# Juvenile systemic lupus erythematosus and acute pancreatitis: a rare and potentially fatal presentation

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To the editor,

A 15-year-old Caucasian girl, with irrelevant prior medical history, was admitted to our hospital with a 4--week history of intermittent fever and anorexia, weight loss of 3 kg in 2 weeks, asthenia and polyarthralgia for 2 weeks. Over the last 2 days the patient also complained of mild abdominal pain. Physical examination showed a malar rash, periungual erythema, swollen and painful cervical and inguinal lymph nodes and synovitis of the wrists and proximal interphalangeal joints of the hands. There was no history of use of alcohol, drugs or toxic substances.

Initial investigations revealed: hemoglobin 11.1 g/dL, white cell count of $2.04\times10^9$/L with $0.71\times10^9$/L neutrophils, $1.00\times10^9$/L lymphocytes and $77\times10^9$/L platelets; erythrocyte sedimentation rate 43 mm/hour, C-reactive protein 0.7 mg/L. The liver and pancreatic enzymes were elevated: aspartate aminotransferase 994 U/L (normal 10-31), alanine aminotransferase 425 U/L (normal 10-31), alkaline phosphatase 177 U/L (normal 47-119), G-GT 209 U/L (normal 7-32), amylase 101 U/L (normal 22-880), lipase 96 U/L (normal 7-60). Blood urea nitrogen, creatinine, potassium, sodium, total and free calcium were within normal limits. Urinalysis revealed a protein to creatinine ratio of 1.82 with no other changes.

Chest radiography, electrocardiogram and abdominopelvic ultrasound findings were unremarkable. The echocardiogram showed a minimum pericardial effusion. During the second day of hospitalization, severe epigastric pain associated with nausea and vomiting ensued and at physical examination the epigastrium was tender to deep palpation without signs of peritoneal irritation. Amylase and lipase levels showed a marked increase (amylase 1317 U/L and lipase 6990 U/L). Abdominal computed tomography (without contrast) revealed diffuse pancreas globosity, medium volume ascites and signs suggestive of peritonitis. Concomitant infection was ruled out by negative cultures of blood, bone marrow, urine and negative serologies. Within 24 hours she developed nephritic syndrome with oligoanuric renal failure. The patient received intravenous fluid therapy, analgesia, empiric antibiotherapy, and was kept nil *per os*. Due to the suspicion of autoimmune disease-related pancreatitis, corticosteroids were initiated with pulses of methylprednisolone 1 g for 3 consecutive days, followed by oral prednisolone (1 mg/kg/day).

Further investigations confirmed a systemic lupus erythematosus (SLE) diagnosis with positive antinuclear, anti-double-stranded-DNA, anti-Smith and anti-ribonucleoprotein antibodies and low complement fractions levels. Anticardiolipin antibodies and lupus anticoagulant were negative. A renal biopsy performed after clinical improvement showed class IV lupus nephritis, and induction therapy with mycophenolate mofetil was started. The patient rapidly improved with normalization of pancreatic enzymes, and achieved clinical remission (SLE Disease Activity Index score decreased from 21 to 2) within 4 months.

There are few reported cases of juvenile SLE presenting with acute pancreatitis (AP) at the onset. It is an unusual manifestation, occurring in about 0.2–8.2% of SLE patients[1-4]. It typically occurs in females with clinically active SLE, within the first 2 years of disease, as in the case presented[1,5]. It tends to be more severe and is associated with higher mortality in pediatric patients when compared with adults[6,7].

A diagnosis of SLE-related pancreatitis can only be made after the exclusion of other causes of AP, including hepatobiliary disease, alcohol abuse, drug-induced toxicity, hypercalcemia, hypertriglyceridemia and infections[4,5].

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In this case, taking into account the clinical context and the fact that no other identifiable cause of AP could be found, it was considered a primary SLE manifestation.

The pathophysiology of pancreatitis in SLE is unclear, but several mechanisms have been suggested to play a role, including: vasculitis, micro-thrombus formation, anti-pancreatic antibodies, inflammation due to T-cell infiltration, complement activation, immune complex deposition and infections[3-5,8].

This case emphasizes the importance of considering AP secondary to SLE in a patient presenting abdominal pain, as early recognition will guide appropriate treatment and lead to favorable outcomes[1,2,7].

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