-year-old female patient with intermittent arthralgia more than 10 years, rash, edema on double lower limbs for 7 years. The patient's condition was worsened for 10 days, given a diagnosis of SLE nausea, vomiting and abdominal pain after admission. Examinations: blood amylase 612 U/L, urine amylase 3,345 U/L. Ultrasound: pancreas swelled with a change in texture, with ascites (medium). Considered as severe acute pancreatitis, the patient was given treatment with 80 mg methylprednisolone qd, 0.2 g cyclophosphamide ivgtt qod, fasting. It was required of gastrointestinal decompression, acid suppression, somatostatin inhibiting pancreatic secretion, anti-inflammatory treatment, intraperitoneal perfusion of antibiotics and other symptomatic treatments. The symptoms were relieved after 18 days.
Results: Hormones were selected and combined with immunosuppressants to relieve symptoms with the aid of gastrointestinal decompression, acid suppression, somatostatin and anti-inflammatory therapy.
Conclusions: It is critical to summarize the characteristics of LP, diagnose early and give individualized treatment in order to relieve LP.

Key Words: Systemic lupus erythematosus, Acute pancreatitis

1 Medical record

1.1 General information

A female patient, 42 years old, was admitted to hospital on November 14 of 2013, mainly due to "intermittent arthralgia more than 10 years, rash, edema on double lower limbs for 7 years, worsened for 10 days". 10 years ago, the patient unprovokedly developed swelling and pain in double hands and fingers, double wrists, shoulders and knee joints, with physical limitation and morning stiffness. She was given an early treatment in the local hospital and diagnosed as "rheumatoid arthritis" with symptoms relieved after symptomatic treatment. Later, the patient took some drugs from private dispensaries intermittently with poor effects. 7 years ago, the patient developed rashes and photosensitivity on the face and lower limbs, even worse, skin ulceration on the lower limbs, occurring with hair loss, sjogren's syndrome and exfoliated deciduous teeth. 2 years ago, she suffered from chest tightness and shortness of breath after activity. 10 days before admission, the patient began to show symptoms of cough and difficult expectoration with occasional abdominal pain, nausea, no vomiting. Daily food intake was 100-150 g. The frequency and the condition of defecation and urination were normal.

*Correspondence: Dantong Wang; E-mail: doctorwang01@163.com; Address: Department of Rheumatology and Immunology, Baogang Hospital, Baotou, Inner Mongolia, China.

1.2 Physical examination

T 36.5, P 80 beats per min, R 18 breaths per min, BP 90/60 mmHg, clear state of mind, fluent language. Old rashes about the size of a grain of rice were seen to be scattered on the lower limbs and old cicatrices were scattered on both crura. The patient showed symptoms of cyanosis on oral lips, swelled upper lip, dry mouth and a full denture. Bilateral lung breath sounds rough with no abnormality in heart rate and abdomen. Mild tenderness appeared when touching proximal interphalangeal and metacarpophalangeal joints of two hands, as well as knee joints. It was showed that myatrophy was present on both crura with tenderness (+), severe edema on both insteps and lower limbs. Auxiliary examination: Routine blood test showed WBC 4.68×10^9, Bpc 116×10^9. Routine urine test showed PRO (+) and BLD (-). 24-hours proteinuria showed 1.2 g per 24 hours. Blood gas analysis showed PH 7.387, PCO_2 36.2 mmHg, PO_2 66.6 mmHg and BE -3.1 mmol/L. Blood biochemical items showed ALT 77 U/L, AST 349 U/L, ALB 27 g/L, C3 22.4 mg/dl, C4 5.52 mg/dl. Immune indexes were as follows: ANA: 1: 640 homogeneous pattern, cytoplasmic pattern; ds-DNA: 178.7 IU/m; ENA: anti-nRNP (+), anti-Ro52: weak (+), anti-SSA (+), anti-SSB (+), ANUA: weak (+), AHA: weak (+), anti-rRNP: weak (+), ACL (-), ANCA (-). ESR showed 48 mm/h. Pulmonary function indicated the presence of small airway dysfunction and restrictive ventilatory disorder. Heart color Doppler ultrasound indicated that pulmonary artery and it branches were widened with no pulmonary and tricuspid regurgitation. Systolic pressure of pulmonary artery was 23 mmHg.

1.3 Primary diagnosis

Systemic lupus erythematosus (SLE), lupus with kidney injury, vasculitis and interstitial lung disease.

