Science World Publishing

Open Access Journal | Volume: 2.1

SCIENCE WORLD JOURNAL OF PEDIATRICS

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Cardiac Tamponade Revealing Systemic Lupus Erythematosos in Children: About Two Cases

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Article Information

Article Type: Case Report

Journal Type: Open Access

Volume:2 Issue: 1

Manuscript ID: SWJP-2-1122

Publisher: Science World Publishing

Recieved Date: 26 Feb 2024

Accepted Date: 18 Apr 2024

Published Date: 22 Apr 2024

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Citation:

Aoussar C (2024). Cardiac Tamponade Revealing Systemic Lupus Erythematosos in Children: About Two Cases. Sci World Journ of Pediatrics, 2(1);1-5

1. Abstract

Initial cardiac tamponade revealing systemic lupus erythematosus (SLE) is rare and represents a diagnostic and therapeutic challenge. We report two cases presenting cardiac tamponade as the first manifestation of SLE, echocardiography confirmed cardiac tamponade, and biological assessment revealed markers of autoimmunity. Our two cases highlight the importance of considering SLE as a differential diagnosis in patients presenting with cardiac tamponade, even in the absence of classic symptoms of the disease. Early and appropriate treatment, particularly general corticosteroid therapy, is essential to prevent recurrence of tamponade.

2. Introduction

The occurrence of cardiac tamponade in systemic lupus erythematosus (SLE) is a rare but potentially fatal complication that may require urgent medical intervention. While pericarditis is a common manifestation of SLE, cardiac tamponade initially revealing this autoimmune disease is extremely rare, posing a diagnostic and therapeutic challenge for clinicians. Here, we present two cases in which cardiac tamponade was the initial presenting sign of SLE in children, underscoring the importance of recognizing this atypical presentation for early diagnosis and management [1,2].

3. Observation 1

We present the case of a 12-year-old girl, with no family history of autoimmune diseases, born to healthy, non-consanguineous parents. The disease history spans 5 months, starting with bilateral cervical lymphadenopathy accompanied by anorexia, asthenia, and weight loss. A lymph node biopsy revealed chronic lymphadenitis with suppuration foci, ruling out tuberculosis or malignancy. However, the patient was subsequently lost to follow-up. Subsequent symptoms included abdominal distension, dyspnea, chest pain, lower limb edema, and arthralgia, in a context of apyrexia with a noticeable decline in general health. Clinical examination revealed dyspnea, pallor, fever (38.5°C), asthenia, tachycardia (127 beats/min) with muffled heart sounds, ascites, pleural effusion, hepatosplenomegaly, and lower limb edema. The biological assessment showed anemia at 6.4g/l normochromic normocytic, lymphopenia at 600/mm3, platelets at 284,000/mm3, a C-reactive protein at 49.9mg/l, a sedimentation rate was accelerated to 143mm at the 1st hour, the direct Coombs test was positive, hypo albuminemia at 14.76 g/l, impaired renal function with urea at 1.34g/l and creatinine at 12.5mg/l, leishmaniasis serology was negative and proteinuria from 24h to 2.28g/24h. Chest x-ray (Figure 1) revealed

cardiomegaly and pleural effusion. Echocardiography (Figure 2) revealed a large circumferential pericardial effusion with a very dilated inferior vena cava measuring 14 mm and diastolic obstruction of the right cavities, confirming the diagnosis of cardiac tamponade. Pericardial drainage was performed urgently. Bacteriological examination of the pericardial fluid and the search for Koch's bacillus were negative, as were the cultures. Cytological examination of this fluid did not show any neoplastic cells. The immunological assessment showed the presence of antinuclear antibodies (ANA) at a titer of 1/320, anti-DNA antibodies were positive with hypocomplementemia. Renal biopsy indicated proliferative lupus nephropathy, with the specific class indeterminable due to insufficient glomeruli. Ophthalmological examination was normal. A diagnosis of systemic lupus erythematosus was established. The patient responded well to methylprednisolone boluses followed by oral prednisone (1 mg/kg/day), achieving complete resolution of pericardial effusion during echocardiographic monitoring. Due to persistent renal function deterioration, monthly cyclophosphamide boluses were initiated.

