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Case Report

Cardiac Tamponade as The Initial Presentation of Childhood Systemic Lupus Erythematosus: A Case Report

***Kevin Bassey[1], Frances SamOkpokowuruk,.[1,2], Ifunanya Ularinma Ebiekpi, [1], Idorenyin Diana Etebong[1]**

[1]Department of Paediatrics, University of Uyo Teaching Hospital, Uyo, Nigeria, [2]Department of Paediatrics, University of Uyo, Uyo, Nigeria

Abstract

Systemic Lupus erythematosus (SLE) is an autoimmune disorder characterized by the proliferation of autoantibodies and immune dysregulation resulting in damage to many body organs. Pediatric SLE usually presents with fever, joint pain, rashes, and lupus nephritis. It is uncommon to have large pericardial effusions in children with SLE and cardiac tamponade as the initial presentation of SLE is even rarer.

An 11-year-old female presented to our Children Emergency Unit with fever and fast breathing for two weeks, bilateral leg swelling of four days, and cough of two days duration. She was acutely ill, tachypneic, and dyspneic with marked orthopnea, bilateral leg edema, and raised JVP. She was tachycardic with a diffuse apex beat. Chest X-ray showed a large globular heart. 2D-Echocardiography showed a large circumferential pericardial effusion with a dilated non-collapsing IVC and diastolic collapse of the right ventricle. She had a pericardiotomy done and 650mls of serous pericardial fluid was drained. The inner pericardium had a fibrinoid exudate with a "bread-and-butter" appearance. Pericardial fluid cytology showed no malignant cells while pericardial biopsy showed suppurative granulomatous inflammation. Antinuclear antibody (ANA) was strongly positive. The patient was managed with corticosteroids, colchicine, and hydroxychloroquine, and has remained stable on follow-up.

While cardiac tamponade as an initial presenting complaint in SLE is rare, it is important that children presenting with large pericardial effusions and tamponade be evaluated for rheumatologic disorders. This can be crucial to revealing the correct diagnosis and instituting appropriate care.

Keywords: Systemic Lupus Erythematosus, Pericardial Effusion, Cardiac Tamponade

***Correspondence:** Dr Bassey Kevin, Department of Paediatrics, University of Uyo Teaching Hospital, PMB 1136 Uyo, Nigeria
E-mail: edemkevin@gmail.com

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Introduction

Systemic Lupus Erythematosus (SLE) is an autoimmune disorder that presents with manifestations in all the organ systems of the body. This complex disorder is characterized by the proliferation of autoantibodies, and immune dysregulation resulting in damage to body organs.[1] Pediatric SLE accounts for 10-20% of all cases and the disorder typically presents in adolescence and early adulthood although it has been reported in infants and toddlers as well.[2] Worldwide prevalence of pediatric SLE ranges from 3.3-9.7/100,000 with an incidence of 0.3-2.2/100,000 child-years and the disorder is found to be most common among those of African ancestry and females (M:F ratio of 8-9:1).[3]

Pediatric SLE most commonly presents with fever, joint pain, rash, malaise, or prolonged fatigue.[1] Cardiovascular manifestations are known to occur in up to 50% of patients[4] with SLE with pericarditis being the most common presentation.[5] Pericarditis was noted to be the presenting condition in 4-24% of children with SLE compared to 45-77% who present first with lupus nephritis and 20-36% who present with neuropsychiatric manifestations.[5] It is however uncommon to have large pericardial effusions and even cardiac tamponade reported in patients with SLE[4,6-9] and even rare to have massive pericardial effusions/cardiac tamponade as the initial presenting condition for pediatric SLE.

There is a paucity of data regarding cardiovascular manifestations of pediatric SLE in Africa. A 28-year retrospective review from South Africa revealed that pericardial effusions were present in one-quarter of children who had been diagnosed with SLE and about 3% had developed cardiac tamponade.[8]In this case report, we present and discuss an 11-year-old Nigerian child who presented with a massive pericardial effusion as the initial manifestation of pediatric SLE.

Case History

An 11-year-old female was referred to the children's emergency room of our hospital with complaints of high-grade continuous fever and fast breathing of two weeks' duration, bilateral leg swelling of four days, and cough of two days. There was associated difficulty in breathing which occurred both at rest and on exertion as well as orthopnea. Cough was also associated with chest pain and reported to be worse on lying flat. Leg swelling was associated with abdominal swelling but there was no facial puffiness. She was initially taken to a secondary health facility and referred when symptoms worsened.

