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Acute pancreatitis as an initial presentation of systemic lupus erythematosus: a case report

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Abstract

Background Systemic lupus erythematosus is a systemic autoimmune disease affecting different organ systems. Gastrointestinal symptoms in patients with systemic lupus erythematosus are common. But systemic lupus erythematosus-related acute pancreatitis is a rare presentation. Particularly, it is extremely rare to observe acute pancreatitis as the initial presentation of systemic lupus erythematosus combined with antiphospholipid syndrome.

Case presentation Here, we report a case of abdominal pain as the initial symptom of systemic lupus erythematosus in a patient who was finally diagnosed with systemic lupus erythematosus-related acute pancreatitis. Our patient was a 47-year-old Han female with epigastric pain, nausea, vomiting gastric contents, and loss of appetite. She did not mention any relevant medical history and did not consume alcohol nor greasy food. She was successively diagnosed with acute cholecystitis, acute pancreatitis, and acute appendicitis, but relevant therapeutic interventions proved to be ineffective in improving gastrointestinal symptoms. Renal pathology, along with positive antinuclear antibody and anti-double stranded DNA tests, supported the diagnosis of systemic lupus erythematosus. In addition, the presence of positive anti-cardiolipin antibodies and lupus anti-coagulant, along with thrombosis in vein and internal carotid artery occlusion, supported the diagnosis of antiphospholipid syndrome. Corticosteroid and cyclophosphamide therapy led to resolution of abdominal manifestations, and the patient was discharged with methylprednisolone and hydroxychloroquine. Aspirin was used to treat antiphospholipid syndrome.

Conclusion Systemic lupus erythematosus-related acute pancreatitis should be considered in the differential diagnosis of patients with acute pancreatitis after exclusion of other causes. The patient was given treatment as soon as possible. Corticosteroids combined with cyclophosphamide are an effective treatment.

Keywords Systemic lupus erythematous, Acute pancreatitis, Case report

Introduction

Systemic lupus erythematosus (SLE) is a systemic autoimmune disease affecting different organ systems, and it classically occurs in young to middle-aged women [1, 2]. On the basis of the 2019 European League Against Rheumatism/American College of Rheumatology classification criteria for SLE, SLE is defined as: at least one positive antinuclear antibody (ANA) test as a compulsory entry criterion, followed by additive weighted standards grouped in seven clinical (renal, constitutional, neuropsychiatric, hematological, musculoskeletal, serosal, and mucocutaneous) and three immunological (antiphospholipid antibodies, complement proteins, and SLE-specific antibodies) domains weighted from 2 to 10, with patients accumulating ≥ 10 points [3]. Frequent clinical manifestations of SLE include symptoms of hematological, skin, respiratory, cardiac, and renal involvements [2]. In addition, gastrointestinal symptoms in patients with SLE are common, specifically, abdominal pain [4]. However, SLE-related acute pancreatitis is rare, and the estimated rate of SLE-related pancreatitis is 0.7–4% [5, 6]. Patients

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with SLE-related pancreatitis initially experience a special abdominal symptom that has been described in case reports and small series [7–10]. In addition, antiphospholipid syndrome (APS) is often secondary to SLE [11]. However, studies about acute pancreatitis as the initial presentation of SLE combined with APS are lacking.

Herein, we report a case of a middle-aged woman diagnosed with SLE with acute pancreatitis as an initial presentation in the presence of concurrent APS.

Case presentation

A 47-year-old previously healthy Han woman presented to the department of nephrology with a 1-month history of epigastric pain, nausea, and vomiting. She reported that before the onset of the epigastric pain, she had experienced no fever or diarrhea but reported severe nausea and vomiting gastric contents. Hence, she sought local medical attention to help. Her pancreatic enzyme determinations revealed a blood lipase level of 216.2 U/L (above a normal level), with normal blood and urine amylase levels. Computed tomography (CT) and ultrasonic scans of the abdomen revealed inflammation and fluid accumulation of pancreatitis, as well as effusion in the peritoneal cavity. She was diagnosed with cholecystitis and pancreatitis. Treatment with intravenous fluids, inhibiting pancreas secretion, and instructions to not eat or drink were given; her nausea and vomiting were relieved, but she still experienced epigastric pain and loss of appetite.

