

Cardiac tamponade as an initial manifestation of systemic lupus erythematosus in early childhood

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Abstract

Cardiac tamponade is rare as an initial manifestation of systemic lupus erythematosus (SLE), and even more so in paediatric patients. This paper reports an 8 year old girl with SLE with several unusual features: unusual age of presentation, unusual initial organ manifestation and recurrent cardiac tamponade as a complication.

Systemic lupus erythematosus (SLE) is an uncommon disease in childhood with an estimated prevalence of 0.6 in 100 000.¹ Cardiac involvement is rare as an initial manifestation of the disease, more so in paediatric patients.²⁻³ It is rarer still to encounter cardiac tamponade as the initial manifestation in SLE. Thirteen cases of cardiac tamponade in SLE have been reported.⁴ In only six of these was cardiac tamponade the initial presenting feature and all of these patients were older than 12 years.⁴⁻⁶

We report here an 8 year old girl who presented with recurrent cardiac tamponade; the first episode of this was at about three years of age.

Case report

An 8 year old girl was referred to us with a five year history of recurrent episodes of breathlessness. At the age of three years she was diagnosed as having pericardial effusion and 200 ml of fluid was aspirated twice. She was treated with antituberculous drugs (isoniazid 10 mg/kg daily and rifampicin 10 mg/kg daily) for one year. She remained asymptomatic for the next four and half years. She then again developed breathlessness, this time accompanied by pain in the small and large joints and hands and feet. Pericardial effusion was present. About 1000 ml fluid was aspirated twice at an interval of two months. Again, a provisional diagnosis of tuberculous pericarditis was made and anti-tuberculous treatment was reinstated. Her condition progressively worsened and she was referred to this hospital.

Physical examination showed a sick child with a pulse rate of 120/min and blood pressure of 100/70 mm Hg. The pulse volume was low and pulsus paradoxus of 16 mm Hg was present. Her respiratory rate was 50/min and the jugular venous pulse was raised with an absence of diastolic descent. Cardiovascular examination revealed cardiomegaly with muffled heart sounds. Ewart's sign was positive and there was hepatomegaly of 10 cm below the right costal margin. In view of the patient's

medical history a working diagnosis of recurrent cardiac tamponade as a complication of SLE was made.

Investigations revealed haemoglobin 137 g/l and a total leucocytic count of $8.8 \times 10^9/l$ with 57% neutrophils and 40% lymphocytes. The erythrocyte sedimentation rate was 23 mm/h. A chest radiograph showed an enlarged cardiac silhouette with low voltage impulses on the electrocardiogram. Echocardiography showed a massive pericardial effusion with features of cardiac tamponade (figure). An immediate pericardiocentesis was performed under echocardiographic guidance and 500 ml of fluid aspirated. Biochemical analysis of the pericardial fluid revealed 57 g/l protein. The fluid was acellular on microscopic examination. No microorganism was seen on gram staining and a Zeihl Neelsen stain of the smear. Cultures for aerobic bacteria and *Mycobacterium tuberculosis* were sterile. A tuberculin test was non-reactive to 1 TU (PPD RT-25 with Tween 80). The serum was strongly positive for antinuclear antibodies and the dsDNA antibody. On fluorescent microscopy, the antinuclear antibodies had a membranous and speckled pattern. The rheumatoid factor and lupus erythematosus cell were negative. The concentration of C₃ in serum was 0.92 g/l and that in the pericardial fluid was 0.42 g/l (control 1.00 g/l). A urine analysis for protein and formed elements was negative.

The diagnosis of SLE was confirmed and prednisolone 2 mg/kg daily in three divided doses was started. The dyspnoea and arthralgia responded. Repeat echocardiographic examination showed a decrease in the amount of pericardial fluid and the patient was discharged. However, three weeks later she was again readmitted with signs of pericardial effusion



Figure Echocardiograph showing massive pericardial effusion in an 8 year old patient with SLE.

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with cardiac tamponade. She required pericardiocentesis three times during this admission. In view of the recurrent cardiac tamponade, an anterior pericardiotomy was performed. Histopathology revealed features of chronic non-specific pericarditis.

Discussion

This patient shows several unusual features of SLE: unusual age of presentation (prepubertal onset), unusual initial organ manifestation of SLE, and an unusual complication of recurrent cardiac tamponade. Very few children under five years show an onset of the disease; most are older than 10 years when diagnosed.⁷ Fever with non-erosive deforming arthritis is the most common presenting feature of SLE.

Reports of cardiac tamponade as an initial manifestation of SLE are rare.²⁻³ It has been described in only 11 (0.8%) of 1332 patients in a combined series representing cases over a 50 year period.⁸ Of these, only two cases occurred in children in their second decade.⁵⁻⁶ Our patient is the youngest child with SLE with recurrent cardiac tamponade. Surgery was resorted to in the last episode of cardiac tamponade, which occurred three weeks after prednisolone treatment had begun. Although prednisolone promptly brings relief to such patients, there are reports of the failure of pericardial effusion to reduce after treatment with corticosteroids, and massive pericardial effusion can occur while on adequate doses of corticosteroids.⁵

This patient fulfils the 1982 revised criteria for the diagnosis of SLE in children (arth-

ralgias, serositis, haematological manifestations, immunological manifestations, and positive antinuclear antibodies).⁹ This patient had received isoniazid, a drug reported to cause SLE.¹⁰ We did not consider the diagnosis of isoniazid induced SLE because the first episode of pericardial effusion occurred before any drug treatment. In addition, her symptoms recurred despite discontinuation of the drug and antinuclear antibodies remained positive, showing a membranous rim with a speckled pattern on fluorescent microscopy. This correlates well with the trypsin sensitive non-nucleic acid antibody which is considered specific for SLE. The presence of the dsDNA antibody confirms the diagnosis of SLE. Low complement levels in the blood and pericardial fluids further support this diagnosis.

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