

SYSTEMIC LUPUS ERYTHEMATOSUS (SLE) PRESENTING AS ACUTE PANCREATITIS - A CASE REPORT

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ABSTRACT

Acute pancreatitis is an uncommon manifestation of systemic lupus erythematosus (SLE). In most reported cases, it occurs in the setting of SLE with multi-organ involvement. We report an unusual case of SLE in a Sri Lankan woman presenting with acute pancreatitis with the pancreas as the sole major organ affected. The diagnosis of acute pancreatitis was based on clinical features, serum and urinary amylase levels and computerised tomography. She responded well to high-dose corticosteroid. We also review the literature and discuss the prevalence, theories of pathogenesis and treatment of this condition.

Keywords: acute pancreatitis, systemic lupus erythematosus, amylase, computerised tomogram, corticosteroid

SINGAPORE MED J 1996; Vol 37: 323-324

INTRODUCTION

Acute pancreatitis is an uncommon manifestation of SLE. It is extremely rare for a SLE patient to have acute pancreatitis as the initial presentation. It usually manifests as an acute surgical abdomen. In a young female with acute pancreatitis without the usual aetiological causes of gallstone disease and alcohol abuse, a high index of suspicion of lupus will lead to prompt diagnosis and appropriate management. We report the clinical history of one such patient, followed by a literature review of pancreatitis in lupus.

CASE REPORT

HMD, a 30-year-old Sri Lankan woman, was apparently well until a week before presentation in December 1993 when she had symptoms of an upper respiratory tract infection (URTI). She has been working as a domestic help in Singapore for four years. She did not consume alcohol and had not travelled recently. She is nulliparous and does not suffer from any illnesses. She was prescribed only paracetamol and chlorpheniramine by a private practitioner for her URTI. She developed a rash on her trunk a week later and because it was thought to be due to drug allergy, she was warded at a skin centre. When she subsequently developed vomiting, diarrhoea and epigastric pain, she was transferred to the surgical department of Tan Tock Seng Hospital on the same day for the management of a suspected acute abdomen.

She was febrile on arrival. There was diffuse alopecia and a maculopapular rash over the trunk. There was no malar rash, photosensitivity, discoid lesions, arthritis, aphthous ulcers or ankle swelling. There was epigastric tenderness, but no visceromegaly or evidence of an acute abdomen.

Investigations suggested a diagnosis of acute pancreatitis. The haemoglobin was 11.1 g/dL and total white count was $8.1 \times 10^9/L$. The urea was 3.0 mmol/L, creatinine 48 $\mu\text{mol/L}$, sodium

129 mmol/L, potassium 3.9 mmol/L, chloride 113 mmol/L and glucose 9.7 mmol/L, the total protein was 67 g/L (normal range 62 - 82), albumin 29 g/L (normal range 37 - 51), total bilirubin 19 $\mu\text{mol/L}$ (normal range 3 - 24), alkaline phosphatase 442 U/L (normal range 32 - 103), alanine transaminase (ALT) 185 U/L (normal range 7 - 36), aspartate transaminase (AST) 103 U/L (normal range 15 - 33), gamma-glutamyl transferase (GGT) 334 U/L (normal range 5 - 26), HBSAg and anti-HIV were negative. The serum amylase was 253 U/L (normal range 30 - 100) on admission; it peaked at 310 U/L on the seventh day. The urinary diastase was 3613 U/L (normal range 32 - 641) and it peaked at 5796 U/L on the sixth day. Abdominal ultrasound showed no space-occupying lesion, gallstone or biliary ductal obstruction. The common bile duct was 0.4 cm in diameter. The patient was treated conservatively and discharged on the eighth day.

Nine days later, she was re-admitted for fever and diarrhoea for three days. Her general condition was fair, her temperature was 40°C, pulse 108/min, blood pressure 100/70 mmHg. A generalised confluent macular rash with areas of desquamation was noted. The abdomen was normal.

The haemoglobin was 8.9 g/dL, total white $6.3 \times 10^9/L$, platelet $122 \times 10^9/L$, the urea was 7.1 mmol/L, creatinine 59 $\mu\text{mol/L}$, sodium 130 mmol/L, potassium 4.2 mmol/L, chloride 108 mmol/L and glucose 4.1 mmol/L. The total protein was 67 g/L, albumin 25 g/L, total bilirubin 5 $\mu\text{mol/L}$, alkaline phosphatase 151 U/L, ALT 27 U/L, AST 56 U/L and GGT 98 U/L. The serum amylase was 297 U/L, the urinary diastase 3613 U/L, and they were 250 U/L and 1464 U/L respectively three days later. The serum iron and binding capacity were both low.

She was diagnosed to have acute relapsing pancreatitis, and was given supportive treatment. Intravenous ceftriaxone and metronidazole was administered empirically. She improved and was discharged nine days later.

Outpatient review two weeks later showed that the patient was febrile (37.8°C) and had myalgia. She also developed left ankle arthritis, and was admitted for further investigations.

