Pericardial effusion associated with left ventricular hypertrophy and macrophage activation syndrome revealing systemic lupus erythematosus (a case report)

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 ventricular hypertrophy (LVH) in whom lupus was manifested by macrophage activation syndrome (MAS). 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 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 9OOcc of citrine-yellow exudative fluid. A MAS test was carried out coming back positive. The diagnosis of systemic lupus erythematosus (SLE) complicated by macrophage activation syndrome (MAS) 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 SEL 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 echocardiographic studies. Patients with SLE have an increased prevalence of left ventricular hypertrophy. The studies suggest that inflammation-mediated arterial stiffening is likely to be the underlying mechanism of left ventricular hypertrophy in SLE. Thus, the occurrence of MAS at the same time as lupus appears to be a rare but seems to define a severe form of SLE. Early therapy with high-dose intravenous corticosteroids is usually used in MAS of autoimmune origin.

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: Pericardial effusion, left ventricular hypertrophy, macrophage activation syndrome, systemic lupus erythematosus

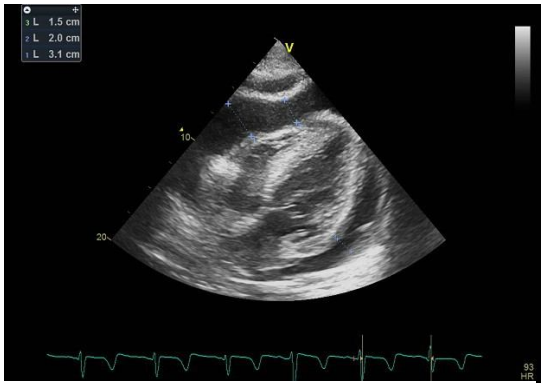
Introduction:

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 ventricular hypertrophy (LVH) in whom lupus was manifested by macrophage activation syndrome (MAS).

Case presentation:

A 22-year-old female patient with no significant past medical history was admitted to the emergence department. She presented with fever, chest pain and progressive dyspnoea. 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 radiograph. 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 (Figure N.1), concentric LVH with an LV mass of 126g/m2 with preserved LV systolic function without valvular abnormalities (Figure 2).



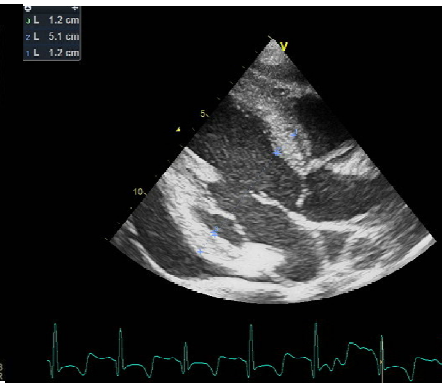


Figure N.2 Figure N.3

• Figure N.2: Subcutaneous view on TTE showed large pericardial effusion:
  - 31mm towards RA
  - 20mm towards RV
  - 15mm towards LV
- Figure N.3: PSGA view showed a concentric LVH with an LV mass of 126 g / m2.

Faced with this brutal scenario (pre-tamponade), the patient underwent pericardiocentesis with extraction of 9OOcc of citrine-yellow exudative fluid. The ADA and genexpert tests were negative. Biological tests showed normocytic normochromic anaemia 9g/dl, lymphopenia 600/mm3 and thrombocytopenia, proteinuria 3g/d, frank inflammatory syndrome with C-reactive protein 160mg. A macrophage activation syndrome (MAS) 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 MAS 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 (SLE) complicated by macrophage activation syndrome (MAS) 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. Therapy prevents recurrence of symptoms.

Discussion:

Systemic lupus erythematosus (SLE) 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 echocardiographic studies [2]. Patients with SLE have an increased prevalence of left ventricular 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 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]. It is therefore likely to be a progressive predictor of cardiac morbidity and mortality in patients with SLE. Thus, the occurrence of MAS 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 SLE with a risk of relapse and frequent lupus flares that are difficult to control with prolonged immunosuppressive therapy [8].

The results suggest that more aggressive, early therapy with high-dose intravenous corticosteroids is usually used in MAS of autoimmune origin. Thus, early targeted therapy may be needed to control the inflammation-mediated effects on vascular stiffness leading to left ventricular hypertrophy [9].

Conclusions:

LES manifested by an activation syndrome and increased LV mass are two progressive indicators of cardiac morbidity and mortality that require 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.

REFERENCES

1 . Tselios K., Urowitz MB Cardiovascular and Pulmonary Manifestations of Systemic Lupus Erythematosus. Revues actuelles de rhumatologie. 2017 ; 13 (3):206–218. doi : 10.2174/1573397113666170704102444

2. Doherty NE, Siegel RJ Cardiovascular manifestations of systemic lupus erythematosus Journal américain du cœur . 1985; 110 (6):1257–1265. doi : 10.1016/0002-8703(85)90023-7.

3. Usalan C., Buyukhatipoglu H., Tiryaki O. Systemic lupus erythematosus complicated by dilated cardiomyopathy and severe heart failure .Rhumatologie Clinique . 2007 ; 26 (1):125–127. doi : 10.1007/s10067-005-0123-z

4. Janice Pieretti, Mary J. Roman, Richard B. Devereux et al. Systemic Lupus Erythematosus Predicts Increased Left Ventricular Mass . Circulation. 2007;116:419–426 . doi.org/10.1161/CIRCULATIONAHA.106.673319

5 Levy D, Garrison RJ, Savage DD, Kannel WB, Castelli WP. Prognostic implications of echocardiographically determined left ventricular mass in the Framingham Heart Study. N Engl J Med. 1990;322:1561–1566. DOI: 10.1056/NEJM199005313222203.

6 . Casale PN, Devereux RB, Milner M, Zullo G, Harshfield GA, Pickering TG, Laragh JH. Value of echocardiographic measurement of left ventricular mass in predicting cardiovascular morbid events in hypertensive men. Ann Intern Med. 1986;105:173–178. DOI: 10.7326/0003-4819-105-2-173

7. Wong K. F., Hui P. K., Chan J. K., Ha S. Y. The acute lupus hemophagocytic syndroine. Ann lntern Med. 1991 ; 1 14:387-90 . DOI: 10.7326/0003-4819-114-5-387

8. Dhote R., Simon J., Papo T., Detournay B., Sailler L., Andre M. H., et al. Reactive hemophagocytic syndrome in adult systeinic disease: report of twenty-six cases aiid literature review. Arthritis Rheuin. 2003;49:633-9 . DOI: 10.1002/art.11368

9. Fukaya S, Yasuda S, Hashimoto T, et al. Clinical features of haemophagocytic syndrome in patients with systemic autoimmune diseases: analysis of 30 cases. Rheumatology (Oxford). 2008;47:1686–91. DOI: 10.1093/rheumatology/ken342