Next, she was admitted to the tertiary hospital; repeated pancreatic enzyme determinations revealed increased pancreatic enzyme levels, including: 1046 U/L urine amylase (reference range, < 600 U/L); 161 U/L blood amylase (reference range, 35–135 U/L); and 81 U/L blood lipase (reference range, < 60 U/L). After treatment with somatostatin and esomeprazole, the pancreatic enzyme levels were normal. A repeated CT scan of abdomen revealed a mildly swollen pancreas. She continued to report epigastric pain and loss of appetite. Meanwhile, she gradually began to experience body swelling and coughing, and blood clots formed in the upper right limb blood vessels. However, there were no rashes or joint swelling, fever or photosensitivity, headaches, seizures, or changes in personality. Another CT scan revealed localized shadows obscuring areas of the lung and effusion in left and in right pleura. A complete blood count showed a white blood cell count of 2.18×10^9/L (reference range, 3–9.5×10^9/L) with 65% neutrophils (reference range, 40–75%), 21% lymphocytes (reference range, 20–50%), and 12.8% monocytes (reference range, 3–10%). Hemoglobin was 91 g/L (reference range, 115–150 g/L); hematocrit 25.4% (reference range, 35–45%); and platelets 33×10^9/L (reference range, 125–350×10^9/L). Urinalysis showed urine with a protein level of 1+ and blood in urine (BLD) 1+, but no white cells. Proteinuria was 7.9 g/24 h (reference range, < 150 mg/24 h). She accepted undergoing bone marrow biopsy and aspiration because of pancytopenia, which revealed that: bone marrow hyperplasia was active, the number of megakaryocytes was normal, platelets were rare, granulocytes were megaloblastic, and peripheral leukopenia was observed. She was suspected of having kidney and autoimmune disease. Therefore, she went to the department of nephrology to further examination. The patient had no relevant medical history. She neither smoked nor consumed alcohol. She also was not allergic to food nor any drugs. The patient's family history of autoimmune diseases and pancreatitis was unremarkable.

The patient had the following vital signs and measurements: temperature: 36.2 °C; blood pressure: 119/71 mmHg; respiratory rate: 18 per minute; pulse rate: 86 beats per minute; and weight: 102 kg. Her abdomen was soft with no rebound pain, but positive for tenderness in epigastrium. Furthermore, the neurological examination, musculoskeletal examination, and skin examination results were normal. Examination of other systems revealed no other abnormalities.

Her laboratory examinations were as follows: connective tissue showed an antinuclear antibody (ANA) titer of 1:1280, positive anti-double-stranded DNA (dsDNA) antibody and anti-SSA, negative anti-Smith antibody, anti-ribonucleoprotein, and anti-SSB. Immunoglobulin (Ig)G level was 24.30 g/L (reference range, 7–16 g/L); immunoglobulin G4 was 0.409 g/L (reference range, 0.03–2.01 g/L); complement 3 (C3) was 0.56 g/L (reference range, 0.9–1.8 g/L); complement 4 (C4) was 0.06 g/L (reference range, 0.1–0.4 g/L); erythrocyte sedimentation rate was 82 mm/h (reference range, 0–20 mm/h); and anti-cardiolipin antibodies and lupus anti-coagulant were positive. The blood creatinine was 38.9 μmol/L (reference range, 41–81 μmol/L); protein was 61.4 g/L (reference range, 65–85 g/L); and albumin was 30.1 g/L (reference range, 40–55 g/L). The C-reactive protein was < 10 mg/L (reference range, 0–10 mg/L); procalcitonin was 0.066 mg/L (reference range, < 0.064 mg/L). Ultrasound of both upper extremity veins showed thrombosis in the right axillary vein segment and the subclavian vein. Because she showed nephrotic range proteinuria. After exclusion of contraindications such as a high risk of bleeding, she was given a renal biopsy, which revealed: IgG^+, IgM^{++}, IgA^{++}, $C3^-$, $C1q^+$, FRA^-, κ^{++}, and λ^{+++} cluster was deposited in mesangial region. Ischemic shrinkage of glomerular basement membrane was observed in 12% glomeruli; mild hyperplasia of glomerular mesangial cells and stroma was observed in 84% glomeruli; and fuglobin deposition was observed

in the mesangial area (Fig. 1). The diagnosis was mesangial lupus nephritis (WHO Class II). Electron microscope results of renal biopsy also are consistent with lupus nephritis (Fig. 2).

