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Brief Report

Cardiac tamponade as an initial manifestation of systemic lupus erythematosus in a child

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Abstract Cardiac involvement is a rare initial presentation of systemic lupus erythematosus. An 11-year-old girl was described to have massive haemorrhagic pericardial effusion and cardiac tamponade, which was later diagnosed as systemic lupus erythematosus. Therefore, in children presenting with cardiac tamponade, systemic lupus erythematosus should be considered as one of the differential diagnoses, as morbidity and mortality associated with cardiac tamponade can be dramatically reduced with early diagnosis and use of steroids.

Keywords: Cardiac tamponade; pericardial effusions; pericarditis; systemic lupus erythematosus

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Systemic lupus erythematosus is an autoimmune disease characterised by the presence of auto-antibodies directed against the self-antigen of the body. Cardiac involvement as an initial manifestation is rare in patients with systemic lupus erythematosus, especially in the paediatric age group, and presenting as cardiac tamponade is even more rare.[1] In this report, we present the case of an 11-year-old girl who presented with cardiac tamponade due to massive pericardial effusion in the emergency department. The child improved on pericardiocentesis along with steroids, and the diagnosis of systemic lupus erythematosus was established.

Case history

An 11-year-old female patient was admitted in another hospital with a recent history of dyspnoea and petechial rash on both lower limbs. There was no history of fever or any bleeding manifestation. The child had a history of epilepsy at 18 months of age, and thus a contrast-enhanced computerised tomography of the head and electroencephalography were done at that age, both of which were normal. Anti-epileptic treatment was given for 2 years, and since then the child has been apparently well.

An echocardiogram was performed and a massive pericardial effusion was found. Ultrasonography was done, which showed moderate pericardial effusion and mild pleural effusion. Her haemodynamic status deteriorated and she was transferred to our hospital. On arrival, physical examination revealed a sick child with a pulse rate of 120/min, blood pressure of 106/68 mmHg, and a low pulse volume with a pulsus paradoxus of 20 mmHg. Her respiratory rate was 45/min and the jugular venous pulse was raised. Cardiovascular examination revealed cardiomegaly with muffled heart sounds. The liver was palpable 7 cm below the right costal margin. Low-voltage electrocardiogram was reported. The chest X-ray showed cardiomegaly with clear lungs (Fig 1). The child had petechiae and purpuric rash over both feet and legs. Echocardiography showed massive pericardial effusion with features of cardiac tamponade, and thus pericardiocentesis was performed under echocardiographic guidance and 500 ml of haemorrhagic fluid was aspirated. Meanwhile, the child became clinically stable, but after 20 h features of cardiac tamponade developed and pericardiocentesis was done. About 300 ml of haemorrhagic fluid was aspirated and the child was put on continuous drainage, which led to a drainage of 250 ml in 2 days.

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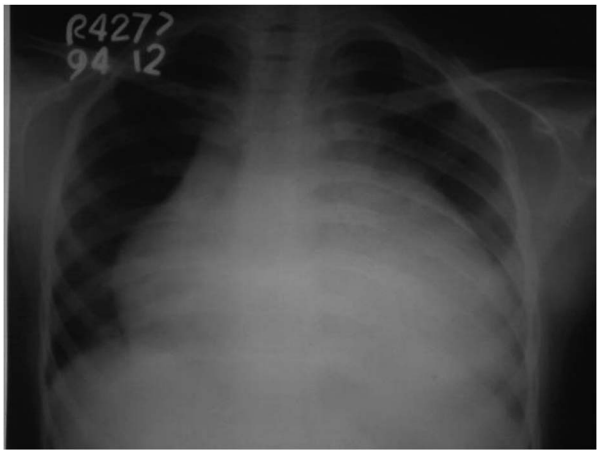

Figure 1.
Chest X-ray showing a massive pericardial effusion.