There was a history of recurrent urticaria-like rash for the last year. There were no routine medications or known drug allergies. She was not exclusively breastfed and had yet to attain menarche. She was a twin, and the first of three children, with no similar illness in twin or other siblings. Her father also had a history of recurrent urticaria-like rashes. Parents belong to the upper socioeconomic class.

At presentation, she was acutely ill with severe respiratory distress and marked orthopnea, febrile (37.8^{o}C), anicteric, acyanosed, and not dehydrated, she had bilateral cervical and submandibular lymphadenopathy and bilateral leg oedema up to both knees. She had an $SPO_2$of 92% and a weight of 32kg (25th percentile for age).

Her pulse rate was 132bpm, BP - 110/60mmHg, and pulsus paradoxus could not be demonstrated due to severe tachypnoea and dyspnea. The jugular venous pulse was 7cm.The apex beat was diffuse along the anterior axillary line. Heart sounds were muffled and distant. She was dyspneic with a respiratory rate of 58cpm. There was dullness to percussion and decreased breath sounds in the left middle and lower lung zones. There was a discoid skin rash on the upper limbs and trunk but no joint swelling.

Chest X-ray revealed a large, globular heart with bilateral pleural effusions. The electrocardiogram showed sinus tachycardia with low voltage complexes. A 2D Echocardiogram revealed a large circumferential pericardial effusion with a dilated non-collapsing inferior vena cava and fibrinous strands within the effusion.

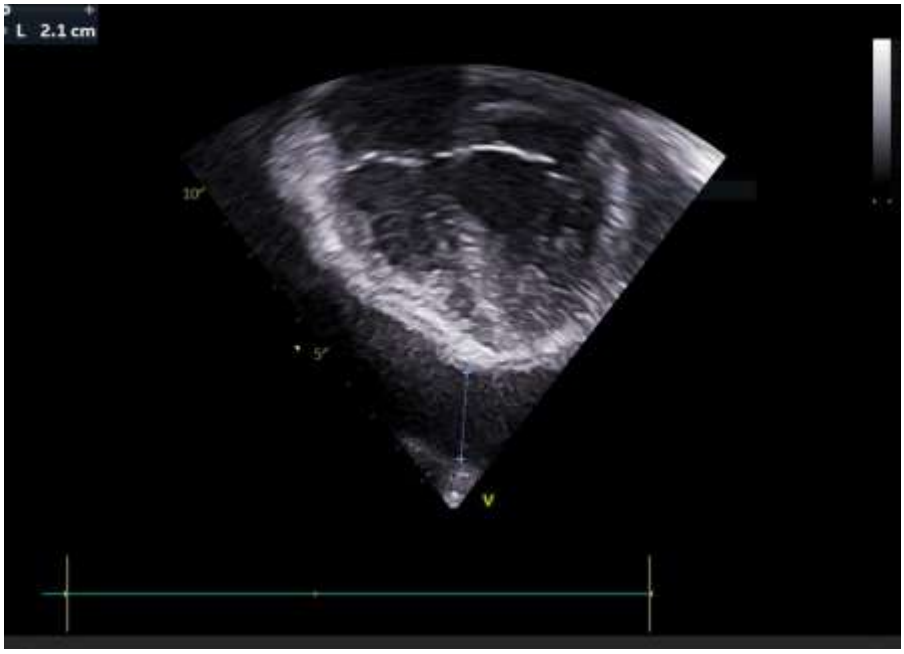

Figure 1: The image shows a large circumferential pericardial effusion. The heart was seen to be "swinging freely in the pericardial fluid."

