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REPEATED CARDIAC TAMPONAGE IN SYSTEMIC LUPUS ERYTHEMATOSUS: CASE REPORT

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Abstract: Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease that can present with several clinical manifestations. Cardiac tamponade is a rare condition, but it can be fatal if not caught early. Our case report highlights a 22-year-old female patient diagnosed with recent SLE who presented pericardial effusion followed by recurrent cardiac tamponade. A pericardial-pleural window was performed associated with immunosuppression with corticosteroids and cyclophosphamide, with good clinical response. By presenting this report, we emphasize the importance of diagnostic recognition and early treatment of cardiac tamponade to avoid morbidity and mortality in patients with SLE.
Keywords: Systemic lupus erythematosus, cardiac tamponade, pericardial effusion

INTRODUCTION

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease that can present with several clinical manifestations. As for pathophysiology, it is characterized by a defect in innate immunity, culminating in the breakdown of self-tolerance and, thus, promoting the creation of antibodies by B cells of adaptive immunity together with the synthesis of immunocomplexes that are deposited in tissues (1). The disease in question is more common in women compared to men, at a ratio of 10:1, with one of the possible explanations being the difference in hormonal composition between the sexes (2). Thus, as it affects all body tissues, it can have different presentations, making its diagnosis a challenge (3).

Cardiac tamponade is a rare but potentially fatal complication of systemic lupus erythematosus (SLE), with a prevalence of 4 to 4.5% of patients (3,4,5). However, as it is a rare presentation, the hypothesis is often not considered, leading to late treatment, which contributes to higher mortality (5).

Therefore, knowledge about this possible complication is important for an early diagnosis and adequate treatment. The present study aims to record the case of a patient who was hospitalized with SLE and presented recurrent cardiac tamponade as a complication.

CLINICAL CASE

A 22-year-old woman, born and living in Itapajé-CE, married, housewife, was diagnosed with SLE in 2022, during a medical evaluation at a hospital in the city, had no follow-up, was awaiting referral to a tertiary hospital. Initially, she presented with a skin condition characterized by a malar rash, associated with symmetrical and additive arthritis of the wrists, knees and ankles. At that time, treatment with prednisone 40 mg/day and hydroxychloroquine 400 mg/day was started. Six months after this diagnosis, the patient started with moderate dyspnea progressive on exertion, associated with stabbing chest pain, unrelated to exertion and with no dependent ventilatory pattern, oliguria and frothy urine. In addition, the patient reported alopecia, weight loss and fever. Due to these symptoms, she sought medical assistance, being admitted to an Emergency Care Unit (UPA), and later transferred to a tertiary-level health care hospital, a reference in rheumatology, in the capital of the State of Ceará, Brazil, to follow up.

At the tertiary level hospital, on physical examination, he had a heart rate of 148 bpm, respiratory rate of 38 breaths/min, blood pressure of 91/59 mmHg, temperature of 37.3°C, and oxygen saturation of 96% in air environment. The recorded body mass index was 17.9 kg/m 2 and he was pale. Cardiovascular examination revealed a pancardial murmur 4+/6+ and jugular swelling. The radial pulses were symmetrical but decreased in amplitude

and the extremities were slightly cool. Physical respiratory examination revealed decreased breath sounds at the base of the right lung. Abdominal evaluation showed hepatomegaly and ascites. There was arthritis in the knees and ankles, edema in the lower limbs with pitting 3+/4 and lymph node enlargement in the cervical chains. Laboratory tests showed hemoglobin 6.8 g/dL, positive Coombs test, albumin 2.2 g/dL, creatinine 4.8 mg/dL, urea: 178 mg/dL, urine summary showed proteinuria 3+/4+ and erythrocytes 2+/4+, 24-hour proteinuria demonstrated a proteinuria of 1.8 g/day, Complements (c3: 86 mg/dL and c4: 4 mg/dL), LDH 278 U/L, total bilirubin 0, 9 mg/dL, anti-dna >240 (reagent), HIV and negative serologies for hepatitis B and c (table 1). The findings of the complementary exams suggested acute renal failure due to lupus nephritis and an important autoimmune hemolytic anemia. A simple chest X-ray showed bilateral pleural effusion, greater on the right, associated with a significantly increased cardiothoracic index (Figure 1). The electrocardiogram showed sinus tachycardia. (Figure 2). The transthoracic echocardiogram revealed a massive pericardial effusion with significant diastolic restriction. As a result, an evaluation by the thoracic surgeon was requested and the patient underwent echocardiogram-guided pericardiocentesis, in which 460 ml of serosanguineous fluid were drained (Figure 3). Repeated echocardiography after the procedure showed the presence of a small volume of pericardial fluid without hemodynamic repercussions. The analysis of the pericardial fluid showed 298 leukocytes and 300 red blood cells per mm 3, differential cellularity with a predominance of neutrophils 49% and biochemistry showed glucose discreetly consumed. Both bacteriological and GeneXpert cultures were negative. After 72 hours of the procedure, the patient presented hemodynamic and respiratory worsening, characterized by heart rate of 153 bpm, respiratory rate of 42 breaths/min, blood pressure of 90/57 mmHg, temperature of 37.1°C and maintained oxygen saturation of 91%, and a new urgent transthoracic echocardiography was performed, which showed a new voluminous pericardial effusion, therefore opting for a surgical approach and performing a pericardial-pleural window with pericardial biopsy. In this new procedure, 940 ml of serosanguinous fluid were emitted intraoperatively, and empirical treatment was performed for infectious pericarditis due to contiguity. New assessment of the pericardial fluid showed no difference in relation to the previous analysis. Seven days after the last surgical procedure, the patient presented a tonic-clonic convulsive episode, treated with intravenous diazepam. This intercurrence was attributed to neurolupus, after neuroimaging and CSF analysis, which ruled out the possibility of neuroinfection. We opted for corticosteroid adjustment to a dose equivalent to 1mg/kg/day of prednisone and infusion of human immunoglobulin 400mg/kg/day for 5 days. After this therapy, cyclophosphamide 500mg/m2 of body surface was started, once a month, after clinical improvement of the presumptive pulmonary infectious condition, which was treated with broad-spectrum antibiotic therapy. The patient evolved with a significant improvement in dyspnea, and was discharged with renal replacement therapy and outpatient follow-up in the rheumatology and cardiology services of the tertiary hospital.

