Acute lethal pancreatitis in childhood systemic lupus erythematosus

Childhood systemic lupus erythematosus (SLE) is a multisystem disease with protean manifestations. Pancreatitis in adult-onset lupus is a well recognized but rare complication. We report a child with SLE who developed fatal pancreatitis shortly after presentation. We believe that there are only three other reports of this complication in the literature, and our report is the first case of fatal pancreatitis on initial presentation [1–2].

A 14-yr-old girl presented to a district general hospital with a 4-week history of rashes on her face, later spreading to the trunk. She had stomatitis, vomiting, diarrhoea, anorexia and marked weight loss. On examination, her weight was 40 kg (along 9th centile). She was unwell, listless and febrile and had severe gingivostomatitis, a scaly butterfly rash on her cheeks and punched-out vasculitic lesions on her forehead and both knees. She also had vasculitic lesions over the tragus and index finger, petechiae over the buccal mucosa and palate, retinal haemorrhages and joint involvement. Her blood pressure was 110/70 mmHg. Systemic examination revealed no other abnormalities. The only family history of note was that her mother had type 1 neurofibromatosis.

Initial investigations showed a haemoglobin concentration of 10.4 g/dl, a white cell count of $1.4 \times 10^{9}/μl$, a platelet concentration of 61 000/μl and an erythrocyte sedimentation rate of 55 mm/h (normally < 12 mm/h). The direct Coomb’s test was positive and the activated partial prothrombin time was increased (40.8 s) (Control-32). Her renal parameters showed sodium 139 mEq/l, potassium 4.2 mEq/l, urea 11.3 mmol/l and creatinine 112 μmol/l. Liver function analyses showed an albumin concentration of 25 g/l, alanine transaminase 170 IU/l (normal range 0–40 IU/l) and serum
bilinear 22 µmol/l. Urine analysis revealed haematuria and proteinuria. Concentrations of complement components were very low [CH50 < 7 U/ml (normal range 25–45 U/ml), C3 0.20 g/l (normal range 0.80–1.40 g/l), C4 0.03 g/l (normal range 0.15–0.40 g/l)] and immunology screening showed strong positivity for double-stranded DNA antibodies (>100 IU/l), immunoglobulin G (IgG), cardiolipin antibody (35 gpu; normal range 0–10 gpu) and IgG antinuclear antibodies (1/1000). Echocardiogram showed moderate left ventricular hypertrophy and mild mitral valve regurgitation with prolapse.

The patient was transferred to our tertiary centre, where a clinical diagnosis of SLE was made, and in view of her neutropenia she was commenced on intravenous methylprednisolone at 15 mg of her neutropenia she was commenced on intravenous methylprednisolone at 15 mg/kg/day and antibiotics. Over the next few days, she developed diffuse abdominal tenderness and haematemesis with raised serum amylase (848 IU/l; normal range 12–87 IU/l). Barium meal showed no significant lesion. Laparoscopy performed after she had developed an acute abdomen with peritonitis showed pancreatitis with free peritoneal fluid and multiple areas of fat necrosis.

Post-operatively, she became anuric and hypotensive, resulting in acute renal failure with severe metabolic acidosis requiring haemodialysis. Ultrasound examination of her kidneys showed loss of corticomedullary differentiation, which was consistent with acute renal failure. Her methylprednisolone dose was increased to 30 mg/kg/day. She later developed breathing difficulty and her chest X-ray revealed bilateral pleural effusions, for which paracentesis was performed. Pericardial rub was noted on examination and repeat echocardiogram showed changes of left ventricular hypertrophy with a thin rim of pericardial fluid suggestive of myocardial involvement. Her blood sugars rose to 16.8 mmol/l and insulin treatment was started.

She again complained of abdominal pain and developed another acute abdomen. Laparotomy showed two jejunal perforations that were repaired and histology showed a severe necrotizing vasculitis of the jejunal wall and fat necrosis secondary to acute pancreatitis in the omentum. Post-operatively, she required ventilation in intensive care. A urinary tract infection with candida was treated with amphotericin and haemodialysis was continued for persistent metabolic acidosis and azotaemia. The patient continued to deteriorate, with persistence of disease activity despite the use of high-dose steroids and daily plasma exchanges, and she developed Aspergillus fumus infection in her upper respiratory tract, for which fluconosine was commenced. She developed large collections of clot and fluid, one in the pelvis and another in the upper abdomen. She later developed bradycardia and went into asystole, from which she was initially resuscitated; however, she remained hypotensive and died soon afterwards. The course of her illness from presentation was less than 1 month and she remained in hospital throughout, mostly in the intensive care unit.

At post-mortem, widespread petechial rashes were noted on her arms, legs and trunk. Her peritoneal cavity contained several litres of fresh blood and blood clots, her pericardial cavity contained 100 ml of serous fluid and a small endocardial haemorrhage was present on the posterior wall of her left ventricle. Both her lungs were totally consolidated, with thick green pus exuding from the cut surfaces of bronchi. Severe haemorrhagic necrosis with dense adhesions were present between bowel loops and the anterior and posterior abdominal walls. Gross fat necrosis was present throughout the abdominal cavity. Her bile ducts and pancreas could not be identified in the necrotic tissue present in the abdomen. Her liver was markedly enlarged, with a focal area of necrosis in the right lobe, and both kidneys were generally enlarged, pale and necrotic. Her uterus, Fallopian tubes and ovaries appeared totally necrotic and were covered in dense adhesions in a haemorrhagic mass in the pelvis. The final pathological diagnosis was fulminating SLE affecting the gastrointestinal tract, pancreas, skin and kidneys with terminal pulmonary aspergillosis.

SLE is a multisystem disease with the potential to involve almost any organ in the body. Pancreatitis is a rare complication in adult patients with lupus [4–7]. The aetiology of pancreatitis in lupus patients is still unclear. Immunosuppressive medications, such as corticosteroids, cyclosporin and azathioprine, have been implicated in several case reports [8, 9]. Vasculitis with ischaemia of the pancreas has also been implicated [10].

Our patient, who presented with classical signs of childhood lupus, deteriorated rapidly and died from fulminant pancreatitis. We believe that severe vasculitis with ischaemia of the pancreas was the explanation for the fulminant pancreatitis in our patient. We would like to highlight this extremely rare, albeit serious, complication in childhood lupus. We are aware of only two other case reports of pancreatitis in childhood lupus [1, 2]. In both reports, immunosuppressive therapy (primarily steroids) was thought to have precipitated pancreatitis. We believe that this is the first case report of severe pancreatitis secondary to vasculitis presenting at the onset of lupus disease in a child. In our opinion, this extremely rare complication must be borne in mind when managing children with lupus, particularly in the presence of any abdominal symptoms.

A. V. Ramanan, A. D. Thimmarayappa, E. M. Baildam

Department of Paediatric Rheumatology, Royal Manchester Children’s Hospital, Charlestown Road, Blackley, Manchester M9 7AA, UK
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Correspondence to: A. V. Ramanan, Flat 508, 77 Elm Street, Toronto, Ontario, Canada M5G 1H4.


