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Pericardial Effusion Associated with Left Ventricle Hypertrophy and Macrophage Activation Syndrome Revealing Systemic Lupus Erythematosus: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Introduction: Systemic lupus erythematosus is an autoimmune disease of unknown etiology. Despite the rarity of clinical manifestations, cardiac involvement is one of the major causes of mortality.

Case Presentation: We report the case of a patient with severe pericardial effusion and concentric left ventricle hypertrophy (LVH) in whom lupus was manifested by macrophage activation syndrome. she was admitted to the emergence department for fever, chest pain and progressive dyspnea. She also reports having developed inflammatory polyarthralgia for 1 year. Cardiomegaly was noted on chest radiograph. The ECG showed microvoltage and sinus tachycardia. The

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echocardiography showed a severe pericardial effusion with diastolic collapse of the right ventricle and respiratory variations. Faced with this brutal scenario (pre- tamponade), the patient underwent pericardiocentesis with extraction of 900cc of citrine-yellow exudative fluid. A macrophage activation syndrome test was carried out coming back positive. The diagnosis of systemic lupus erythematosus complicated by macrophage activation syndrome was retained (according to the ACR/EULAR 2019 criteria). The patient was treated by corticotherapy. The evolution was favorable with disappearance of the pericardial effusion. This therapy prevents recurrence of symptoms.

Discussion: Cardiovascular manifestations of systemic lupus erythematosus may involve all heart structure specially pericardium, valve, conduction system and coronary arteries. Pericardial involvement is the first to occur in 11% to 54% of cases according to some studies. Patients with systemic lupus erythematosus have an increased prevalence of left ventricle hypertrophy. The studies suggest that inflammation-mediated arterial stiffening is likely to be the underlying mechanism of left ventricle hypertrophy in systemic lupus erythematosus. Thus, the occurrence of macrophage activation syndrome at the same time as lupus appears to be a rare but seems to define a severe form of systemic lupus erythematosus. Early therapy with high-dose intravenous corticosteroids is usually used in macrophage activation syndrome of autoimmune origin.

Conclusions: Systemic lupus erythematosus revealed by an activation syndrome as well as increased LV mass present two progressive indicators of cardiac morbidity and mortality requiring targeted and early treatment.

Keywords: Pericardial effusion; left ventricle hypertrophy; macrophage activation syndrome; systemic lupus erythematosus.

1. INTRODUCTION

Systemic lupus erythematosus is an autoimmune disease of unknown etiology. Despite the rarity of clinical manifestations, cardiac involvement is one of the major causes of mortality. We report the case of a patient with severe pericardial effusion and concentric left ventricle hypertrophy (LVH) in whom lupus was manifested by macrophage activation syndrome.

2. CASE PRESENTATION

A 22-year-old female patient with no significant past medical history was admitted to the emergence department. She presented fever, chest pain and progressive dyspnea. She also reports having developed inflammatory polyarthralgia for 1 year. Physical examination reveals a pale patient, febrile at 39°C, blood pressure 92/65mmHg, muffled heart sounds, tachycardia 120bpm, polypnea 32c/min, hepatomegaly with splenomegaly and distended jugular veins. Cardiomegaly was noted on chest radiography. The ECG showed microvoltage and sinus tachycardia. The echocardiography showed a severe pericardial effusion with diastolic collapse of the right ventricle; respiratory variations were noted (Fig. N.1). Moreover, she had a concentric LVH with a with an LV mass of 126 g/m2, preserved LV systolic function without valvular abnormalities (Fig. N.2).

Faced with this brutal scenario (pre-tamponade), the patient underwent pericardiocentesis with extraction of 900 cc of citrine-yellow exudative fluid. The Adenosine deaminase (ADA) and genexpert tests were negative. Biological tests showed normocytic normochromic anemia 9g/dl, lymphopenia 600/mm3 and thrombocytopenia, proteinuria 3g/d, frank inflammatory syndrome with C-reactive protein a 160mg. A macrophage activation syndrome test was carried out, which came back positive with an H-score indicating a diagnostic probability of between 80 and 88%. A renal biopsy is now scheduled for our patient.

