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Acute Recurrent Pancreatitis: An Uncommon Manifestation of Systemic Lupus Erythematosus in children

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Abstract

Systemic lupus erythematosus (SLE) is known as a complex multi-organ autoimmune disorder, which is characterized by systemic inflammation and presents with a variety of clinical manifestations. The relationship of SLE with pancreatitis in children is an uncommon and possibly life-threatening manifestation. Most of the previously reported cases showed that pancreatitis occurs in the setting of SLE with multi-organ involvement. In this report, we present a case of an eleven-year-old girl with the diagnosis of SLE-pancreatitis, which was an acute recurrent pancreatitis phenotype. In our case, pancreatitis was assumed to be a part of SLE exacerbation, accompanied by abnormalities in renal, hematologic and immunology tests. The diagnosis of acute recurrent pancreatitis was based on the signs and symptoms, laboratory tests such as serum and urinary amylase levels and computed tomographic findings. Treatment with high dose steroids and other forms of immunosuppressant drugs showed improvement in the patient. Physicians should consider SLE as an avoidable cause of pancreatitis in any SLE patient with complaints of gastrointestinal symptoms and elevated pancreatic enzymes.

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Introduction

Systemic lupus erythematosus (SLE) is known as a complex multi-organ autoimmune disorder, which is characterized by systemic inflammation resulting from antibodies directed against self-antigens, immunity dysfunction, and immune complex formation. It is predominantly found in females. SLE cases are diagnosed in 10% to 20% during childhood, where most studies use 18 years as the upper age limit. Childhood-onset SLE (cSLE) patients have more systemic involvements with active disease at presentation and overtime hence leading to bad outcomes [1]. Involvement of the pancreas is an uncommon cSLE manifestation which is potentially life-threatening [2, 3]. SLE-pancreatitis can appear early in the disease course of SLE, most frequently within the first two years of disease onset [2-4] and may be observed at the initial manifestation of disease [4]. The diagnosis of SLE pancreatitis can be given only after ruling out the common causes of pancreatitis, such as mechanical obstruction and toxic-metabolic insults related to pancreatitis, for example, alcohol, gallstones, hypertriglyceridemia, hypercalcemia, and medication [5, 6]. The possible mechanisms for SLE pancreatitis include vasculitis, microthrombi, immune complex deposition, and vascular intimal thickening and ischemia [4, 5]. The diagnosis of SLE pancreatitis is usually made on the basis of signs and symptoms, laboratory tests such as serum and urine amylase levels, and computed tomographic findings. Herein, we present a case history of an eleven-year-old Chinese girl who suffered from SLE nephritis with pancreatitis. The purpose of this case report is to draw attention to the risk of pancreatitis in children with SLE.

Case report

An eleven-year-old Chinese girl was admitted to our hospital with complaints of red erythematous rash on the face for twenty days. One week before admission, the patient had yellowish discoloration of skin and sclera associated with pale stool and dark urine. She also had oral ulcers, which were not painful. Upon admission, the patient was conscious and alert. Her vital signs showed: body temperature 36.8 °C, pulse rate 107 beats per minute, respiratory rate 25 times per minute and blood pressure 102/69 mmHg. The general physical examination showed mild pallor and icterus. A skin examination was also notable for malar rash characterized by erythema involving the cheeks and nasal bridge but sparing the nasolabial folds. The oral cavity showed diffuse and purulent ulcers. Examinations on the respiratory, cardiovascular and central nervous system revealed no abnormality. Laboratory results demonstrated a white blood cell count (WBC) 3.97×10^9/L, lymphocytes 29%, neutrophils 60.9%, hemoglobin 94g/L, platelet count 171×10^9/L, erythrocyte sedimentation rate (ESR) 22 mm/h, alanine aminotransferase 91 U/L, aspartate aminotransferase 563U/L, total protein 50.9g/L, albumin 19.5g/L, total bilirubin 161.8μmol/L, direct bilirubin 153.4umol/L, indirect bilirubin 8.4 μmol/L, creatinine 63umol/L, triglyceride level 7.96mmol/L, cholesterol level 6.26mmol/L, C3 0.263g/L, C4 <0.0688g/L. Urinalysis showed red blood cell count 153/μL, WBC 42/μL, urine protein 2+, occult blood 2+, bilirubin 2+, urine protein: creatinine ratio 3.62, amylase 559.98 U/L. Abdomen ultrasound showed edematous gallbladder wall and gallbladder bed with increased thickness and no gallstones. An abdominal plain computed tomography (CT) also showed gallbladder wall and gallbladder bed thickening, and the Glisson system was widened.

