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CASE REPORT

Systemic sclerosis: A rare cause of heart failure?

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Abstract Systemic sclerosis (SS) is a chronic disease in which there may be multisystem involvement. It is rare (estimated prevalence: 0.5–2/10 000) with high morbidity and mortality, and there is as yet no curative treatment.

We report the case of a young woman newly diagnosed with SS, in whom decompensated heart failure was the main manifestation.

PALAVRAS-CHAVE
Esclerose sistémica;
Cardiomiopatia;
Insuficiência cardíaca

Esclerose sistémica: uma causa incomum de insuficiência cardíaca?

Resumo A esclerose sistémica (ES) é uma doença crónica com possível apresentação multi-sistémica. É considerada uma doença rara (prevalência estimada: 0.5-2/10,000) com alta morbilidade e mortalidade para a qual não há cura hoje em dia.

Relatamos o caso de uma jovem mulher, recém diagnosticada de ES por afetação pleural e cutânea, com insuficiência cardíaca global no momento da consulta.

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Case report

A 38-year-old woman without cardiovascular risk factors, diagnosed with SS one year previously, who had presented with simultaneous involvement of the skin and pleura and was under treatment with steroids and calcium channel blockers, was admitted to the emergency department because of progressive dyspnea at rest accompanied by orthopnea, abdominal distension, and lower limb edema for three weeks.

On physical examination, her blood pressure was 117/81 mmHg and heart rate was 111 beats/min. The presence of microstomia, sclerodactyly, and diffuse cutaneous involvement were noted. Cardiac auscultation was normal without murmur or rub. Breath sounds were absent at the lung bases on both sides. Jugular venous engorgement at 45° was present, with positive hepatojugular reflux. Other physical signs were tender hepatomegaly with bilateral and symmetrical lower limb edema extending up to the knee joint.

The 12-lead electrocardiogram showed sinus tachycardia, incomplete right bundle branch block with left anterior hemiblock and first-degree atrioventricular block (PR interval 210 ms, QRS interval 111 ms).

Baseline arterial blood gas analysis showed hypoxemia as the only abnormality.

Blood tests revealed microcytic anemia (hemoglobin 9.9 g/dl, mean corpuscular volume 69.0 fl) and slight abnormalities in coagulation parameters (INR 1.5) without renal dysfunction.

The chest X-ray showed findings consistent with pleural effusion, mainly at the right base (Figure 1).

Transthoracic echocardiography showed mild left ventricular (LV) dilation (end-diastolic diameter [EDD] 55 mm, end-systolic diameter 49 mm), normal LV thickness, asynchronous LV wall motion, LV ejection fraction 37% by the Teichholz method, LV diastolic dysfunction (restrictive pattern), right ventricular (RV) dilation (EDD 48 mm in 4-chamber apical view) and impaired RV systolic function (tricuspid annular plane systolic excursion 9 mm), without evidence of significant pulmonary hypertension (Figure 2).

During her hospital stay the patient received conventional heart failure therapy, with a favorable response. Pro-BNP levels were found to be high (8191.0 pg/ml).

Cardiac magnetic resonance imaging (MRI) showed a slightly dilated LV with severe LV systolic dysfunction (LV ejection fraction 21%), together with anterolateral subendocardial delayed enhancement, possibly due to fibrosis, and slight RV dilatation (EDD 50 mm) with severe systolic dysfunction (Figure 3).

Diagnostic workup was completed with cardiac catheterization, which showed no significant coronary artery stenosis or signs of significant pulmonary hypertension (peak pulmonary artery systolic pressure 30 mmHg, mean 17 mmHg, and pulmonary capillary wedge pressure 15 mmHg) (Figure 4).

The patient was eventually discharged, clinically stable, with a diagnosis of congestive heart failure and biventricular systolic dysfunction in the context of cardiomyopathy, possibly related to SS. This diagnosis was made on the basis of the absence of other potential causes (coronary disease, hypertension, or family history of heart disease), since a definitive diagnosis based on histological criteria was not available.

Diuretics, angiotensin-converting enzyme (ACE) inhibitors, beta-blockers and angiotensin receptor blockers (ARBs) were added to her routine SS treatment.

Discussion

SS, also called systemic scleroderma, is a chronic disease in which there may be multisystem manifestations, as can be seen in the case presented. There is still debate concerning the role of myocardial fibrosis as a causative mechanism of heart failure. Current hypotheses include arteriolar endothelial injury resulting in fibrosis and vessel obliteration. One theory is that necrotic cardiomyocytes induce recruitment of fibroblasts and their differentiation into myofibroblasts. According to another theory, evolution of the disease with permanent arteriolar wall damage will result in irreversible myocardial ischemia and fibrosis (''coronary Raynaud's phenomenon'').[1–3]

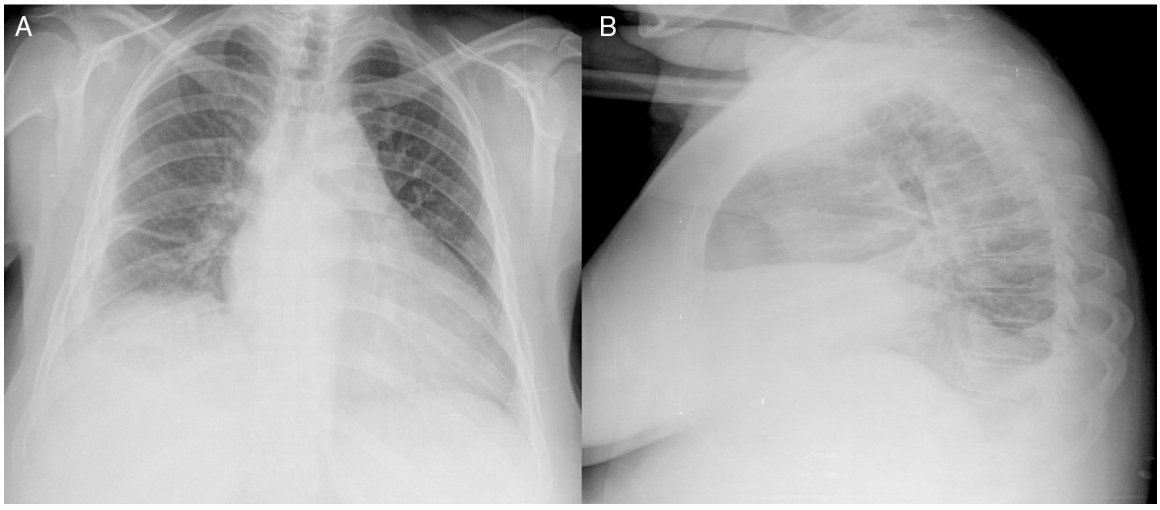

Figure 1 Chest radiograph.