On admission, the haemoglobin was 9.0 g/dL, total white $7.3 \times 10^9/L$, platelet $170 \times 10^9/L$, the reticulocyte count 1.0% and the erythrocyte sedimentation rate (ESR) 127 mm/hour. Total protein was 73 g/L, albumin 30 g/L, total bilirubin 9 $\mu\text{mol/L}$, alkaline phosphatase 113 U/L, ALT 18 U/L and AST 36 U/L. The antinuclear antibody (ANA) was positive at a titre > 1/640 with a homogeneous pattern, the double-stranded deoxyribonucleic acid antibody (anti-ds DNA) was raised at 414.4 IU/mL (normal < 12.2) and the anti-cardiolipin antibody was positive at 34.1 gpl (normal < 11.0). The VDRL was negative.

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The triglyceride and calcium levels were normal. The extractable nuclear antibodies (anti-Sm, anti-Ro, anti-La and anti-ribonuclear antibodies) were negative. The computerised tomogram (CT scan) of the abdomen showed that the pancreas, particularly the tail, was bulky and enlarged with ill-defined outlines. The pancreatic duct was not dilated and slight thickening of the adjacent loops of bowel was noted. There was no pseudocyst formation.

The patient was diagnosed to have acute pancreatitis with SLE. She was treated with prednisolone at a dose of 1 mg/kg/day. She was discharged after a week's stay. Her symptoms improved, including the alopecia. She did not have any recurrence of the abdominal pain and she could resume her work. She has been on regular review for one year and a half at the time of writing and the prednisolone dose is being tapered gradually.

DISCUSSION

Acute pancreatitis is known to be an uncommon manifestation of SLE since its first description in 1939⁽¹⁾. The prevalence has previously been reported to range from 4% to 8.3%⁽²⁻⁴⁾. However, when cases with co-incident risk factors are excluded, such as alcohol abuse, surgery and gallstones, the number purely attributable to SLE are very few indeed⁽⁵⁾.

Seventy-two cases of acute pancreatitis^(1,2,4,6-10) and three cases of chronic pancreatitis^(11,12) in SLE have been described in the literature so far. Almost all cases of acute pancreatitis occurred in the setting of disseminated, active, multi-organ lupus, and the diagnosis has only been made post-mortem in some of them^(1,4).

Our patient is the sixth reported patient with SLE in the literature whose initial manifestation was acute pancreatitis^(4,6,13-15), the 13th who was on no significant medication before the illness⁽⁶⁾, and the first Indian. In addition, the major organ involvement in her case appears to be limited solely to the pancreas, to such an extent that the diagnosis of SLE was missed initially.

We are confident of a diagnosis of acute pancreatitis in her even though the serum amylase was only raised to three times the upper limit of normal and the amylase isoenzymes and lipase levels are not determined (these tests being unavailable) because of the typical history and the oedematous pancreas visualised on the CT scan⁽¹⁶⁾.

Our patient did not have any other conditions that may predispose to pancreatitis like trauma, surgery, gallstones, alcoholism, hypercalcaemia or hypertriglyceridemia. She was on paracetamol and chlorpheniramine for one week, but these drugs are not known to cause pancreatitis. Although the patient had symptoms of URTI initially, we think that a viral aetiology is unlikely because besides mumps, which is occasionally associated with pancreatitis, the role of other viruses is speculative⁽¹⁷⁾. We conclude that the pancreatitis is secondary to SLE.

The patient does not have secondary antiphospholipid antibody syndrome though the anti-cardiolipin antibody (ACA) is positive because there are no associated features of thromboses, recurrent abortions, thrombocytopenia or false-positive VDRL. We do not think that the ACA has a pathogenetic role in this case.

The theories of the pathogenesis of acute pancreatitis in SLE have recently been reviewed by Petri⁽⁵⁾. She listed the possible mechanisms of pancreatic vascular events (thrombosis and vasculitis), inspissated secretions attributed to corticosteroids and necrotising pancreatitis. Hypothetical mechanisms include complement activation, hypotension and pancreatic antibodies. The role of the anti-phospholipid syndrome is unclear at the present time^(5,10).

Another controversial issue is that some of the drugs used to treat SLE are believed to cause pancreatitis. These include

frusemide, thiazide, azathioprine, danazol and especially corticosteroids. However, Steinberg et al concluded that corticosteroids are unlikely to cause pancreatitis, based on their analyses of published case reports and animal studies⁽¹⁸⁾.

The mainstay of treatment is use of corticosteroid and supportive care. Our experience in this case is similar to those of previous authors who have used high-dose prednisolone to treat SLE patients with pancreatitis; both the abdominal and systemic features resolved. Recently, a 15-year-old girl with severe SLE with multi-system involvement (including acute pancreatitis and pseudocyst-formation) who did not respond to high-dose steroids was successfully treated with plasmapheresis⁽⁹⁾.

This patient reminds us that SLE must be considered as a possible underlying diagnosis in every case of apparently idiopathic acute pancreatitis in a young female, even in the absence of obvious clinical features of SLE.

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