Immunological tests showed positive anti-nuclear antibodies (ANA) titer 1:1000, anti-double-stranded DNA positive, anti-histone antibody positive and anti-nucleosome antibody (ANuA) positive. According to the 1997 American Rheumatism Association diagnostic criteria, this patient fulfilled 4 criteria out of 11, so she was given the diagnosis as SLE with obstructive cholestasis. She was initially treated with oral prednisolone (2mg/kg/day in divided doses), reduced glutathione, compound glycyrrhizin and other supportive treatment. One day after the admission, she developed epigastric pain, which was acute in onset and progressive in nature associated with vomiting. The physical examination revealed slight abdomen distension with diffuse tenderness, especially on the epigastrium. She also had oliguria (urine volume <0.5 ml/kg/h) and high blood pressure 150/110mmHg. Tests showed serum amylase 793U/L, urine amylase 3454U/L, urine total protein 1.67 g/L and urine creatinine 4072umol/L, serum calcium <1.73 mmol/L. An abdominal plain computed tomography (CT) showed that the pancreas was enlarged and edematous, the density was not uniform and peripancreatic fluid was present (Fig. 1).

We further considered the patient to have acute pancreatitis with acute kidney injury (AKI). She had recurrent abdomen pain. The patient's

condition did not improve even after the third day of her hospital stay so she was kept on nothing per oral, parenteral nutrition and nasogastric suction were initiated. Intravenous imipenem, intravenous methylprednisolone, diuretics, magnesium sulfate, analgesics and other supportive treatments were given after which her abdominal pain improved. Urine volume increased to 1000ml/day. Repeated tests showed that the serum amylase was 258 U/L, and urine total protein was 0.78 g/L, whereas prothrombin time (PT) 47.1 seconds, International Normalized Ratio (INR) 4.27, creatinine 224umol/L, and BUN 10.7mmol/L were found. She was treated with methylprednisolone pulse therapy (at the time of acute presentation and during the episode of disease flare-ups) for 3 days and continuous renal replacement therapy (CRRT) for 6 days. Intravenous cyclophosphamide (CYC) pulse therapy at a dose of 0.2g was added. She gradually resumed her oral feeding. Her liver function tests returned to near normal level and jaundice disappeared after a month. Her abdomen ultrasound showed no abnormality of the gallbladder. She started complaining of abdomen pain again, which got better with intravenous fluids, antibiotics and supportive management for pancreatitis. After the patient was clinically stable, under local anesthesia, a kidney biopsy was performed. Immunofluorescent microscopic examination showed active and chronic lesions which were diffused proliferative glomerulonephritis with full-house staining pattern suggestive of Class IV (acute/chronic) Lupus Nephritis. Her test showed recurrent high amylase level, which was slowly decreased. On discharge, methylprednisolone was changed to oral prednisolone at a dose of 45mg per day which was in a tapering dose and hydroxychloroquine at a dose of 0.1g once a day was added.

Two months later, she was again admitted to the hospital for follow-up. This time, she had a fever for 2 days with high CRP 69mg/L and again developed paroxysmal abdomen pain. Serum amylase was 550U/L. Her CT abdomen was improved. She was managed as before for treating acute recurrent pancreatitis and was symptomatically relieved. Intravenous CYC had to be stopped because of her low CD4+/CD8+ratio 0.37. She was treated with tacrolimus at a dose of 0.5g twice a day and prednisolone 45mg per day, which was kept in a slow tapering dose. After the stay in the hospital for 5 weeks, her symptoms were improved and her amylase returned to near normal level.