Finally, on the basis of clinical symptoms, laboratory and imaging examinations, and renal pathology, the patient received the diagnosis of SLE with acute pancreatitis as an initial presentation in the presence of concurrent APS.

Before renal biopsy, she received 20 g per day of human immunoglobulin intravenous injection for 7 days. Next, she received 40 mg per day of methylprednisolone intravenous injection and cyclophosphamide 0.8 g. After 3 days of treatment, her gastrointestinal symptoms completely disappeared. She received oral methylprednisolone at an initial dose of 32 mg per day and hydroxychloroquine 200 mg per day on discharge. A total of 1 month later, the patient returned to the hospital and received cyclophosphamide 0.8 g. Because computed tomography angiography (CTA) prompts intracranial aneurysm and internal carotid artery occlusion, she only received 100 mg of aspirin per day to treat APS.

In July, during a routine follow-up, 2 months after disease onset, she had no SLE related clinical complaints. However, she had some hormonal side effects, such as hallucinations, shaking hands, and weight gain. Her laboratory examinations results and CT findings before and after treatment are shown in Figs. 3 and 4. She continued on 400 mg/d hydroxychloroquine and 24 mg/d methylprednisolone. She has been undergoing follow-up and receives cyclophosphamide 0.8 g per month. In addition,

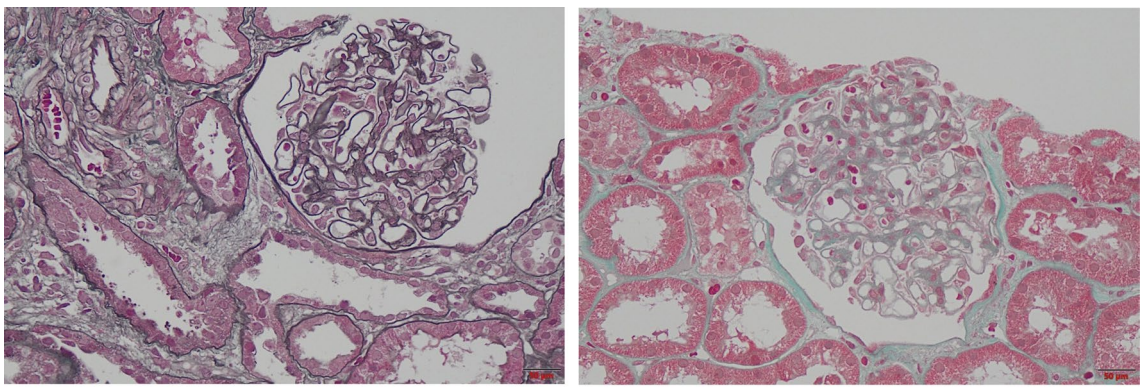

Fig. 1 Histopathological findings of the kidney in light microscopy. **A** Kidney biopsy revealing mild hyperplasia of glomerular mesangial cells and stroma in glomeruli (periodic acid-silver methenamine, 400×); **B** fuglobin deposition was observed in the mesangial region (Masson's trichrome stain, 400×)