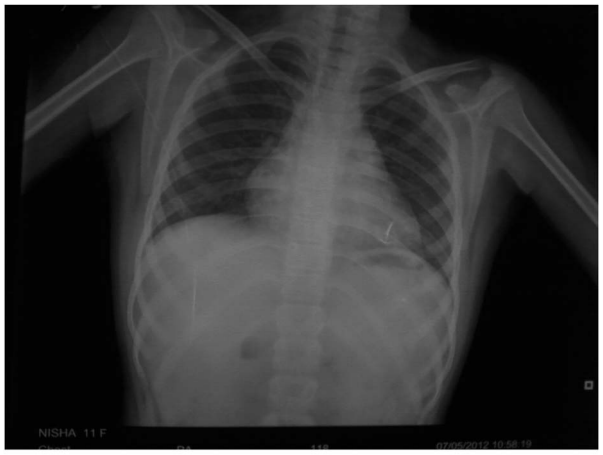

Figure 2.
Chest X-ray after pericardiocentesis and start of steroids.

Investigations revealed a haemoglobin concentration of 106 g/L, a total leucocyte count of 8.8×10^9/L with 38% neutrophils and 60% lymphocytes, and a platelet count of 50,000. The erythrocyte sedimentation rate was 60 mm/h. Biochemical analysis of the pericardial fluid revealed 60 g/L protein. The pericardial fluid was serosanginuous (340 cells with 80% polymorphs and 20% lymphocytes) on microscopic examination. No microorganism was seen on Gram staining, nor on the Ziehl–Neelsen staining of the smear. Fluid polymerase chain reaction for mycotuberculosis and common viruses was negative. The tuberculin test was non-reactive. Her coagulation profile was normal. Serum was strongly positive for double-stranded DNA antibody 250 (normal <30) and the antinuclear antibodies 9.8 (positive >1.4); C_3 was low 0.5 (normal 0.9–2.1). Urinalysis and kidney function tests were normal.

The diagnosis of systemic lupus erythematosus was confirmed as per the 1997 revised classification criteria for systemic lupus erythematosus, and thus prednisolone 2 mg/kg daily in three divided doses was started. The child responded well to treatment, and thus the repeat chest X-ray picture was improved (Fig 2), with echocardiographic examination showing a minimal amount of pericardial fluid. Platelet count was on the increasing trend and became normal within 4 days of starting steroids. The child was discharged on daily steroids, that is, prednisolone 1 mg/kg.

Discussion

An 11-year-old female child who was apparently well presented with sudden onset cardiac tamponade along with purpuric rash over the lower limbs with no present or past history of any febrile episode, and thus the possibility of rheumatological cause (systemic lupus erythematosus), although rare, is considered as part of our work-up. Subsequently, positive antinuclear antibody levels and anti-double-stranded DNA Levels supported our diagnosis. As per 1997 revised criteria for diagnosis of systemic lupus erythematosus, which include 4 out of 11 (malar rash, discoid rash, photosensitivity, oral ulcers, nonerosive arthritis, pleuritis or pericarditis, positive antinuclear antibodies, immunological (anti-DNA or anti-Sm or positive anti-phospholipid antibodies false positive Venereal Disease Research Laboratory, renal involvement, neurologic disorder and haematological involvement (haemolytic anaemia or lymphopenia or thrombocytopenia) our child satisfied criteria with haematological, serositis, immunological and positive antinuclear antibody findings.

Cardiac tamponade is a rare initial manifestation of systemic lupus erythematosus.[1] It has only been described in 0.8% of patients in a combined series representing cases over a 50-year period.[2]

The child had a past history of epilepsy at the age of 18 months. Although seizures can occur in systemic lupus erythematosus, its association with systemic lupus erythematosus could not be established as the child was asymptomatic over the 2-year anti-epileptic treatment period and seizures never occurred afterwards. The pericardial fluid was found to be serosanginuous and haemorrhagic. In some case reports, pericardial fluid is either haemorrhagic or serosanginuous.[3,4] In our child, the fluid was grossly

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haemorrhagic, and on cytological examination the field was full of red blood cells along with the presence of 340 cells – 80% polymorphs and 20% lymphocytes – but repeated culture gram staining and viral PCR of the fluid revealed no organism.

The child had initial improvement with pericardiocentesis but deterioration occurred after 20 hours, requiring pericardiocentesis again followed by continuous drainage. By the time the child was considered for a second pericardiocentesis, steroids were started, which lead to a drastic improvement in symptomatology along with a rising trend of platelet count.

In conclusion, systemic lupus erythematosus-related pericardial effusion should be kept in mind as a rare cause of cardiac tamponade even in patients without previous diagnosis or history suggestive of systemic lupus erythematosus.

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