Pericardial effusion as the initial presentation of systemic lupus erythematosus – a case report

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Abstract

Systemic lupus erythematosus (SLE) is an autoimmune disease that can involve nearly any organ system resulting in a great diversity in presentation. Pericardial effusion as the initial form of presentation in SLE is rare. We present the case of a 19-year-old female who presented with complaints of shortness of breath, non-productive cough, visual disturbances, asthenia, arthralgia and fever over the past 2 months. On admission, the patient was febrile, had a heart rate of 145 beats/min, a respiratory rate of 42 breaths/min and the physical examination revealed muffled heart sounds, decreased breath sounds at the left lung base, mouth sores and conjunctival hyperemia. Investigations suggested large cardiac and minimal pleural effusion. After a thorough investigation, the diagnosis of SLE was confirmed using the Systemic Lupus International Collaborating Clinics (SLICC) criteria and the therapy with methylprednisolone was initiated. Our case report aims to emphasize the importance of SLE as a differential diagnosis when presented with pericarditis.

Key words: systemic lupus erythematosus, pericardial effusion, autoimmune disease
INTRODUCTION

Systemic lupus erythematosus (SLE) is a complex, heterogeneous disease characterized in 95% by autoantibody production that can involve nearly any organ system resulting in a great diversity of clinical presentation which can be difficult to diagnose in the emergency department.[1] Pericardial involvement can be the initial form of presentation of systemic lupus erythematosus; however, clinically significant pericarditis occurs in less than 30%. [2]. Cardiac tamponade is a medical emergency that develops when a pericardial effusion reaches a critical amount, limiting cardiac inflow and leading to hemodynamic compromise. We present the case of a pericardial effusion as a first manifestation of systemic lupus erythematosus to a 19-year-old female patient for emphasize the importance of systemic lupus erythematosus as a differential diagnosis when presented with pericarditis in the presence or absence of cardiac tamponade.

CASE PRESENTATION

A 19-year-old female patient presented to the emergency department with complaints of shortness of breath aggravated by supine position, non-productive cough, visual disturbances (diplopia), asthenia, arthralgia (especially of the interphalangeal joints) and fever over the past 2 months. She had no past medical history.

On admission, the patient had a heart rate of 145 bpm, a respiratory rate of 42 breaths/min, blood pressure of 105/70 mmHg and a temperature of 38.4˚C. On physical examination, cardiac auscultation revealed muffled heart sounds and the apex beat diffuse and displaced inferiorly. Respiratory auscultation revealed decreased breath sounds at the left lung base, with no other pathological findings. The rest of the examination only proved mouth sores (Figure 1) and conjunctival hyperemia.

Electrocardiography showed sinus tachycardia at a rate of 142 beats/min with diffuse but non-specific repolarization abnormalities. (Figure 2) The chest radiography displayed a diffuse enlargement of the cardiac silhouette and bilateral pleural effusion. Echocardiographic examination revealed the presence of large amount of pericardial fluid (20 mm – maximal diameter at end-diastole), without signs of cardiac tamponade. (Figure 3). We started to administer Anti-inflammatory therapy (1600 mg Ibuprofen daily- according to the pericarditis guideline) and empiric antibiotics (Ciprofloxacin 400 mg iv plus Ceftriaxone 2g iv) and we continued with the other investigations to find out the cause of the pericardial effusion.

Laboratory investigations showed a normocytic moderate anaemia (Hgb 7, 6 g/dl) with a normal white and platelet cell count. Erythrocyte sedimentation rate was 83mm/h, C-reactive protein was > 30 mg/L. Cardiac biomarkers (cTnl 0,038 ng/mL, CK-MB 0,372 ng/mL, NTproBNP 310 pg/mL), renal, liver and thyroid function testing were normal and blood cultures were negative. Uralysis was within normal range and showed no proteinuria. The immunologic panel testing showed that C3, C4, rheumatoid factor, RPR, SSA, SSB, Sm were within the normal range. However, we found that serum antinuclear antibodies (ANA) was positive and antiDs-DNA by ELISA were 67 (normal values < 40U/mL).

Pleural fluid cytology revealed cells of an inflammatory response, with no microorganisms detected on Gram stain or Ziehl-Neelsen stain. On the other hand, there was no evidence of malignancy.

Despite the double antibiotherapy, the patients condition was stationary, with persistent fever. We suspended the antibiotherapy and we started methylprednisolone, initially with an intravenous load, and subsequently per os at a dosage of 32 mg. The fever, dyspnea responded. Repeated echocardiographic examination and chest radiography showed
a decrease in the amount of pericardial and pleural fluid. The diagnosis of SLE was established based on the positive clinical and immunologic findings. The patient satisfied 5 of the 17 Systemic Lupus International Collaborating Clinics (SLICC) for classifying SLE, namely serositis, mouth sores, anaemia, positive serum ANA and positive anti-dsDNA antibodies. The patient was discharged by the 20th hospital day, on a therapeutic regimen of 32 mg/day of methylprednisolone.

**DISCUSSIONS**

Even though pericarditis in systemic lupus erythematosus has already been described in regard to its clinical and echocardiographic features, occurrence of cardiac tamponade as a first manifestation in systemic lupus erythematosus remains a rare onset of this disease. [3]

Studies showed that the most common initial presentations of systemic lupus erythematosus included constitutional complaints such as malaise, fatigue, fever as well as renal disease, musculoskeletal and cutaneous involvement. [4, 5]

Fever in systemic lupus erythematosus remains a challenging clinical problem. About 42% of patients with systemic lupus erythematosus accuse fever as a manifestation of active lupus.[4] In our case, we tried to make all investigations necessary to exclude other causes of fever such as infections, medications or malignancies. Studies showed that lupus is a cause of fever of unknown origin in less than 5% of patients. [5]

In conclusion, this is an interesting case of an atypical presentation in systemic lupus erythematosus that can often lead to major diagnostic delay. By allowing for greater weighting of immunologic criteria the use of Systemic Lupus International Collaborating...
Clinics (SLICC) criteria may be more sensitive for diagnosis, potentially leading earlier diagnostic and treatment.

REFERENCES