Minimal Effort Dyspnea in a Patient With Systemic Lupus Erythematosus

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Clinical Case

A 48 year-old woman was diagnosed with systemic lupus erythematosus (SLE) 12 years prior due to joint, skin, and eye affection and was treated on occasion with steroids and antimalarial drugs. She was being studied as an outpatient due to microcytic anemia.

She came into the emergency department because she complained of minimal effort dyspnea, nocturnal paroxystical dyspnea, and lower limb edema up to the knees, which had lasted a week. She did not refer thoracic pain, symptoms or signs of a lung infection, fever or any other symptomatology.

On physical examination she had a blood pressure (BP) of 110/65 mm Hg (without pulsus paradoxus), heart rate (HR) of 100 beats/min, rhythmic, with tachypnea but without cyanosis. Jugular venous pressure could be found at the angle of the jaw. She had no adenopathy, her heart sounds were dulled and a few rales were found at both lung bases. Her abdomen was without resistance and depressible, she had soft liver enlargement, without signs of peritoneal irritation. Her lower extremities had edema.

Complementary testing showed leukocytes, 6760/μL; neutrophils, 73.90%; lymphocytes, 15%; hemoglobin, 8.10 g/dL; hematocrit, 23.50%; mean cell volume (MCV), 65.30 fL; mean cell hemoglobin (MCH), 21.4; platelets, 355 000/μL. Serum iron, 34 μg/dL; ferritin, 454 ng/mL; transferrin, 202 mg/dL; erythrocyte sedimentation rate (ESR), 84; C-reactive protein, 17 mg/L. Immunologic study: antinuclear antibodies were positive with a homogeneous pattern. The technique employed was IFA Hep-2; antinuclear antibodies titers were, 1/5120; double-stranded anti-DNA antibodies, 158 (0-15); titers of double stranded anti-DNA antibodies 1/20; anti-SS-A antibodies, 7.66 (0-1.1) arbitrary units (AU); anti-SS-B antibodies, 1.33 (0-1.1) AU; anti-RNP antibodies, 0.98 (0-1.1) AU; anti-Sm antibodies, 0.45 (0-1.1) AU; atypical ANCA P were positive; antimyeloperoxidase antibodies were negative (0-25 AU); antiproteinase-3 antibodies were negative (0-20 AU); rheumatoid factor, 44.6 (0-20) U/mL; immunoglobulin G, 3970 (690-1400) mg/dL; immunoglobulin M, 173 (40-240) mg/dL; immunoglobulin A, 340 (70-370) mg/dL; immunoglobulin E, 28.50 (0-100) U/mL; C3, 83.40 (75-140) mg/dL; C4, 9.30 (10-34) mg/dL; creatinphosphokynase and troponin were normal.

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Figure 1. *Posteroanterior chest x-ray in which global cardiac enlargement due to an increase of the size of the 4 chambers can be observed. There is also a mild bilateral pleural effusion.*

Electrocardiogram: diffuse alteration in repolarization with low voltage. An anteroposterior and lateral chest x-ray showed cardiac enlargement (Figure 1).

Progression

Due to the clinical suspicion of a cardiac tamponade, the patient underwent an echocardiogram (Figure 2) where a severe pericardial effusion with echocardiographic signs of tamponade was documented, after which the patient was subjected to an emergency pericardial fluid evacuation, obtaining 800 mL of serohematic fluid with the following biochemical parameters: glucose, 111 mg/dL; proteins, 6.70 g/dL; lactate dehydrogenase, 1227 U/L; lactate, 6.70 mmol/L; adenosin-deaminase, 57.10 U/L. The cell count was as follows: leukocytes, 7000 cells/µL; polymorphonuclear cells, 90%; lymphocytes, 10%.
After this procedure the patient underwent treatment with steroids and showed both clinical and radiologic improvement (Figure 3). Pericardial fluid cultures, both for anaerobic microorganisms and for mycobacteria, were negative.

Figure 3. *Posteroanterior chest x-ray: heart enlargement and pleural effusion have disappeared.*

Diagnosis

Cardiac tamponade due to lupus pericarditis. Systemic Lupus Erythematosus with a SLEDAI score on 10.

Discussion

Pericarditis is the most common cardiac manifestation in SLE. Symptoms of pericarditis appear in one third of patients, with echocardiographic alterations in 45% and up to 80% of autopsy reports show histologic changes on the pericardium. However, SLE is a very infrequent cause of cardiac tamponade. A recent series[1] of 325 patients with SLE showed that only 4 (1.23%) presented cardiac tamponade. Occasionally, tamponade is the first manifestation of the disease.[2-4]
Pericardial effusion in SLE is normally and exudate[5] with a moderate amount of neutrophils,[6] with high concentrations of antinuclear antibodies.[7] The latter is not pathognomonic because it can occur in tumor-related pericardial effusion.
In patients with lupus, once the diagnosis of cardiac tamponade is established and confirmed through echocardiography, the next step is usually to evacuate the pericardial fluid or, on occasion, perform a pericardial window procedure, alongside treatment with high dose steroids (prednisone or prednisolone at a dose of 60-100 mg/day).[8] Some authors have also postulated the use of immunosupressants if there is a lack of response to steroids. Progression is usually good and the effusion does not tend to recur nor does it leave constrictive sequelae,[9] although there have been some reports of this complication.

Figure 2. *2D vision echocardiography of the 4 chambers; there is a severe concentric pericardial effusion (3 cm on the anterior and posterior sides). The left ventricle is neither dilated nor hypertrophic.*

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