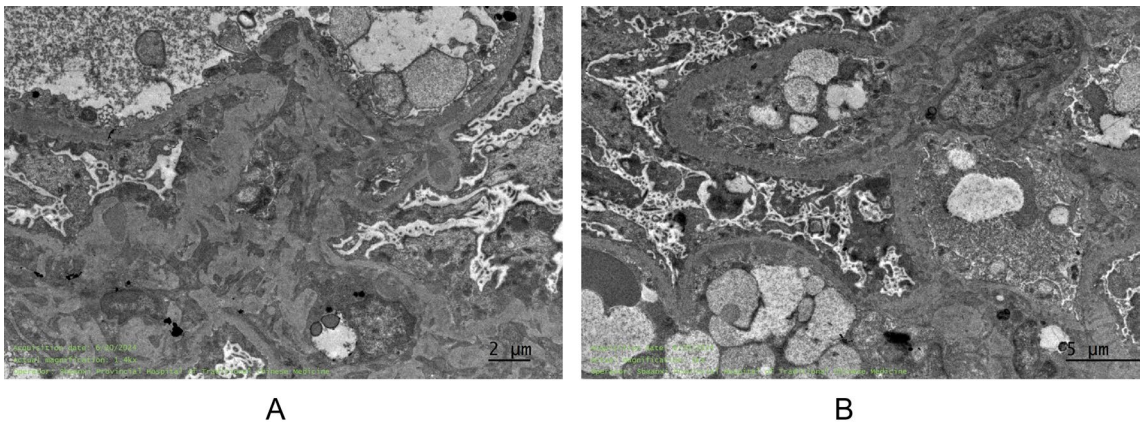

Fig. 2 Histopathological findings of the kidney in light microscopy in electron microscopy. **A** The electron density was deposited subepithelial and subendothelial region in glomeruli (14,000×); **B** the electron density was deposited mesangial region (10,000×)