After neoplastic and infectious causes of macrophage activation syndrome were excluded, immunological tests showed positive antinuclear antibodies. Other positive tests included antinucleosomal antibodies, antiribosomal antibodies and native anti-DNA antibodies (Elisa and IFI). The diagnosis of systemic lupus erythematosus complicated by macrophage activation syndrome was retained (according to the ACR/EULAR 2019 criteria). The patient received 3 boluses of methylprednisolone 1g/d for 4 days, followed by prednisolone 1mg/kg/d. The evolution was favorable with a normalization of the sediment in the urine and the disappearance of the pericardial effusion. This therapy prevents recurrence of symptoms.

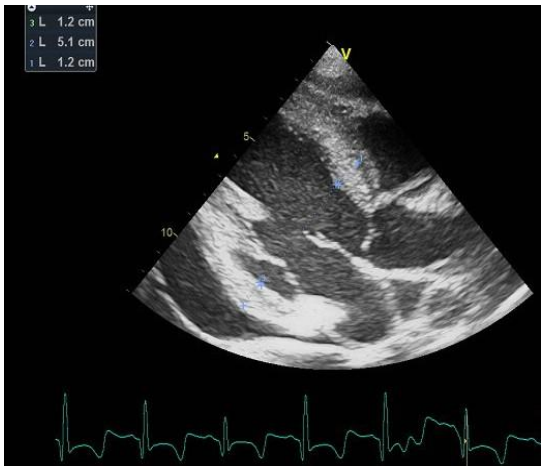

Fig. N.1

Fig. N.1. Subcutaneous view on TTE showed large pericardial effusion:

- **31mm towards RA**
- **20mm towards RV**
- **15mm towards LV**

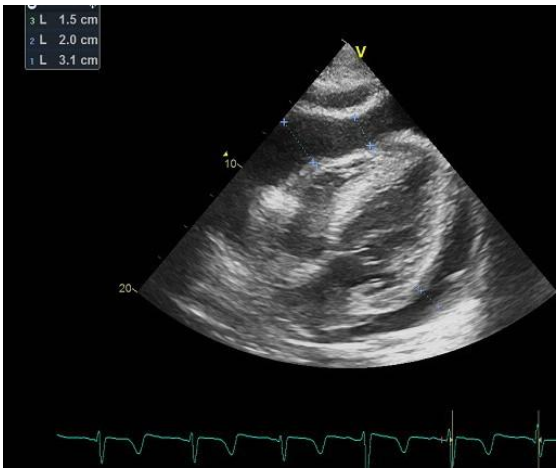

Fig. N.2

Fig. N.2. PSGA view of posterior wall and septal wall with 12 mm hypertrophy., concentric LVH (ERP= 0.42) with an LV mass of 126 g/m2

3. DISCUSSION

Systemic lupus erythematosus is a multisystem connective tissue disease that can have involvement in any organ of the body. Cardiovascular manifestations can affect the pericardium, myocardium, endocardium, valves, conduction system and coronary arteries [1]. Pericardial involvement is the first manifestation in 11% to 54% of cases according to some studies [2]. Patients with systemic lupus erythematosus have an increased prevalence of left ventricle hypertrophy. This is not exclusively the result of concomitant coronary or valvular disease, premature subclinical atherosclerosis or other traditional stimuli, as in our patient [3,4]. The results of studies suggest that inflammation-mediated arterial stiffening is likely to be the underlying mechanism of left ventricle hypertrophy in systemic lupus erythematosus E, with an increased risk of stroke, coronary heart disease, congestive heart failure and sudden cardiac death in various populations [5,6]. It is therefore likely to be a progressive predictor of cardiac morbidity and mortality in patients with systemic lupus erythematosus.

Thus, the occurrence of macrophage activation syndrome at the same time as lupus appears to be a rare presentation described by Wong et al [7]. This association seems to define a severe form of Systemic lupus erythematosus with a risk of relapse and frequent lupus flares that are difficult to control with prolonged immunosuppressive therapy [8]. The studies suggest that more aggressive, early therapy with high-dose intravenous corticosteroids is usually used in macrophage activation syndrome of autoimmune origin. Thus, early targeted therapy may be needed to control the inflammation-mediated effects on vascular stiffness leading to left ventricle hypertrophy [9].

4. CONCLUSIONS

Systemic lupus erythematosus manifested by an activation syndrome and increased LV mass are two progressive indicators of cardiac morbidity and mortality that require targeted and early treatment.

CONSENT

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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