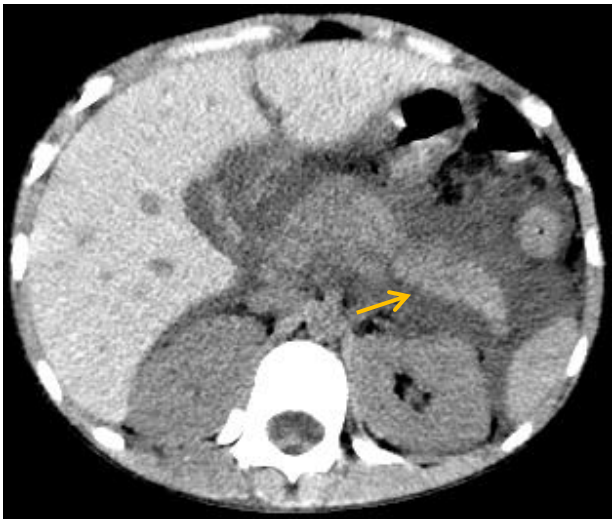

Fig. 1 Plain CT of the abdomen showing enlarged and edematous pancreas, an irregular contour of the pancreatic margins without uniform density and presence of peripancreatic fluid.

Discussion

Very few SLE-pancreatitis have been reported previously in both adult and pediatric SLE. Richer et al. [7] reported SLE pancreatitis in 6% children (12/201). In another study of pediatric SLE patients, 1.1% developed acute pancreatitis [8]. Marques et al. [9] reported a multicenter cohort study from 2012 to 2014 including 852 pediatric patients with cSLE, of whom 22 (2.6%) patients developed pancreatitis. Using the International Study Group of Pediatric Pancreatitis (INSPPIRE) standardized definitions; this study was the first report to typify pancreatitis in cSLE patients. In another study, 20 (91%) of the patients had acute pancreatitis, 2 (9%) had acute recurrent pancreatitis and none of them had chronic pancreatitis which revealed acute pancreatitis being the predominant type [9]. In many studies, the incidence of patients with SLE pancreatitis may be undervalued because patients who have gastrointestinal symptoms such as abdominal pain and vomiting may be thought as gastritis and treated accordingly rather than considering as pancreatitis hence, the pancreatic enzyme levels were not measured [10]. cSLE pancreatitis is often severe and has a significantly higher mortality rate ranging from 25% to 54% [5, 8, 9]. So it should be suspected in any patient with the symptom of abdomen pain. Early diagnosis and proper treatment are very important for a favorable prognosis. Our patient had acute recurrent pancreatitis, with more than 3 separate episodes of

acute pancreatitis with complete recovery of pain or complete normalization serum pancreatic enzyme levels, prior to the following occurrence of acute pancreatitis.

In our case, we had no evidence of gallbladder stone from abdominal ultrasound, no history of alcohol intake, no raised level of hypercalcemia, hypertriglyceridemia or hyper cholesterol which can cause pancreatitis. The patient initially had SLE along with obstructive jaundice as shown by the symptoms, laboratory tests and ultrasound of the abdomen. After the second day of admission, the patient was diagnosed to have acute pancreatitis. Her jaundice disappeared after a month and abdomen ultrasound showed no abnormality in the gallbladder, but there were still recurrent episodes of acute pancreatitis. The pancreatitis was considered to be a part of SLE exacerbation, accompanied by abnormalities in renal, hematologic and immunologic tests. Eaker et al. [11] showed controversy in SLE pancreatitis and steroid because steroid usage has been concerned to be the cause of SLE-pancreatitis. Whereas, studies also have reported SLE patients whose acute pancreatitis was improved and resulted in favorable outcomes with the administration of high doses of steroids [3, 12, 13]. The mortality rate of SLE patients who initially started on steroids following the diagnosis of pancreatitis was 20% compared to 61% among those without steroids [3]. In our case, the patient was given steroids since the first day of admission for the treatment of SLE, her pancreatitis was diagnosed 2 days after admission; however, we continued steroid at a high dose which was tapered slowly as pancreatitis improved. She was also prescribed with CYC for a certain time, which was stopped after her low CD4+/CD8+ratio and risk of infection. CYC was replaced with another immunosuppressant tacrolimus.

Conclusions

SLE-pancreatitis is an uncommon disease associated with a high mortality rate and may be under-recognized. SLE-pancreatitis should be considered as the differential diagnosis when SLE is active with acute abdomen pain. We should timely diagnose the condition of the patient and give proper treatment with steroids, in addition to other forms of immunosuppression and disease-modifying agents which is crucial in managing and improving outcome in SLE-pancreatitis.

Conflict of Interest

There are no conflicts of interest.

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