DISCUSSION

SLE is a chronic autoimmune disease that affects many organs and systems in the body, including the skin, joints, kidneys, heart and nervous system (1,2). Although cardiac involvement is present in 50% of patients' cases, the most prevalent manifestations are

pericardial effusion and pericarditis, unlike cardiac tamponade, which is extremely rare, occurring in less than 1% of these patients(8). Most cases of cardiac tamponade in SLE are related to lupus syndromes induced by drugs, such as hydralazine, procainamide, isoniazid and carbamazepine (8-10).

The risk to life in cardiac tamponade in patients with SLE represents greater severity, requiring, in some emergency cases, surgical intervention by pericardiocentesis (14). The low incidence of tamponade, despite the high prevalence of pericarditis in SLE, can be attributed in part to the widespread use of steroids and non-steroidal anti-inflammatory drugs (NSAIDs), which effectively reduce the pericardial inflammatory process.

The analysis of the pericardial fluid can often be compromised, both by the professional's technique when performing the pericardiocentesis procedure, and also by the large amount of red blood cells that may be present in the sample, impairing the cellularity analysis, considering that the RBCs may be counted as neutrophils by some programs during laboratory analysis (11). The liquid from the pericardial effusion in lupus patients is commonly an exudate, with the presence of high cellularity and low levels of complement (C4) and, when the presence of antinuclear antibodies in this liquid is confirmed, it can be considered pathognomonic of SLE (13).

Nonsteroidal anti-inflammatory drugs can be used to treat mild pericarditis. However, cardiac tamponade in SLE must be treated with high-dose corticosteroids and pericardiocentesis, except in cases where there is active infection. In these situations, the use of immunoglobulin can be used initially, avoiding immunosuppression in patients with active infection and risk of severe sepsis. Immunosuppressants such as mycophenolate mofetil and azathioprine are also required for treatment of severe serositis (15, 16). In the reported case, findings during the physical examination, such as: tachycardia, low electrical voltage on the ECG, distension of the jugular veins, hypotension and hypophonesis of heart sounds (characterizing Beck's triad), suggested significant cardiac involvement, which required surgical intervention of emergency. In addition, other physical examination and laboratory findings indicated severe systemic activity of the underlying disease.

CONCLUSION

Cardiac tamponade in SLE can be fatal. Attending physicians must be alert and suspect this complication in SLE patients with dyspnea, muffled sounds, hypotension, and jugular swelling. The assertive diagnosis of cardiac tamponade is essential to reduce the mortality associated with this complication. The rarity of repetitive cardiac tamponade in a short period of time is also highlighted in this case report.

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EXAM	OBSERVED VALUE	REFERENCE VALUE
Hemoglobin	6,8 g/dL	11,5 - 15,7 g/dL
Direct coombs	Positivo	Negative
Albumin	2,2 g/dL	3,5 - 5,7 g/dL
Creatinine	4,8 mg/dL	0,6 - 1,3 mg/dL
Urea	178 mg	15 - 43 mg/dL
EAS	Protein 3+/4+ and hemoglobin 2+/4+	Missing protein and missing hemoglobin
24 hours proteinuria	1,8 g/day	Up to 0,14g/dia
LDH	278 U/L	140 - 271 U/L
total bilirubin	0,9 mg/dL	Up to 1,2 mg/dL
HIV	Non-reactive	Non-reactive
Anti- DNA	Reagent (> 240)	Non-reactive: < 20UI/mL Undetermined: 20 to 24.9UI/mL Reagent: > 25 UI/mL
HBsAg	Non-reactive	Non-reactive
ANTI-HBc IgM	Non-reactive	Non-reactive
ANTI-HCV	Non-reactive	Non-reactive
Complement C3	86 mg/dL	90 - 180 mg/dL
Complement C4	4 mg/dl	8 - 150 mg/dL