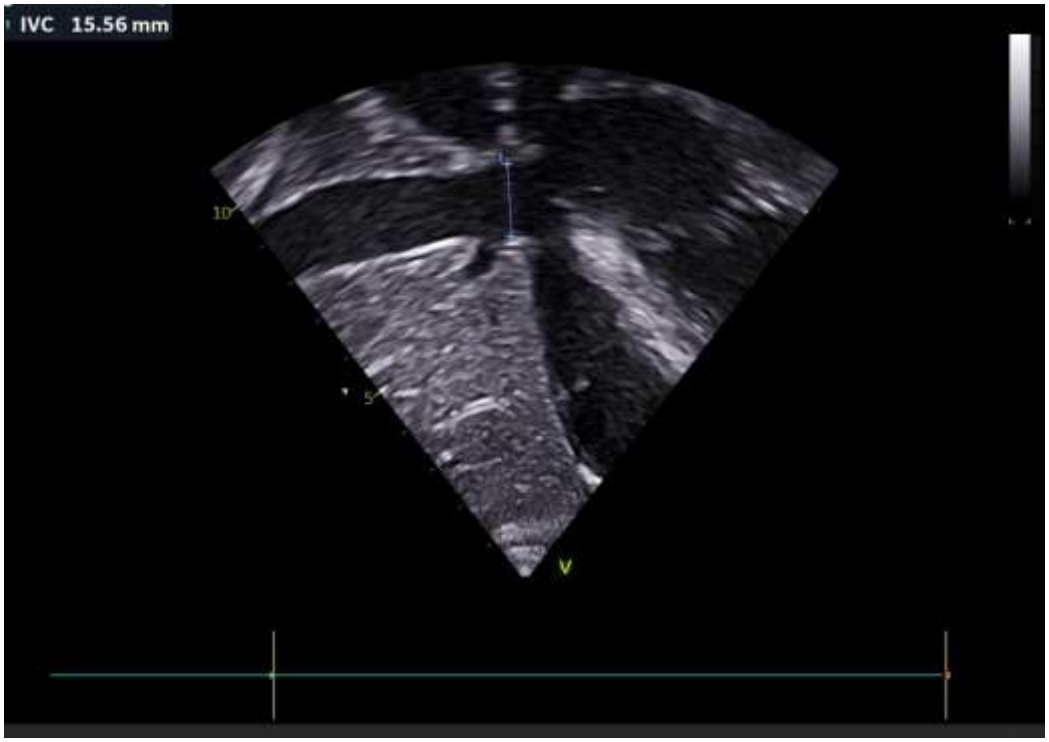

Figure 2: The Inferior Vena Cava is shown and is dilated. It was also seen to be barely collapsible on inspiration.

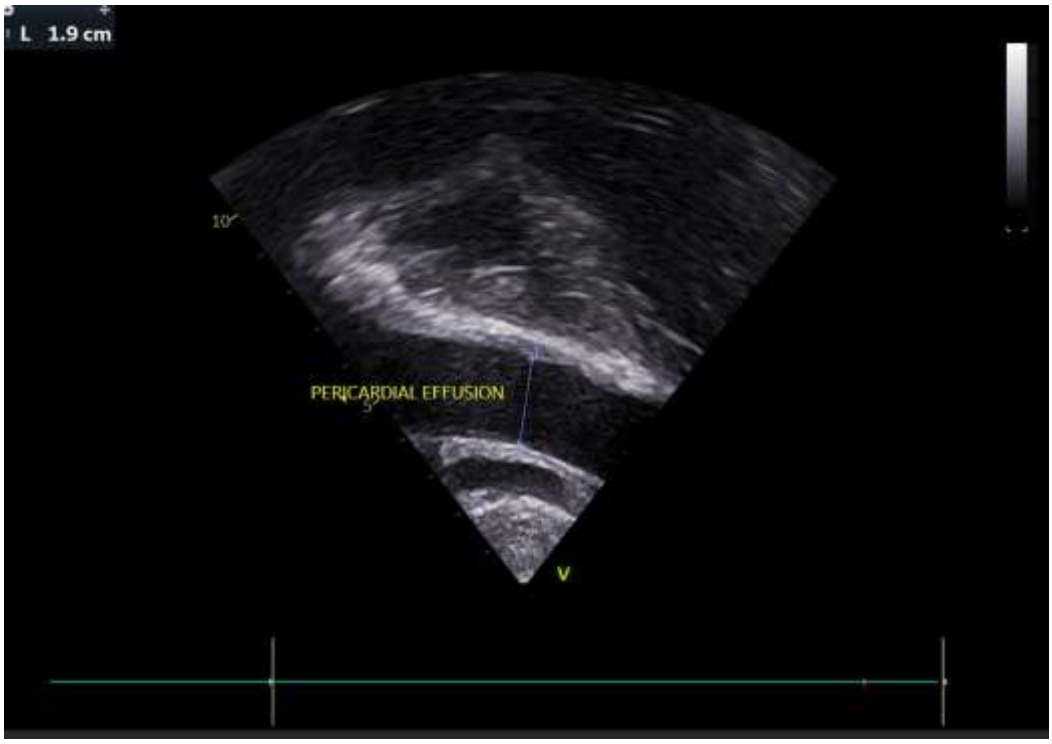

Figure 3: The image shows pericardial effusion particularly related to the right ventricle. The associated diastolic collapse of the right ventricle was seen.

