Myopericarditis and severe myocardial dysfunction as the initial manifestation of systemic lupus erythematosus

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Abstract

Pericarditis is the most frequent cardiac manifestation of systemic lupus erythematosus (SLE). However, a large pericardial effusion as the initial manifestation of the disease is infrequent, especially when it is associated with myocardial damage. We describe an unusual case of a young female with pleuropericarditis and severe myocardial dysfunction as the initial manifestation of SLE.

Keywords

Pericarditis, myocarditis, pleural effusion, systemic lupus erythematosus, clinical onset

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Introduction

Pericarditis is the most commonly recognized cardiac complication in systemic lupus erythematosus (SLE).¹ Pericardial involvement has been demonstrated in more than 30% of patients with SLE in imaging and necropsy studies.² However, severe pericardial effusion with myocardial dysfunction as the initial manifestation of SLE is uncommon. We describe a case of a 23-year-old woman who presented with myopericarditis and severe myocardial dysfunction secondary to SLE.

Case report

A 23-year-old woman born in the Philippines and living in Barcelona from the past five years, presented to the Emergency Department (ED) with acute onset of chest pain that had started 10 hours before. She denied any other symptoms. Her family history included a brother who died suddenly at 12 years of age. She did not have any other relevant medical history. On admission, her physical examination revealed a pericardial rub without any other abnormalities. Her vital signs were normal. The *electrocardiogram* (ECG) showed a normal sinus rhythm with T-wave inversion in leads V1–3 and isoelectric ST-segment. Laboratory analysis including C-reactive protein (CRP), troponin-I and blood

cell count were normal. She was diagnosed with acute pericarditis and started on intravenous (IV) non-steroidal anti-inflammatory drug (NSAIDs). After a 12-hour period of observation the patient was discharged home, asymptomatic and under oral acetylsalicylic acid treatment.

One week later, the patient returned to the ED with chest pain, dyspnea and palpitations. Her physical examination showed body temperature of 38.3°C, heart rate of 120 bpm and normal blood pressure. Cardiovascular examination was remarkable for tachycardia and pericardial rub, without any signs of congestion or low output. The ECG (Figure 1(A)) revealed sinus tachycardia, negative T-wave in V1–4 leads. An upright portable chest x-ray showed cardiomegaly and a mild left pleural effusion that

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Figure 1. (A) 12-lead electrocardiogram (ECG) showing sinus tachycardia, diffuse ST segment elevation <0.05 mV in leads I, II, III, aVF and V5–V6. Negative T-wave in V3–4 and DIII; (B) Chest X-ray that depicts slightly increased cardiothoracic index (0.6) and mild left pleural effusion; (C) Thoracoabdominal CT-scan revealed a mild left pleural effusion and a severe pericardial effusion; (D) Contrast-enhanced cardiac magnetic resonance (CE-CMR). Left: short-axis view without late delayed enhancement after injection of gadolinium; right: T2-weighted sequence short-axis view without evidence of significant myocardial edema.

was not present during the first episode (Figure 1(B)). Troponin I was slightly elevated (0.035 ng/ml). Portable transthoracic echocardiogram demonstrated a severe pericardial effusion, with biventricular systolic dysfunction secondary to global hypokinesia, without signs of cardiac tamponade. The patient was admitted with a diagnosis of myopericarditis and severe pericardial effusion, and treatment with IV 1.8 g lysine acetylsalicylate, oral colchicine and IV 60 mg methylprednisolone was started. During the following days the patient worsened abruptly, developing acute heart failure and low output state that required dobutamine infusion. A new echocardiogram ruled out cardiac tamponade. To clarify the etiology of the myopericarditis, several tests were performed. Pleural effusion analysis depicted glucose 48 mg/dl, proteins 53.1 g/l, lactate dehydrogenase (LDH) 1163 UI/l, adenosine deaminase (ADA) 30.4 UI/l with mild cellularity and without atypical cells. Polymerase chain reaction (PCR) for detection of *Mycobacterium tuberculosis* in pleural fluid was negative. Blood and pleural effusion cultures were negative. Thoracoabdominal CT-scan (Figure 1(C)) revealed a mild left pleural effusion and a severe pericardial effusion. Cardiac magnetic resonance imaging (MRI) showed pericardial effusion and a low ejection fraction of both

ventricles without the presence of delayed enhancement (Figure 1(D)). Microbiological tests were also taken to investigate a possible infectious etiology. Mantoux skin test and viral serology were negative. A sample of the pleural exudate was cultured in Lowenstein-Jensen medium, being negative. However, the QuantiFERON-TB® blood test was positive. Despite the fact that Mantoux skin test was negative, this finding indicated at least a latent tuberculosis (TB) infection. Thus, treatment with levofloxacin, rifampicin and ethambutol was initiated.

Concomitantly, laboratory examination for immunological biomarkers included a positive titer for antinuclear antibodies (ANA), Ro, Sm and U1 ribonucleoprotein antibodies (RNP). Double-stranded DNA and complement antibodies were negative. With these results, and considering the clinical course of the disease, SLE was considered as the main diagnosis. At this point, given that the patient was already under treatment with tuberculostatics, there was no added risk in initiating immunosuppression; therefore treatment with an IV pulse of cyclophosphamide was started, maintaining high-dose IV methylprednisolone.

Over the following days, the patient's clinical status improved allowing the withdrawal of dobutamine. Repeated transthoracic echocardiogram revealed an improvement in

the ejection fraction of both ventricles with a left ventricle ejection fraction of 53%. The patient was discharged home with oral corticosteroids and acetylsalicylic acid.

Discussion

SLE is a multisystemic autoimmune disease with remarkable variability in clinical presentation. Cardiac involvement is the second most frequent manifestation behind kidney involvement, being diagnosed in nearly 50% of patients. Pericarditis is the most prevalent cardiac manifestation, appearing in 30–50% of patients during the course of the disease. However, pericardial involvement with severe myocardial dysfunction is uncommon.^{3–6}

Initial differential diagnosis included infectious and autoimmune diseases. As a consequence of the clinical suspicion of *Mycobacterium* infection and the requirement of corticosteroid treatment for symptom control, the positive result of Interferon gamma release assay (IGRA) forced us to start treatment for TB. IGRA test has a greater sensitivity than the skin test to detect latent TB,⁷ however active TB infection was reasonably ruled-out with a low ADA⁸ in pericardial fluid, a negative PCR and a negative Lowenstein-Jensen culture, and the patient was diagnosed with latent TB infection.

Clinical and immunological testing led us to the final diagnosis of SLE. Four (abnormal antinuclear antibodies, immunological disorder, arthritis and serositis) criteria for classification of SLE according to the American College of Rheumatology were met.⁹

Other diagnoses investigated, though much less probable, were lymphoma and a pleural tumor. A lymphoma could cause a disseminated disease with similar symptoms; however, the laboratory tests and the absence of adenopathies in the thoracoabdominal CT-scan made this possibility improbable. Pleural malignancies could have explained the exudate; however, this was ruled out because of the absence of cell atypias and the concomitant presence of pericardial effusion.

Contrast-enhanced cardiac magnetic resonance (CE-CMR) is useful in patients with cardiomyopathies, depicting several patterns highly specific of myocardial damage.¹⁰ Although the patient presented clinically with severe left ventricle

systolic dysfunction, CR-CMR did not reveal delayed enhancement at the left ventricle. More experience is needed to correlate the CE-CMR results with anatomopathological findings.

In conclusion, an organized diagnostic algorithm rather than a unique test result led us to proper diagnosis and management of a critically ill patient with a very unusual debut of SLE.

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Conflict of interest

None declared.

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