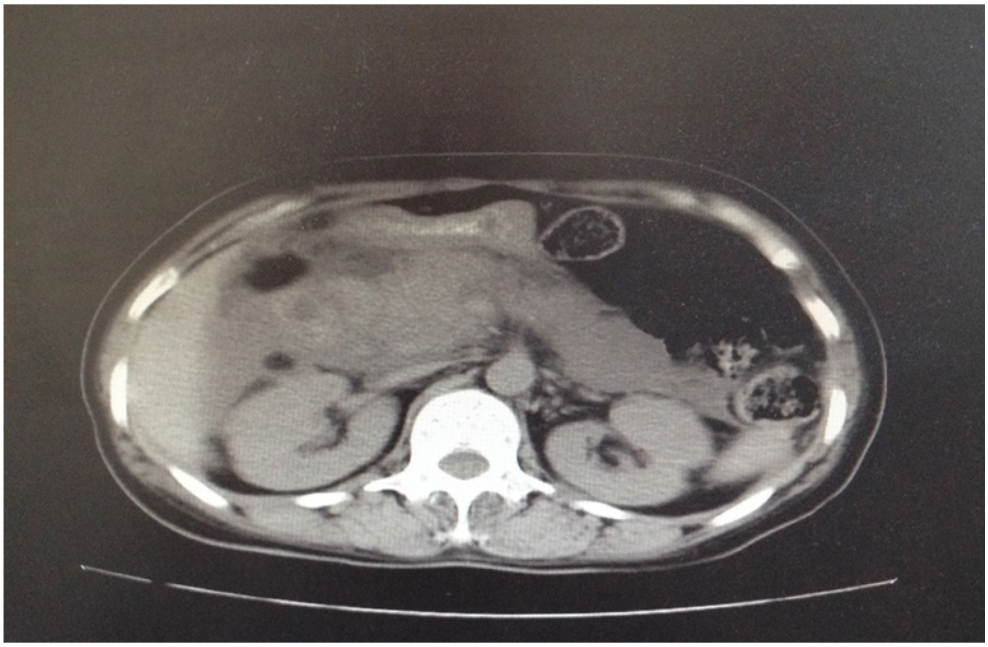

Figure 1: CT manifestation of pancreatitis

1.4 Treatment

After admission, the patient was given treatment with 50 mg prednisone acetate qd po, 0.2 g hydroxychloroquine bid po and 0.2 g cyclophosphamide ivgtt qod. The patient was obviously relieved of chest tightness, shortness of breath and edema on double lower limbs. 5 days after admission, the patient suddenly developed nausea and vomited for several times, and the vomitus was gastric contents. Emergent examination showed blood amylase 612 U/L and urine amylase 3,345 U/L. It was considered as acute pancreatitis. The patient was given treatment with 80 mg methylprednisolone qd, 0.2 g cyclophosphamide ivgtt qod, fasting. It was required of gastrointestinal decompression, acid suppression, somatostatin inhibiting pancreatic secretion, anti-inflammatory treatment and other symptomatic treatments. 3 days later, the abdominal pain was relieved with abdominal distension aggravated. Re-examination showed blood amylase 559 U/L, pancrelipase 639 U/L and urine amylase 3,582 U/L. B-scan ultrasonographic findings showed that pancreas swelled with a change in texture, with ascites (medium); abdominal CT scan indicated pancreas swelled obviously with reduced density, blurred edge, and effusion in minimum amount around pancreas (see Figure 1). Abdominocentesis was conducted to determine the level of amylases (1,659 U/L). Considered as severe acute pancreati-

tis, the patient was given treatment with peritoneal catheter drainage, intraperitoneal perfusion of antibiotics and other symptomatic treatments. The symptoms of digestive tract disappeared completely after 18-day treatment, and the re-examination showed blood amylase 94 U/L and pancrelipase 691 U/L, LFT and the level of urine amylase recovered to normal. Re-examination of color Doppler ultrasound showed ascites were completely absorbed. The patient was discharged from hospital with symptoms relieved.

2 Discussions

SLE is a multi-system autoimmune disease, in which multiple organs are involved. Acute pancreatitis (AP) can be induced by many precipitating factors, 80% of which are associated with biliary tract disease and overeating. Hyperlipidemia, hypercalcemia, infections and drugs can also be included in predisposing factors. The predisposing factor about the patient in this case can be excluded from above factors.

Lupus with pancreatitis (LP) is a rare disease. After analyzing clinical characteristics in 28 cases of patients with pancreatitis related to autoimmune diseases, Wang Q et al.[1] conclude that: pancreatitis is very common in women; can reflect the activity of diseases; is of high-morality; can be controlled by hormones + immunosuppressants. Chronic pancreatitis is less common than AP.[2]