A window pericardiotomy with drainage was done and the findings included: serous pericardial effusion of about 650ml, fibrinoid exudate on the inner side of fibrous pericardium, bread, and butter appearance of the parietal pericardium, left pleural effusion about 200ml of serous fluid, adhesion of left lung to pericardium but no intrapericardial adhesions. A drainage tube was left *in situ.*

Pleural and pericardial fluid acid-fast bacilli and MTB GeneXpert were both negative. Pericardial fluid cytology was negative for malignant cells and pericardial tissue biopsy showed suppurative granulomatous inflammation. The sample was negative on PAS, AFB, and Gram staining. Urinalysis was positive for blood and protein and the estimated 24-hour urine protein was 1000mg. ESR was 130mm/hr and the CRP was elevated (46mg/L). HIV I and II screening was negative and Full Blood Count showed a microcytic anemia (Hb - 7g/dl) with normal white cell count and differential. Direct Coombs test was positive. Her serum tested strongly positive for antinuclear antibodies (titer 1:>5120) with a homogenous pattern. The diagnosis of SLE was established based on the positive clinical and immunological criteria present. The patient satisfied 5 of the 17 Systemic Lupus International Collaborating Clinics criteria[10] namely skin rash, serositis, nephritis, positive serum ANA, and positive Direct Coombs test. Window pericardiostomy and closed thoracostomy drainage were done on admission then the patient had IV methylprednisolone 500mg daily for three days which was followed up by oral prednisolone at 1mg/kg/day daily. Colchicine 0.6mg daily and hydroxychloroquine 250mg daily were also administered. Repeat Echocardiogram showed no significant residual effusion.

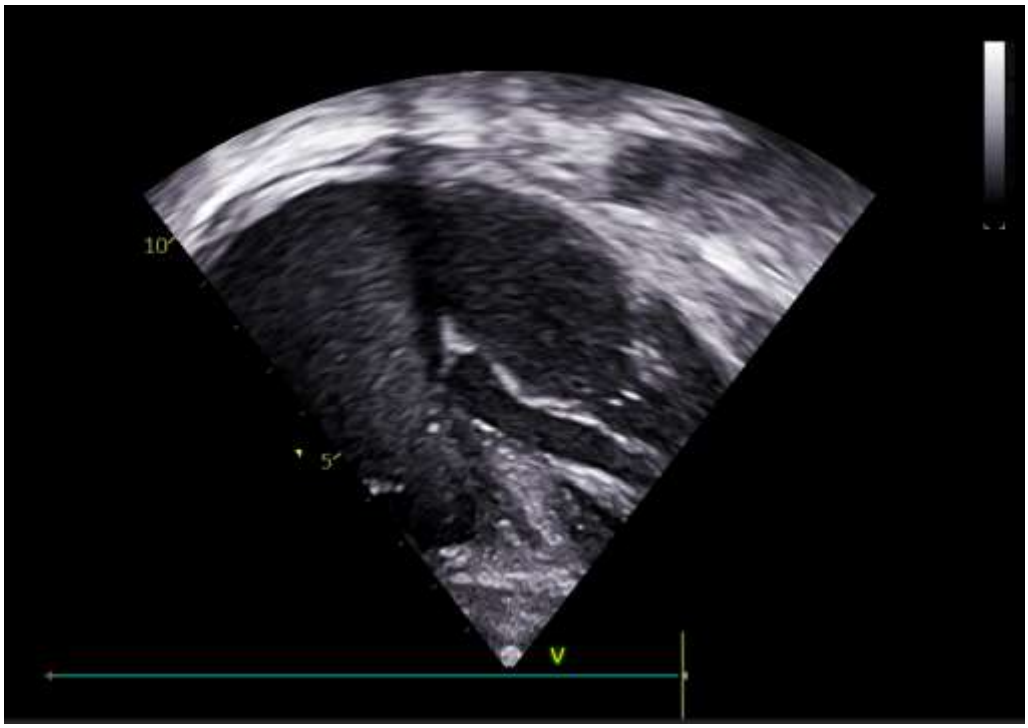

Figure 4: The image shows the apical 4-chamber view post pericardiotomy. Only minimal effusion is noted around the atria.