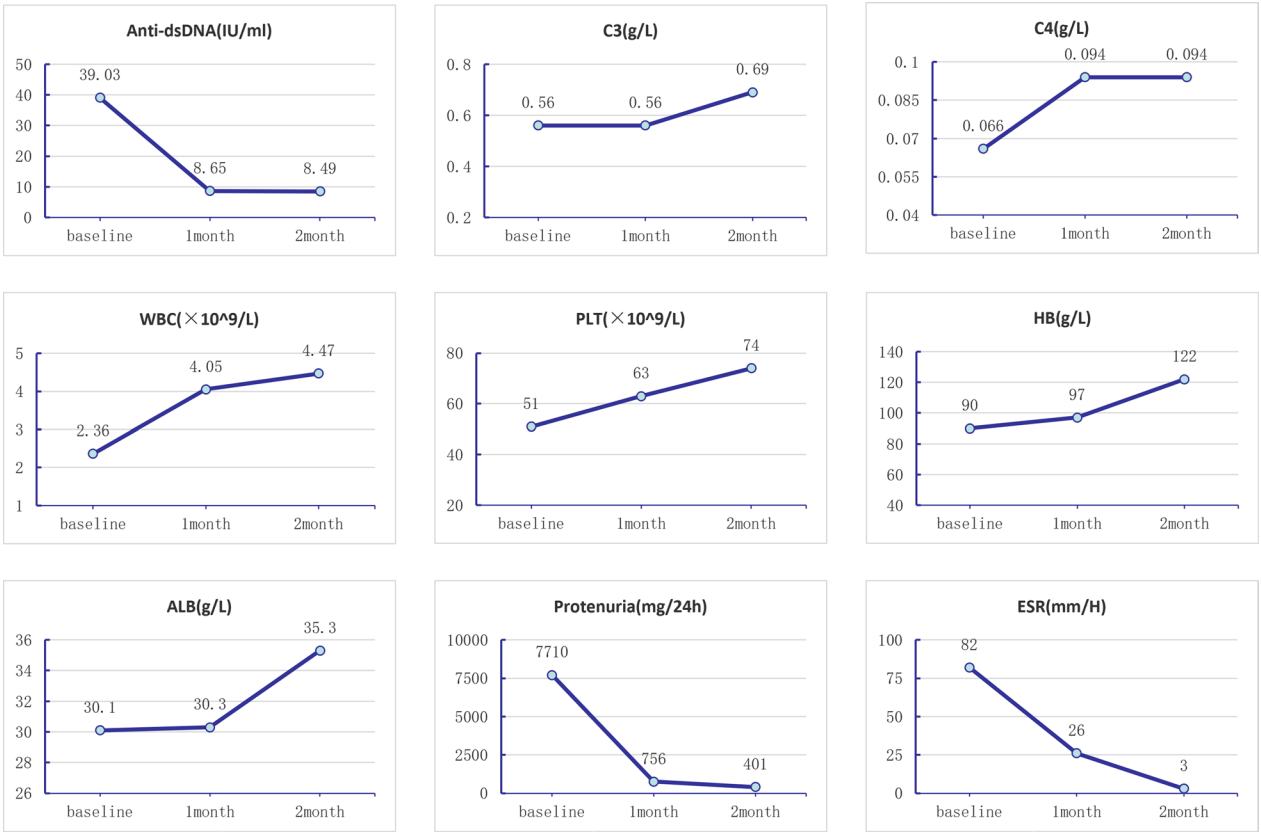

Fig. 3 Laboratory examinations results before and after treatment. *Anti-dsDNA* anti-double-stranded DNA antibody, *C3* complement 3, *C4* complement 4, *WBC* white cell count, *PLT* platelets, *HB* hemoglobin, *ALB* albumin, *ESR* erythrocyte sedimentation rate

the dosage of methylprednisolone will be gradually reduced.

Discussion

On admission the patient was positive for ANA and anti-dsDNA, albuminuria, thrombocytopenia, and hypocomplementemia, which fulfills the European League against Rheumatism Association criteria for the diagnosis of SLE. The main manifestation of the patient was gastrointestinal symptoms in the early stage of the disease. She was repeatedly diagnosed with and treated for acute pancreatitis, but her gastrointestinal symptoms stayed the same. Her gastrointestinal symptoms had completely disappeared after corticosteroid and human immunoglobulin therapy, which is consistent with SLE-related pancreatitis. At this point, autoimmune pancreatitis (AIP) also should be considered as a differential diagnosis. Most cases of AIP are type 1 AIP characterized clinically by frequent manifestation with obstructive jaundice, with or without a pancreatic mass [12]. It is a pancreatic manifestation of IgG4-related disease in which serum levels of IgG4 have the highest diagnostic value [12]. The patient showed neither obstructive jaundice nor a pancreatic mass. In addition, the immunoglobulin G4 was normal. Her manifestations, serological findings, and renal pathology results did not fulfill the criteria for the diagnosis of AIP. Systemic inflammatory response syndrome (SIRS) is a clinical response arising from a nonspecific insult such as pancreatitis [13]. Although the patient showed a white cell count of 2.18×10^9/L, her normal temperature (36.2 °C), respiratory rate (18 per minute), and pulse rate (86 beats per minute) did not fulfill the definition of SIRS. The patient did not show a remarkable increase in pancreatic enzyme levels in our case, which may be related to somatostatin therapy in the early stage of disease. Our case demonstrates the complicated manifestations of SLE and a rare initial manifestation as acute pancreatitis with APS.

Gastrointestinal involvement is a common complaint observed in 40–60% of patients with SLE, but acute pancreatitis as an initial manifestation is extremely rare [4, 14]. SLE pancreatitis might result from vasculitis, and histological features are both vasculitis and thrombosis [4]. Concomitant antiphospholipid antibodies (aPL) were found in SLE pancreatitis in a minority of cases [15, 16]. It is a life-threatening manifestation. There is no specific