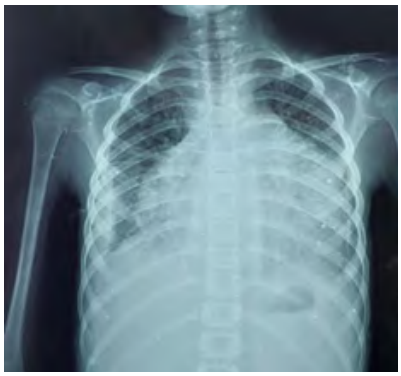

Figure 1: chest x-ray showing cardiomegaly and right pleurisy

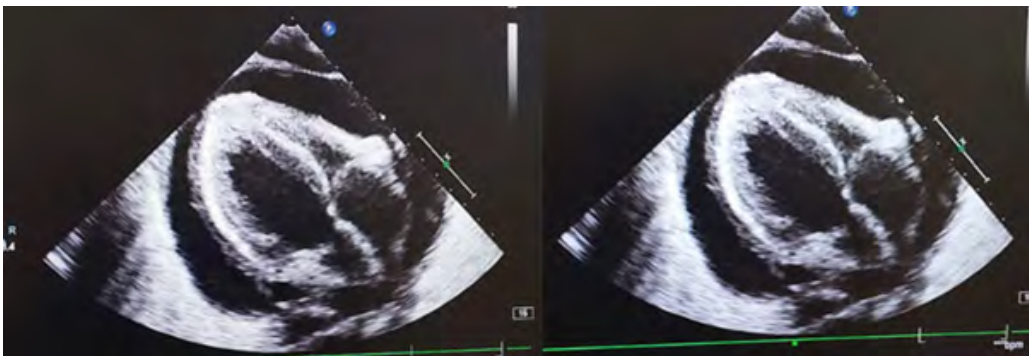

Figure 2: Echocardiography revealed a large circumferential pericardial effusion

4. Observation 2

We present the case of a 15-year-old girl, born to healthy first-degree consanguineous parents with no family history of autoimmune diseases. The disease history spans 1 month, beginning with polyarthralgia and alopecia. Subsequently, she developed abdominal distension, dyspnea, chest pain, and fever, accompanied by a decline in general health. Clinical examination revealed dyspnea, pallor, fever (39°C), asthenia, tachycardia (120 beats/min) with muffled heart sounds, moderate ascites, and pleural effusion syndrome.

The biological assessment showed anemia at 6g/l normochromic normocytic, lymphopenia at 400/mm3, platelets at 256,000/mm3, a C-reactive protein at 85.4mg/l, a sedimentation rate was accelerated to 97 mm at the 1st, the direct coombs test was positivef, ahypoalbuminemia at 29 g/l,ahypocomplementemia,normal renal function with urea at 0.30 g/l and creatinine at 6 mg/l, 24-hour proteinuria at 2.18g/24h. Chest x-ray (Figure 3) revealed cardiomegaly. Echocardiography revealed large pericardial effusion, very dilated inferior vena cava and grade II mitral regurgitation. hasgrade III confirming the diagnosis of cardiac tamponade.Pericardial drainage was performed urgently. Bacteriological examination of the pericardial fluid and the search for Koch's bacillus were negative, as were the cultures. Cytological examination of this fluid did not show any neoplastic cells. The immunological assessment showed the presence of anti-DNA antibodies, anti-nucleosome antibodies and anti-histone antibodies. Renal biopsy revealed class IV lupus nephropathy with an activity index of 6 and a chronicity index of 2. Ophthalmological examination was normal. A diagnosis of systemic lupus erythematosus was made. The patient responded well to methylprednisolone boluses followed by oral prednisone (1 mg/kg/day), achieving complete resolution of the pericardial effusion. Three months later, the patient presented memory disorders. Brain angioMRI revealed moderate diffuse

enlargement of the cortical furrows and demyelinating deep white matter lesions, potentially related to her lupus disease. Due to class IV lupus nephropathy, immunosuppressive therapy was initiated: monthly cyclophosphamide boluses for 6 months followed by oral azathioprine.

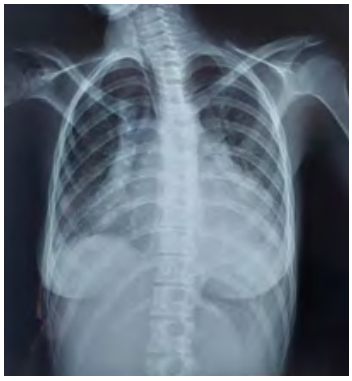

Figure 3: chest x-ray showing cardiomegaly

5. Discussion

Systemic lupus erythematosus (SLE) is an autoimmune disease that can affect various organs. Its incidence in children is estimated between 0.28 and 2.22 per 100,000 children, with a prevalence between 6.3 and 9.73 per 100,000 children. Girls are often affected by this disease. Serous involvement is common in SLE and has been consistently included in the American College of Rheumatology (ACR) lupus classification criteria in 1982 and 1997, as well as in the 2015 Systemic Lupus Collaborating Clinics (SLICC) classification criteria (see Table 1). Pericardial effusion is one of the most common manifestations of SLE, found in approximately 50% of patients. However, cardiac tamponade is rare, estimated to occur in less than 1% of SLE patients. Additionally, cardiac tamponade as the initial manifestation of the disease is even rarer, with only a few cases reported in the literature [3,4,5,7]. In systemic lupus erythematosus (SLE), cardiac tamponade appears to be rare, both as the first manifestation of the disease and during its course. Data on its specific prevalence are limited, but pericarditis, which can eventually lead to cardiac tamponade, is considered one of the earliest cardiac manifestations of SLE. Studies suggest that cardiac tamponade occurs in only a few patients with SLE, indicating that it is an uncommon phenomenon in this autoimmune disease.