Due to non-compliance with blood pressure medications, the patient developed seizures associated with markedly elevated blood pressure and was readmitted twice over a three-month period. Blood pressure was controlled using IV labetalol both times and oral methyl-dopa, enalapril and amlodipine were given on follow-up. She has since remained stable on follow-up for the last eight months.

Discussion:

Pericardial effusions are frequent cardiac complications of SLE but massive effusions and/or cardiac tamponade such as was the case in our patient are rarely ever the presenting condition for pediatric SLE. There are less than 20 documented cases of pediatric SLE presenting with tamponade in the literature[5] and only three reported from Africa in the last thirty years.[8] Adult studies have shown a similar picture with a large review from India revealing that about 3% of all adults with SLE over a 30-month period had cardiac tamponade as a presenting feature.[11] Although SLE presenting with large pericardial effusions is rare, it would be pertinent to note the possibility of occurrence and to have a high index of suspicion for pediatric SLE in children presenting with features of pericarditis particularly in resource-poor settings such as Africa where diagnostic capabilities are limited, expensive and may need to be prioritized.

Our patient presented with a history of fever, difficulty in breathing, and cough. The key signs on examination were marked tachycardia, tachypnoea, dyspnea, and orthopnea, an elevated JVP and muffled heart sounds. Studies have shown these to be the key signs of cardiac tamponade and in addition to cardiomegaly on chest radiographs are reportedly more sensitive than other signs, including hypotension (which was absent in our patient), in diagnosing cardiac tamponade.[12] Our patient fulfilled the criteria, with a total score of 13.5, for immediate drainage of pericardial fluid (score of 6 or more) as set forth in the triage strategy by the European Society for Cardiology Working Group.[13] A window pericardiotomy with continuous drainage rather than a pericardiocentesis was however carried out due to the presence of fibrin strands within the effusion which may predict difficulty with pericardiocentesis.[14] The suspicion of an underlying systemic disorder at the time and the presence of a left pleural effusion also necessitated continuous drainage to prevent acute recurrence.

The findings of "bread-and-butter pericarditis" and adherence of the left lung to the pericardium may have suggested rheumatic pericarditis or tuberculosis respectively, both of which are more common causes of pericarditis in the tropics[15-16] and could present a diagnostic conundrum. In the index patient, both the MTB GeneXpert for the pericardial fluid as well as Ziehl-Neelsen staining on the pericardial fluid and biopsy tissue were found to be negative. It may also be that many cases of lupus pericarditis are misdiagnosed as tuberculous pericarditis or even reported as being of unknown etiology due to the lack of diagnostic facilities for SLE as has been suggested by a large systematic review of pericardial diseases in Africa.[15] The presence of both raised CRP and ESR as seen in our patient is significant as the elevation of both parameters is highly indicative of the presence of serositis in SLE.[17] The presence of a microcytic anemia in our patient while not typical for SLE is not unusual[18] and may also be indicative of the high burden of co-morbid nutritional deficiencies in our environment.

Systemic lupus erythematosus had long been considered a rare condition in Sub-Saharan Africa.[19] The incidence is however on the increase and it is believed that this may be due to increased awareness and increasing availability of diagnostic tools rather than an actual increase in cases.[20] Despite this increased availability of diagnostic tools, confirming SLE diagnosis remains a challenge due to the prohibitive costs of extractable nuclear antigen antibody (ENA) panels in our environment. These costs are largely borne directly out-of-pocket by the patients and their families.[21] The index patient was only able to afford to do the ANA test and was unable to afford the other ENA tests more specific for SLE such as anti-dsDNA, anti-Sm, and antiphospholipid antibodies. The diagnosis was however established with the combination of clinical and immunologic criteria according to the SLICC guidelines.[10] The SLICC criteria has shown better sensitivity in the diagnosis of pediatric SLE and has led to earlier diagnosis and fewer misclassifications when compared to the earlier ACR criteria.[22]

Conclusions

Cardiac tamponade as the initial presenting condition of pediatric SLE is rare. It is however known to occur and children presenting with large pericardial effusions and tamponade should have appropriate testing for rheumatological disorders particularly if associated with pleural effusion, hematologic abnormalities, and elevation of both CRP and ESR.

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