Case report

Cardiac Tamponade, an Unusual First Presentation of Systemic Lupus Erythematosus

Abstract

This case is reported for the rarity of cardiac tamponade as the first manifestation of Systemic lupus erythematosus (SLE). Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease with worldwide prevalence that affects almost all organs of the body.A 27-year-old female presented to our hospital with a two-month history of progressive breathlessness, generalized edematous syndrome and diffuse, non-migratory arthralgias, the patient's shortness of breath was exacerbated with exertion and associated with vague chest tightness.This case is reported for the rarity of cardiac tamponade as the first manifestation of SLE. The transthoracic echocardiography revealed a large circumferential pericardial effusion (PE) with an echo-free space of 34 mm in the apical view and 36 mm in the subcoastal view with swinging heart and diastolic collapse of the right atrium.We highlight the importance of including SLE in the differential diagnosis of patients who present with cardiovascular symptoms because cardiac tamponade and pleural effusion are uncommon early presentations of SLE. For prompt intervention and improved patient outcomes, early detection and diagnosis are essential.

Keywords: Systemic lupus erythematosus, non-migratory arthralgias, multisystem autoimmune disease, conduction system

Introduction:

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease with worldwide prevalence that affects almost all organs of the body (1). The disease is more prevalent among younger women, with a female-to-male prevalence ratio of approximately 10:1(2).Cardiac involvement is common andcan affect the pericardium, myocardium, valvular structures, and the conduction system, pericardial involvement is the most common cardiac manifestation of SLE(3-4).The progression to developing cardiac tamponade is very rare, with an incidence between 1% and 3% (5),and as the first presentation, it is rarer (6),it is a potentially fatal complication if not treated promptly. In this case report, we describe a 27-year-old Moroccan woman whose initial manifestation of SLE was cardiac tamponade.

Case presentation:

A 27-year-old female presented to our hospital with a two-month history of progressive breathlessness, generalized edematous syndrome and diffuse, non-migratory arthralgias, the patient's shortness of breath was exacerbated with exertion and associated with vague chest tightness.Shehad been treated for psoriasis for 8 years. She had no history suggestive of heart failure, renal failure, or liver disease. She had no hair loss, oral ulcers, or rashes. She was not a known patient with diabetes mellitus and/or hypertension. She doesn't drink alcohol or smoke cigarettes.

On examination, the patient's vital signs showed a blood pressure of 90/70 mmHg, a heart rate of 135 beats per minute, a temperature of 36,6°, and oxygen saturation of 98% on room air. The patient was ill-appearing and was in moderate distress. She had facial puffiness, generalized edema.On cardiovascular examination,the heart sounds were muffled, and the jugular veins were distended. A respiratory examination revealed decreased breath sounds at the right lung base.

The electrocardiogram showed sinus tachycardia, low voltages and electrical alternans (fig 1).The chest radiograph showed an increased cardiac silhouette (fig 2)

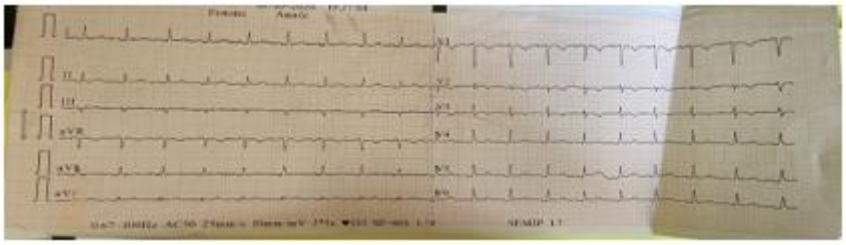


FIGURE 1: EKG demonstrating sinus tachycardia with low voltage QRS and electrical alternans

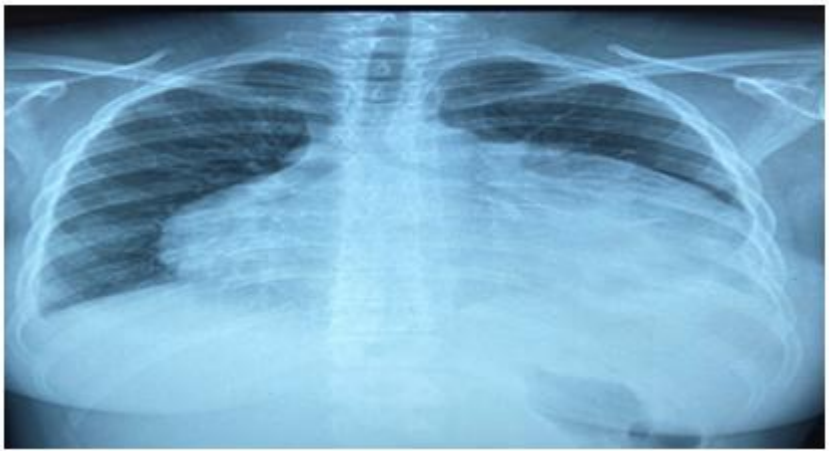


FIGURE 2: Chest X-ray shows an enlarged cardiac silhouette and small right pleural effusion