Cardiac tamponade is a medical emergency that results in impaired blood flow to the heart as well as hemodynamic problems. Typical symptoms include dyspnea, orthopnea, chest pain, pulsus paradoxus, jugular vein distension, and hypotension. The diagnosis, generally made by echocardiography, reveals a pericardial effusion as well as a diastolic collapse of the right ventricle, a systolic collapse of the right atrium, a plethora of the inferior vena cava with minimal respiratory variation, as well as an exaggerated variation of the respiratory cycle in the entry velocities of the mitral and tricuspid valves.

In recent cases of SLE with cardiac tamponade, it is common to observe pleuritis and/or pleural effusion, while bilateral pleural effusions in this context are rare. Hematologic abnormalities such as hemolytic anemia, thrombocytopenia, and leukopenia are also common. In some patients, low serum complements levels have been observed, which could indicate progression to cardiac tamponade [7,8,9,10]. Our two cases present anemia, lymphopenia, high ESR, hypocomplementemia. Pericardial tamponade was confirmed by echocardiography, and detection of anti-nuclear antibodies (ANA) and anti-DNA antibodies in serum confirmed the diagnosis of Lupus.

For the treatment of pericardial tamponade, it is generally recommended to administer an intravenous bolus of methylprednisolone, with an initial dose of 1 gram over a period of three days, followed by prednisolone at a dose of 1 mg/kg/day, in combination with monthly doses of intravenous cyclophosphamide (750 to 1000 mg/dose), appears to be very effective. Data on immunosuppressive regimens specific to lupus-related tamponade are limited, with only one small study reporting the use of cyclophosphamide. Most studies generally only mention the use of prednisolone, and a few studies have explored the use of colchicine, although its use is not specifically for tamponade. When tamponade is accompanied by hemodynamic complications, urgent cardiological intervention is necessary to assess the situation. This evaluation may include procedures such as pericardiocentesis, which involves draining pericardial fluid to relieve pressure on the heart. Other interventions may include creating a pericardial window to facilitate continued drainage or performing pericardial stripping to remove part of the pericardium [3, 4, 10, 11].

Table 1: The 2015 classification criteria of the Systemic Lupus Collaborating Clinics (SLICC) [8]

Acute/subacute cutaneous lupus rash	Up to 2 points
Malar rash	2.p
Subacute cutaneous Lupus erythematosus (SCLE) rash	1.p
Palpable purpura or urticarial vasculitis	1.p
Photosensitivity	1.p
Discoid lupus erythematosus (DLE) rash or hypertrophic Lupus rash	1.p
Non-scarring frank alopecia	1.p
Oral/nasal ulcers	1.p
Joint disease	1.p
Pleurisy and/or pericarditis	1.p
Psychosis and/or seizure and/or acute confusion	1.p
Kidney involvement	Up to 2 points
proteinuria≥3+ or ≥500 mg/day or urinary casts	1.p
Biopsy-proven nephritis compatible with SLE	2.p
Hematology	Up to 3 points
WBC count < 4000/mm3 or lymphocyte count < 1500/mm3 on ≥ 2 occasions or WBC count < 4000/mm3 along with lymphocyte count < 1500/mm3 in	
one occasion	1.p
Thrombocytopenia < 100,000/mm3	1.p
Hemolytic Anemia	1.p
Serologic tests	Up to 3 points
Low titer positive ANA	1.p
High title FANA with homogenous or rim pattern	2.p
Positive anti-ds DNA	2.p
Positive anti-Sm	2.p
Anti-phospholipid antibodies (aPLs)	1.p
Low serum supplement (C3 and/or C4 and/or CH50)	1.p

6. Conclusion

Diagnosing systemic lupus erythematosus (SLE) during initial cardiac tamponade is complex due to the absence of classic symptoms of the disease. Nevertheless, this diagnosis is crucial to guide appropriate therapeutic management. Although surgical drainage may be necessary urgently to ensure the patient's survival, treatment with systemic corticosteroid therapy is essential to prevent recurrence of tamponade [1, 12].

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