Case study

PERICARDIAL EFFUSION ASSOCIATED WITH LEFT VENTRICULAR HYPERTROPHY AND MACROPHAGE ACTIVATION SYNDROME REVEALING SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE REPORT

Abstract

Introduction: SLE is an autoimmune disease of unlabeled etiology. Cardiac involvement represents one of the greatest causes of mortality. We report a case of lupus revealed by macrophage activation syndrome (MAS) in a patient with abundant pericardial effusion and concentric LVH.
Observation: 22-year-old female patient, without any particular medical history, admitted for fever, chest pain and progressively worsening dyspnoea. Furthermore, she reports the notion of inflammatory polyarthralgia evolving for 1 year. The chest radiograph revealed cardiomegaly. The electrocardiogram showed a micro voltage. Echocardiography revealed abundant pericardial effusion with diastolic collapse of the right ventricle; reparatory variations was noted (figure N.1), concentric LVH. We underwent a Pericardiocentesis with extraction of 9OOcc of citrine yellow fluid. A MAS test was carried out coming back positive. The diagnosis of systemic lupus erythematosus (SLE) complicated by macrophage activation syndrome (MAS) . The patient was treated corticotherapy. The evolution was favorable with normalization of the urinary sediment and disappearance of the pericardial effusion.
Discussion: Cardiovascular manifestations of SEL may involve the pericardium, myocardium, endocardium, valve, conduction system and coronary arteries [1]. Pericardial involvement is the first to occur in 11% to 54% of cases according to electrocardiographic studies [2]. also increased prevalence of left ventricular hypertrophy that is not exclusively the result of concomitant coronary artery disease or valvular disease in our patient [3-4].The results suggest that inflammation-mediated arterial stiffening is likely to be the underlying mechanism of left ventricular hypertrophy in SLE .Thus, the appearance of MAS at the same time as lupus seems to be a rare mode of revelation described by Wong et al [7]; this association seems to define a severe form of SLE. The results suggest that more aggressive therapy; and early by intravenous corticosteroid therapy at high doses is usually used in SAM of autoimmune origin; Thus targeted and early therapy may be necessary [9]. Conclusions: LES 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

VENTRICULAR HYPERTROPHY ,Systemic lupus erythematosus ,macrophage activation syndrome

Introduction:

Systemic lupus erythematosus is an autoimmune disease of unlabeled etiology. Cardiac involvement represents one of the greatest causes of mortality and despite the rarity of clinical signs. We report a case of lupus revealed by macrophage activation syndrome (MAC) in a patient with abundant pericardial effusion and concentric LVH.

Observation:

22-year-old female patient, without any particular medical history, admitted for fever, chest pain and progressively worsening dyspnoea. Furthermore, she reports the notion of inflammatory polyarthralgia evolving for 1 year. Physical examination reveals a pale patient, febrile at 39°C, blood pressure was 92/65 mm Hg, muffled heart sounds, tachycardia at 120 beats/min, polypnea at 32 c/min, hepatomegaly with splenomegaly and distended jugular veins. The chest radiograph revealed cardiomegaly. The electrocardiogram showed a micro voltage and a sinus tachycardia. Echocardiography revealed abundant pericardial effusion with diastolic collapse of the right ventricle; reparatory variations was noted (figure N.1), concentric LVH with a LV mass of 126g/m2 with preserved LV systolic function without valvular abnormality (figure 2).

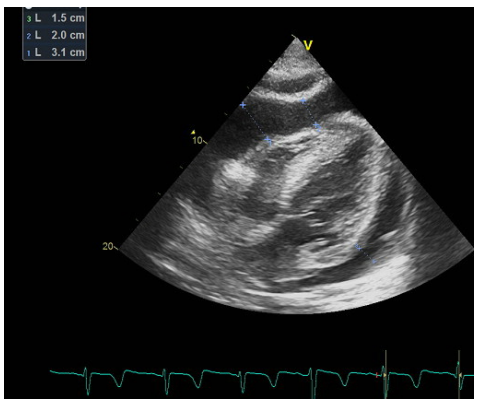

Figure N.1

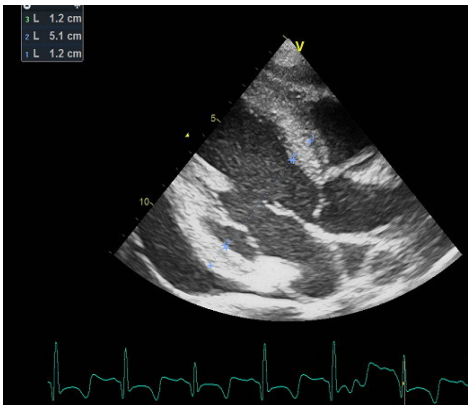

Figure N.2

- Figure N.1: ubcutaneous view in TTE showed a large pericardial effusion: 31 mm looking at RA; 20 mm lookingat RV; 15 mm lookingat LV.
- Figure N.2: PSGA sectiona concentric LVH with a LV Mass at 126g /m2.

In view of this brutal scenario (pre-tamponade) the patient underwent a Pericardiocentesis with extraction of 9OOcc of citrine yellow fluid exudative with negative ADA and genexpert test. The biological tests showed normocytic normochromic anemia at 9

g/dl, lymphopenia at 600/mm3, and thrombocytopenia, proteinuria at 3 g/d, C-reactive protein at 160mg; in front of the frank inflammatory syndrome, a MAS test was carried out coming back positive with an H score indicating a diagnostic probability of 80 to 88%. Our patient is now programmed for renal biopsy.

After ruling out neoplastic and infectious causes of MAS, the immunological tests showed positive antinuclear antibodies and the positivity of others like antinucleosome antibodies, antiribosome antibodies, and native anti-DNA antibodies (Elisa and IFI). The diagnosis of systemic lupus erythematosus (SLE) complicated by macrophage activation syndrome (MAS) was retained (according to ACR/EULAR 2019 criteria).

The patient was treated with 3 boluses of methylprednisolone 1 g/d for 4 days, followed by 1 mg/kg/d of prednisolone equivalent. The evolution was favorable with normalization of the urinary sediment and disappearance of the pericardial effusion.

Discussion:

Systemic lupus erythematosus (SLE) is a multi-systemic connective tissue disease that can affect all organs of the body. Cardiovascular manifestations may involve the pericardium, myocardium, endocardium, valve, conduction system and coronary arteries [1]. Pericardial involvement is the first to occur in 11% to 54% of cases according to electrocardiographic studies [2]. Patients with SLE have an increased prevalence of left ventricular hypertrophy that is not exclusively the result of concomitant coronary artery disease or valvular disease, premature subclinical atherosclerosis, or other traditional stimuli, as in our patient [3-4].

The results suggest that inflammation-mediated arterial stiffening is likely to be the underlying mechanism of left ventricular hypertrophy in SLE with an increased risk of stroke, coronary heart disease, congestive heart failure, and sudden cardiac death in various populations [5,6] and is therefore likely to be a progressive predictor of cardiac morbidity and mortality in patients with SLE. Thus, the appearance of MAS at the same time as lupus seems to be a rare mode of revelation described by Wong et al [7]; this association seems to define a severe form of SLE, with a risk of recurrence, and frequent lupus flares that are difficult to control with prolonged immunosuppressive therapy [8] .

The results suggest that more aggressive therapy; and precocious byIntravenous corticosteroid therapy at high doses is usually used inSAM of autoimmune origins; Thus targeted and early therapy may be necessaryto control inflammation-mediated effects on vascular stiffness that leads toleft ventricular hypertrophy [9]

Conclusions :

LES 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.

DECLARATION OF INTEREST:

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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