Table 1

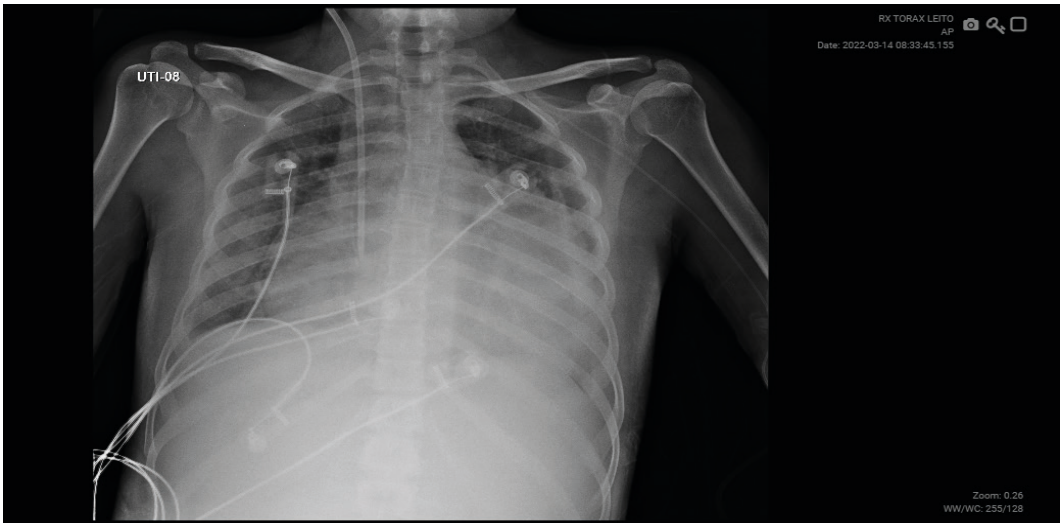

Figure 1

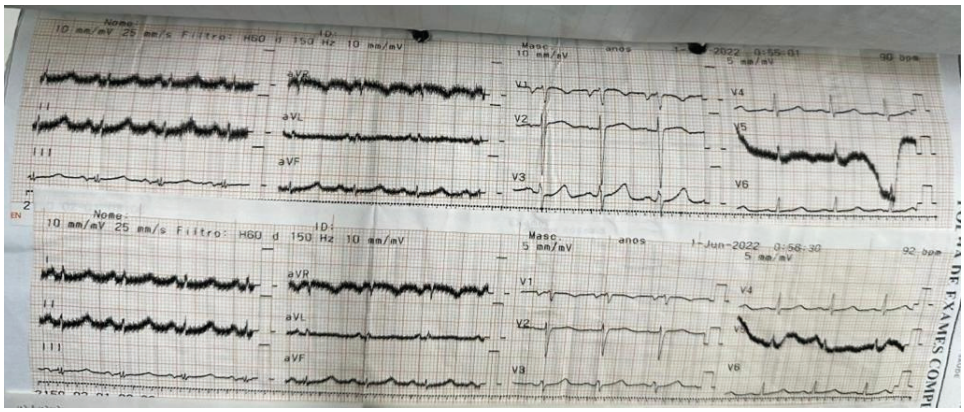

Figure 2

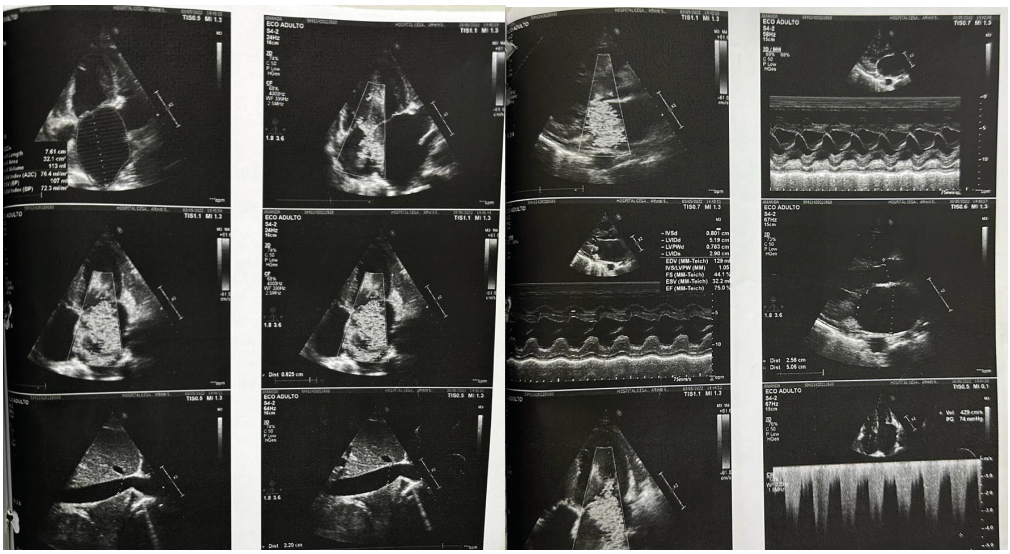

Figure 3