The transthoracic echocardiography revealed a large circumferential pericardial effusion (PE) with an echo-free space of 34mm in the apical view and 36mm in the subcoastal view withswinging heart anddiastolic collapse of the right atrium(Figure 3). There was also a significant inspiratory decrease in the mitral andthe tricuspid valves E wave velocity on pulse wave Doppler tracing,44% and 61% respectively(fig 4) and an IVC dilated to 26 mm non-complying, which confirmed a tamponade.

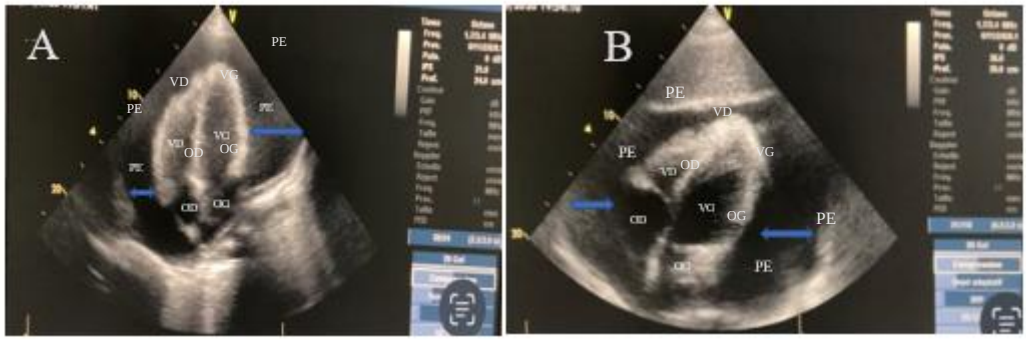


Figure 3: (A) Transthoracic 2-dimensional echocardiogram (apical view) showing a large pericardial effusion (arrow)and of the right atrium. (B) Transthoracic 2-dimensional echocardiogram (subcostal view) showing large pericardial effusion (PE)

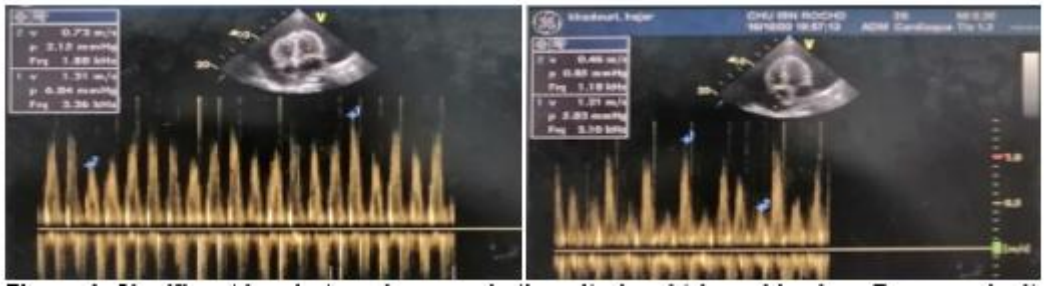


Figure 4: Significant inspiratory decrease in the mitral and tricuspid valves E wave velocity on pulse wave Doppler tracing

The initial emergency behavior was to admit the patient in our intensive care unit, half-seated position, saline infusion andpericardiocentesis withdrawal of 1700 ml of yellow-citrine fluid. We have started symptomatic treatment of the pericardial effusion of colchicine 0.5 mg per day, aspirin 3g per day associated with a proton pump inhibitor. The evolution was marked in our patient by the recurrence of a large posterior effusion, which indicated surgical drainage with a pleuro-pericardial window, the pericardial biopsy showed inflammatory fibrous pericarditis and no malignancy.A biochemical, cytological and bacteriological study was carried out on the punctured fluid, finding an exudative fluid with lymphocytic predominance (90%) and no germs, the culture was sterile and tuberculosis PCR was negative. Complete blood count showed a normochromic normocytic anemia (hemoglobin 10.1 gr/dl), thrombocytopenia at 120000/mm^3 and leukopenia at 1230/mm^3 with an inflammatory syndrome with a CRP of 209 mg/l and hyperfibrinogenemia at 5.95 g/l.Additional laboratory testing was done to further determine the cause of the pericardial effusion. Anti-double stranded DNA (anti-dsDNA) was positive, as were antinuclear antibodies (ANA) at a titer 1280. Complement C3 of 0,45 g/L, and complement C4 <0,06g/l. Positive proteinuria at 0.53 g/24h. Direct Coombs testing was positive. Additional tests revealed an elevation in serum complement levels and cancer antigen 125 (CA-125), but other malignant markers are negative. Serologic tests for hepatitis C virus (HCV), hepatitis B surface antigen (HBsAg), and human immunodeficiency virus (HIV). A CT scan revealed alarge pericardial effusion associated with a small right pleural effusion (fig.5).