Bai et al.[3] retrieved and analyzed 56 cases of LP patients reported domestically during 1983-2009, and the research results showed that SLE with the first clinical manifestation of AP accounted for 32.7% with an overall case fatality rate of 29.3%. The case fatality rate of severe AP can reach up to 68.4%, and its clinical manifestations contained abdominal pain, nausea, vomiting, fever, the increase of blood amylase with increased urine amylase and lipase. Positive rates of swollen pancreas detected by abdominal B-scan ultrasonographic and CT scan were 86.0% and 97.1% respectively. Glucocorticoids are more effective when combined with immunosuppressants in the treatment of LP. Derk CT et al.[4] studied cases of LP patients reported by Thomas Jefferson University during 1982-2002, and concluded that LP accounted for 0.85%, 76% of which were in the active stage of SLE; The number of organs involved was at an average of 4.4 (renal diseases accounted for 56%, hydrothorax 48%, arthritis 44%); 82% of patients were relieved of symptoms clinically and laboratorially after using a large dose of hormones, which cannot aggravate AP. Breuer GS et al.[5] reported that the annual incidence of LP was 0.4‰-1.1‰, 80% of which were women; 97% of diagnoses depended on the measurement of serum amylase (blood amylase) and lipase. This disease had no correlation with hormones + azathioprine. The morality was 27%, which could be reduced by 67% by use of hormones + azathioprine.

2.1 The pathogenesis of LP

(1) Vasculitis: caused by circulating immunocomplex deposited on the vascular walls in pancreas; (2) Diseases of blood vessel caused by microthrombus and antiphospholipid syndrome; (3) Autoantibodies in pancreatic tissues; (4) Medication-induced;[6] (5) Virus infections etc.; (6) Macrophage activation syndrome.[7] Among 11 cases of LP children patients, there were 10 cases accompanied with macrophage activation syndrome. The mutual characteristics are that: over-activated and over-proliferated T-lymphocytes and macrophages secrete a large amount of proinflammatory factors, which aggravate pancreas injury with clinical manifestations such as fever, hypocytosis and ferritin septicemia. Goel R et al.[8] reported that the incidence of LP was associated with epileptic seizure, non-use of hormones and arthritis.

2.2 Clinical manifestations of LP

Pancreatitis can not only occur in the active stage[9] but also the static stage of lupus. Clinically, it is characterized by abdominal pain, nausea, vomiting and abdominal distension, fever and so on. Part of patients easily fall into multiple organ failure with an aggressive clinical course, and the morality is high. The diagnosis is based on laboratory examinations (such as increased levels of blood amylase and urine amylase) and imageological examinations (e.g. swollen pancreas, abscess and pancreatic pseudocyst can be detected by ultrasonography and CT).[10] CT is helpful in the process of determining causes that induce abdominal pain, guiding the treatment and monitoring complications.[11]

2.3 The diagnosis and the differential diagnosis of LP

The diagnosis of LP should be excluded from some causes such as calculus of extrahepatic duct, alcoholic factor, drug-induced factor, hypercalcemia and virus infections etc. A series of tests indicate the increase of blood amylase and lipase, swollen pancreas and inflammatory exudation confirmed by B-scan ultrasonographic and CT scan. Consequently, LP can be diagnosed when drug-induced pancreatitis and IgG4-related autoimmune pancreatitis are excluded.

2.4 The treatment of LP

Glucocorticoid is a controversial focus,[12,13] which most researches have shown that hormones are effective in the treatment of LP.[14–16] However, it was also reported that hormones could induce and worsen AP.[6] The AP mentioned in this case was considered to be induced by protopathy, and 80 mg methylprednisolone was applied in the treat-

ment. The results showed that the application of hormones with an appropriate dose can relieve the symptoms and prevent from the risks that causing drug-induced pancreatitis. A large amount of researches have shown that hormones are more effective when combined with immunosuppressants (cyclophosphamide, mycophenolate mofetil and azathioprine) in the treatment and prognosis of LP.[17–22] The patient in this case was given hormones with combination of cyclophosphamide and hydroxychloroquine, and then relieved of symptoms. A case report abroad showed that single use of CD20 (mAb)[23] or combined use of CD20 and CTX[24] can relieve the symptoms in a short time and prevent from side effects (e.g. infections) caused by a large dose of hormones. Hence, CD20 can be applied to patients who fail in hormone therapy or suffer from severe pancreatitis in the second-line setting. Add-on treatments such as fasting, gastrointestinal decompression, acid suppression, somatostatin inhibiting pancreatic secretion, anti-inflammatory treatment play an important role in the treatment of AP; as for AP patients with digestive tract perforation and hemorrhage, it is recommended to undergo surgical intervention as required,[25,26] and it is critical to the outcome. It was also reported that the application of hormones with combination of plasma exchange therapy succeeded in the treatment.[27] Except for the application of hormones and immunosuppressants, add-on treatments (such as gastrointestinal decompression, acid suppression, somatostatin, anti-inflammatory treatment, intraperitoneal perfusion of antibiotics) are key to fast relief of symptoms.

LP is of high morality, which is associated with AP severity, SLEDAI scoring, multiple organ injury,[28] onset age (children > adults);[29] the elevated level of liver enzymes is another risk factor that induces LP.[30]

Conflicts of Interest Disclosure

The authors have no conflicts of interest related to this article.

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