Heart failure with complete recovery in a patient with systemic lupus erythematosus

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Abstract
We report the case of a 43-year-old female patient who was admitted to the Cardiology Department from the Rheumatology Clinic where she was being treated for multi-organ serositis, fatigue and mild dyspnoea on exertion. The patient had a known medical history of systemic lupus erythematosus (SLE). Following extensive evaluation with blood tests for immunological and viral culprits, cardiac ultrasound, chest and abdominal computed tomography (CT) and heart magnetic resonance imaging (MRI), the diagnosis of effusive constrictive pericarditis secondary to her SLE was made. Treatment with β-blockers, diuretics and corticosteroids was given with excellent results, and one year post discharge the patient remains asymptomatic. Systemic lupus erythematosus patients often manifest cardiac complications such as pericarditis. The practising physician should always bear in mind this possibility when treating such patients.

Key words: systemic lupus erythematosus, acute heart failure, heart failure recovery.

Introduction
Cardiac involvement is one of the main complications substantially contributing to the morbidity and mortality of patients suffering from systemic autoimmune diseases. Such involvement has been recognized since the beginning of the 20th century, but in the last decades, newly recognized clinical entities have been described due to the introduction of very sensitive, non-invasive or semi-invasive cardiac imaging techniques [1]. All of the anatomical heart structures can be affected, and multiple pathogenic mechanisms have been reported.

Non-organ-specific autoantibodies have been implicated in immune complex formation and deposition as the initial triggers for inflammatory processes responsible for disease processes such as Libman-Sacks endocarditis, myocarditis and pericarditis. Several autoantibodies, such as anti-phospholipid antibodies (aPL), anti-SSA/Ro antibodies and anti-endothelial cell antibodies, can become potent mediators of cardiac damage. The consequences of such damage can affect several heart structures such as the valves, myocardium, pericardium, conduction tissue and cardiac arteries in patients suffering from systemic lupus erythematosus (SLE), anti-phospholipid syndrome (APS), Sjögren syndrome (SS) or other autoimmune diseases. Other cardiac manifestations, such as pericarditis, myocarditis, endocarditis and conduction disturbances, when present, are often mild and, usually, subclinical features are more prevalent than clinically apparent disease.

We report the case of a 43-year-old female with known history of SLE, who was admitted to our hospital because of mild symptoms of fatigue and dyspnoea and eventually was diagnosed with effusive constrictive pericarditis secondary to her rheumatic primary disease.

Case report
The patient was originally admitted to the Rheumatology Department because of fatigue and known medical history of SLE. Three months prior to admission, ENA,
anti-SSA and anti-SSB antibodies were found to be elevated. ANA were also positive at that time.

She appeared haemodynamically stable with a blood pressure of 120/85 mmHg. The electrocardiogram (ECG) was unremarkable and heart ultrasound revealed normal left ventricular (LV) wall and cavity dimensions with mildly reduced LV systolic function. The estimated ejection fraction was 45%, confirmed independently by two echocardiographers. The mitral valve inflow pattern was indicative of impaired relaxation (E < A, 'E' < 'A'). Right ventricle tissue Doppler imaging (TDI) indices were normal. There was also a small to medium size pericardial effusion, mainly around the right ventricle and adjacent to the anterior wall of the heart. Finally, the inferior vena cava (IVC) was dilated with no respiratory diameter variation. The patient was transferred to the Cardiology Department for in-depth follow-up of her cardiac function.

Blood tests were unremarkable apart from mild microcytic anaemia (haematocrit – Ht 33, haemoglobin – Hb 10.6, mean corpuscular volume – MCV 76) and raised transaminase values (aspartate aminotransferase – AST 175, alanine aminotransferase – ALAT 383) with marginally increased γ-glutamyl transferase (γGT) and lactate dehydrogenase (LDH). C reactive protein (CRP) was also increased, unlike erythrocyte sedimentation rate (ESR), which was within the normal range. Cardiac troponins were modestly elevated with peak CtnI on the day of admission 0.789 (normal < 0.056). The brain natriuretic peptide (BNP) values were also markedly elevated (up to 14 700 pg/ml, normal laboratory values < 100 pg/ml). Serum total protein and albumin were modestly reduced (3.2 g/dl, normal > 3.5). The patient was further investigated with diagnostic thoracocentesis and lab results confirmed that the aspirated pleural fluid was an exudate. A series of extensive blood tests for viral or immunologic markers had already been contacted at the Rheumatology Clinic including Coxsackie virus, cytomegalovirus (CMV), hepatitis B and C, toxoplasma and the Venereal Disease Research Laboratory (VDRL) test. Finally, a series of tests for cancer markers was also conducted without any remarkable findings apart from a marginal initial cancer antigen CA 12-5 value that was not confirmed at a second sampling a few days later. Serum amyloid A (SAA) was 10.5 mg/l (normal value < 6.4 mg/l) and thyroid hormones were also unremarkable.

The patient was investigated with chest and abdominal CT scans. The chest scan revealed a number of lymph nodes of increased dimensions (up to 1.4 cm) and a medium sized pleural effusion, already known from plain chest radiography on admission. No signs of malignancy were evident. The abdominal scan confirmed the presence of intraperitoneal fluid and also revealed a 4 cm cystic mass at the anatomical position of the right ovary, a finding that was considered for further evaluation.

The differential diagnosis at the time included the possibility of myocardium-involving multiple organ serositis secondary to SLE, amyloidosis, or possibly extrathoracic malignancy (e.g. Meigs syndrome) should the right ovary prove to be neoplastic tissue.

Further diagnostic work up was advised and the patient was programmed for a heart MRI and 24-hour ECG rhythm recording. The MRI showed borderline dilation of both ventricles with normal LV systolic function. No signs of acute myocardial inflammation were evident but signal enhancement in the epicardial layer of the lateral/inferolateral wall was possibly consistent with a non-ischaemic inflammatory/infiltrative disease process (myocarditis/cardiomyopathy). The medium sized pericardial effusion was again shown, as was the normal origin of the coronary arteries and mild mitral valve regurgitation without any dilation of the left atrium. The 24-hour ECG recording did not reveal any noteworthy arrhythmias or conduction disturbances. Further evaluation of the ovaries by the gynaecologists refuted the possibility of ovarian malignancy.

The diagnosis of effusive constrictive pericarditis was made and the patient was treated with diuretics, β-blockers and corticosteroids (prednisone 1 mg/kg/day IV). A low dose of angiotensin converting enzyme (ACE) inhibitor was also prescribed. Patient functional status improved significantly over the following days. The symptoms of dyspnoea on exertion and fatigue were abolished. Further, 16 days after admission, the patient was discharged with instructions for close follow-up at the Cardiology Department. At discharge blood tests showed microcytic anaemia of the same level as on admission, but inflammation markers and transaminase values were normal, as were BNP and proBNP. Heart ultrasound prior to discharge confirmed that LV systolic function was normal (estimated ejection fraction – EF 60%). Her next outpatient appointments with the Cardiology Clinic were arranged initially at monthly intervals. At follow-up her cardiac function remained normal with no fluid accumulation up to 1 year post discharge. She remains on β-blocker treatment (carvedilol 6.25 mg bid), prednisolone at low doses (5 mg/day), and hydroxychloroquine in accordance with the rheumatological consultations. There was no necessity of application of diuretics apart from a modest dose of spironolactone (12.5 mg od).

**Discussion**

Cardiovascular disease has recently been acknowledged as a primary cause of morbidity and mortality in SLE [2], and the heart is frequently involved in this systemic disease: very sensitive methods of cardiovas-
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The authors declare no conflict of interest.

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