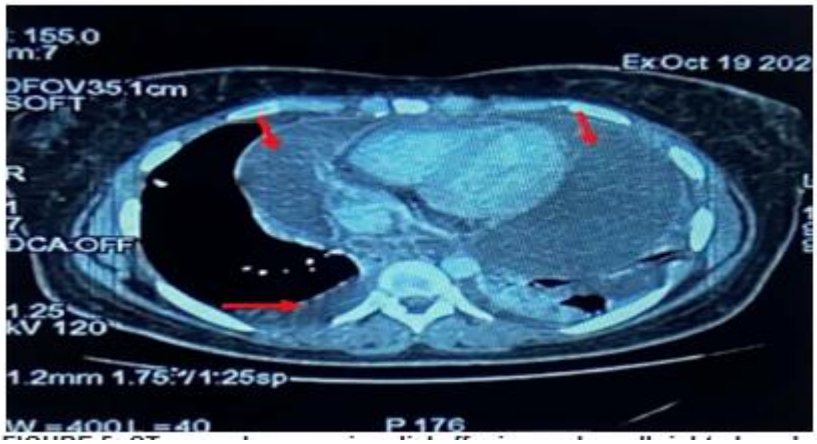
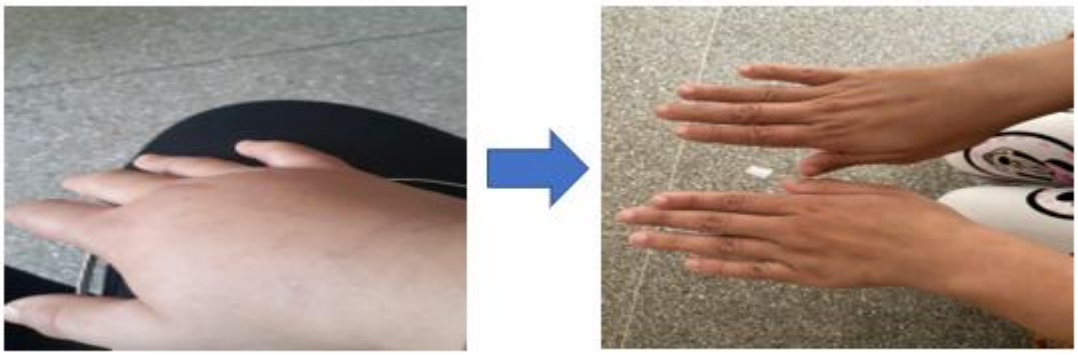


FIGURE 5: CT scan shows pericardial effusion and small right pleural effusion.

The patient had a score of 39 according to the 2019 European Alliance of Associations for Rheumatology/American College of Rheumatology (EULAR/ACR) classification criteria for SLE, confirming the diagnosis of SLE. These included low levels of C4 and C3, affirmativeanti-dsDNA antibodies, proteinuria > 0.5g/24h; tenderness in at least too joint; pericardial effusion and acute pericarditis, thrombocytopenia and autoimmune hemolysis.The patient was started on methylprednisolone at a dose of 1 mg/kg/day. She was also administered hydroxychloroquine orally 200mgx2/day. She had a favorable clinical response with a significant reduction in systemic congestion (fig 6) as well as biological with a CRP at 10 mg/l vs 164 and a 24h proteinuria at 0.16 vs 0.53 g/24h and also regression of the pericardial effusion.She is currently undergoing outpatient follow-up in the rheumatology and cardiology clinics.