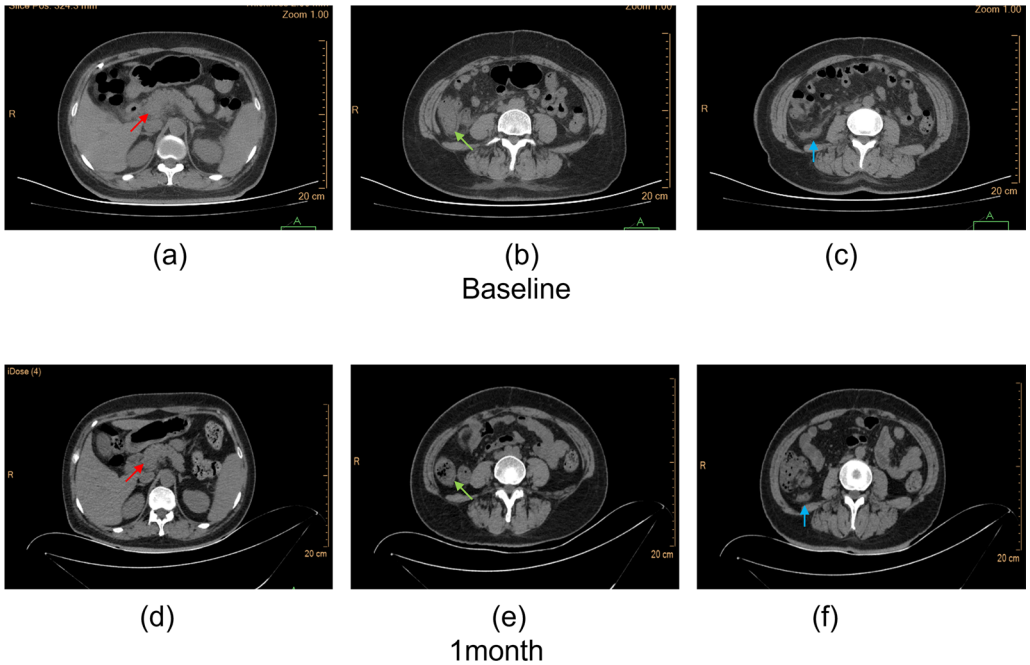

Fig. 4 Abdominal computed tomography findings before and after treatment. **a**–**c** Computed tomography of the abdomen exhibiting the swollen pancreas and inflammatory exudation around the pancreas (red arrow); the intestinal wall edema (double circle) in ascending colon (green arrow); the swollen cecum (blue arrow). **d-f** A total of 1 month after treatment; computed tomography of the abdomen exhibiting no obvious swelling or exudation in pancreas (red arrow), ascending colon (green arrow), or cecum (blue arrow)

treatment guideline. Many studies have confirmed that corticosteroids can be considered as a therapeutic modality and have been shown to decrease mortality in lupus-associated pancreatitis [17, 18]. The majority of patients were treated with corticosteroids and azathioprine, cyclophosphamide, and mycophenolate were commonly experiencing SLE gastrointestinal involvement [6]. On the basis of the treatment experience in our case, corticosteroids combined with cyclophosphamide can be effective in the treatment of SLE-related acute pancreatitis. However, before treating for SLE-related pancreatitis, other causes of pancreatitis should be ruled out in patients with SLE.

This is one of a few papers that report the renal pathology of SLE-related pancreatitis. Strikingly, the renal pathology of the patient was not severe, but the abdominal symptoms were severe. A similar presentation was observed in another patient in our Medical Center. This raises questions for our future research. It is important to balance renal pathology and gastrointestinal involvement when treating this condition.

Conclusion

Gastrointestinal involvement is a common presentation in SLE; however, SLE-related acute pancreatitis is extremely rare, particularly as the initial presentation of the disease. SLE-related acute pancreatitis should be considered in the differential diagnosis of patients with acute pancreatitis after the exclusion of other common causes and given treatment as soon as possible. Corticosteroids combined with cyclophosphamide are effective in the treatment of SLE-related acute pancreatitis.

Abbreviations

SLE	Systemic lupus erythematosus
APS	Antiphospholipid syndrome
CT	Computed tomography
ANA	Antinuclear antibody
dsDNA	Double-stranded DNA
AIP	Autoimmune pancreatitis
SIRS	Systemic inflammatory response syndrome

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Author contributions

STL reported this rare case of SLE. MYL wrote the article and STL edited it. All authors have read and approved the final manuscript.

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Availability of data and materials

Not applicable.

Declarations

Ethics approval and consent to participate
This case report was written and reported after obtaining the patient's informed consent. In addition, the case was approved by the Medical Ethics Committee of the Xi'an Central Hospital Affiliated to Xi'an Jiaotong University (LW-2024–044).

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
No financial or nonfinancial benefits have been received from any party related directly or indirectly to the subject of this article.

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