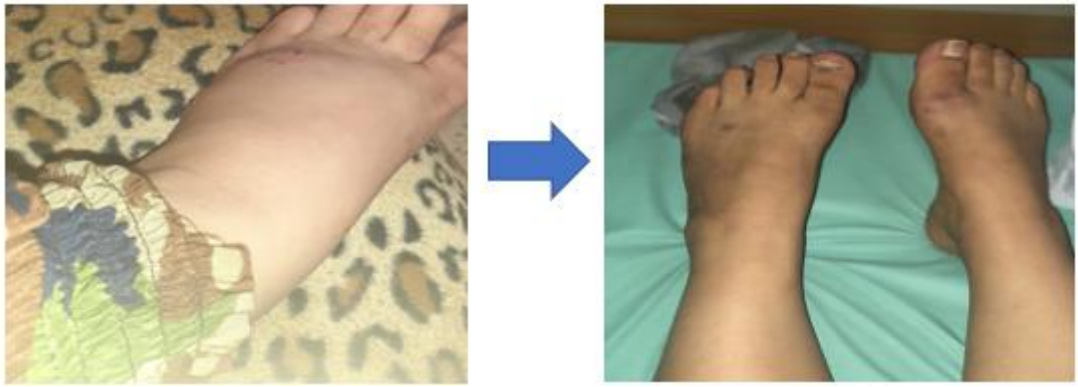


FIGURE 6: Favorable clinical response with a significant reduction in systemic congestion after initiating the treatment

Discussion:

Our patient had severe serositis as an initial presentation of SLE that presented as cardiac tamponade and small pleural effusion.Cardiac tamponade is a pericardial syndrome known as medical emergency (4) characterised by impairment of the diastolic filling of the ventricles causing reduction of cardiac output (7), and if not treated promptly, can lead to cardiac arrest. All the causes of pericardial effusion are also possible causes of cardiac tamponade. In clinical practice the most common etiologies of pericardial effusions are: cancer, tuberculosis and purulent infections, trauma, iatrogenic complication of cardiovascular interventions, acute aortic disease, systemic inflammatory diseases and renal failure (8).The rate and acuity at which these fluids accumulate within the pericardial space are the primary drivers for the development of tamponade physiology(4).The heart is frequently involved in SLE. Approximately half of SLE patients will experience cardiac complications (3). Cardiac tamponade as the first sign of SLE is very rare and only limited to case reports and series (5). In 1949, Curtis and Horne described the first confirmed cases of lupus erythematosus with pericardial effusion as the main manifestation (9).Typical symptoms and signs include dyspnea, orthopnea, chest pain, pulsus paradoxus, jugular venous distension, and hypotension (5). The diagnosis is established by echocardiography, the core echocardiographic findings of pericardial tamponade are pericardial effusion, signs of right atrial and/or right ventricular compression, abnormal respiratory variation in right and left ventricular dimensions and in tricuspid and mitral valve flow velocities usually associated with inferior vena cava plethora (5-8). The low incidence of tamponade despite the high prevalence of pericarditis in SLE may be attributable in part to the widespread use of steroids and nonsteroidal anti-inflammatory drugs (NSAIDs), which effectively reduce pericardial inflammation (5). Several studies have been carried out in an attempt to determine which patients are likely to develop cardiac tamponade. One retrospective study showed that patients with pericardial effusions who developed tamponade had statistically lower C4 complement levels than those who did not develop physiological tamponade (10). Moreover, A retrospective study showed that the presence of pleuritis and anti-nucleosome antibody positivity are significant predictors of progression to cardiac tamponade in SLE patients (11). In treating known cardiac tamponade, efforts to withdraw the pericardial fluid by pericardiocentesis are the treatment of choice (13). The diagnosis of SLE, in our patient, was established based on positive clinical and immunological findings. According to the 2019 classification criteria of the European League Against Rheumatism (EULAR). Generally, the prognosis of lupus serositis is good; it responds well to corticosteroids and immunosuppressants. However, relapse and progression to fibrosis are uncommon (12).We highlight the importance of including SLE in the differential diagnosis of patients who present with cardiovascular symptoms because cardiac tamponade and pleural effusion are uncommon early presentations of SLE. For prompt intervention and improved patient outcomes, early detection and diagnosis are essential.

Conclusion:

SLE affects any organ or system, and its onset ranges from non-specific symptoms to uncommon conditions such as cardiac tamponade, which is a potentially fatal condition and can lead to admission to the emergency department. Clinicians should consider the possibility of SLE when evaluating patients with pericardial effusion. Accurate and rapid diagnosis of this rare form of SLE could save lives. The aim of this case report is to raise awareness of these unusual appearances, to determine the best management approaches and to better understand the mechanisms underlying the different manifestations, hence the need for further research.

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