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CASE REPORT

Global Heart Failure and Fatal Outcome in Scleroderma

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ABSTRACT

The prevalence of heart failure with reduced ejection fraction remains low in scleroderma but the evolution can be fatal. We report the case of a 64-years old age women without cardiovascular risk factors, followed for systemic scleroderma for 3 years which presents a global heart failure with systolic left ventricular dysfunction and atrial tachycardia 2/1 associated with pulmonary arterial hypertension, a Raynaud phenomenon and severe renal failure, unfortunately the evolution is marked by sudden death.

KEYWORDS

Scleroderma; Global hear failure; Reduced ejection fraction; Pulmonary arterial hypertension; Sudden death

1. INTRODUCTION

The heart is one of the major organs involved in scleroderma. Cardiac involvement can be manifested by myocardial disease, conduction system abnormalities, arrhythmias, or pericardial disease. Additionally, scleroderma renal crisis and pulmonary hypertension lead to significant cardiac dysfunction secondary to damage in the kidney and lung. Overt cardiac involvement in systemic sclerosis is associated with a mortality rate of up to 70% over five years [1] and about one-fourth of deaths in patients with systemic sclerosis are from cardiac causes [2]. Many patients with systemic sclerosis have cardiac involvement detectable by MRI even though they have no clinical manifestation, heart failure and pulmonary arterial hypertension are complications of scleroderma that affect the life expectancy of patients [1].

2. CASE REPORT

A 64-years old woman of Caucasian origin followed for 3 years for systemic scleroderma, presented to the emergency for tachycardia and dyspnea stage III NYHA, with signs of right heart failure, the clinical examination on admission found: A blood pressure of 90/60 mmHg, the temperature at 37.2°C, an O_2 saturation in air at 90%, edema of both legs up to the knee , Blood biology reveals inflammatory syndrome (leukocytes

10000/mm^3, CRP 37 mg/l), Creatininemia 26 mg/l, serologies: Syphilis, HBS, HCV, and HIV: Negative, serology COVID-19 (IGm-, IGg-), the electrocardiogram shows a supraventricular tachycardia (Figure 1).

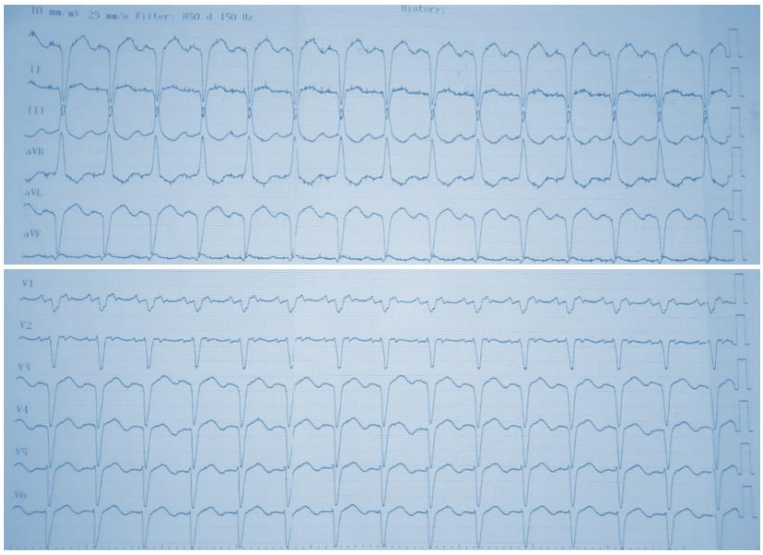

Figure 1: Supraventricular tachycardia.

Transthoracic Doppler echocardiography found LV systolic dysfunction with severe tricuspid insufficiency with probable pulmonary arterial hypertension 45 mmHg, non-elevated LV filling pressures, right atrium surface 20 cm^2, TAPSE 13 mm and inferior vena cava dilated to 23 mm (Figure 2).

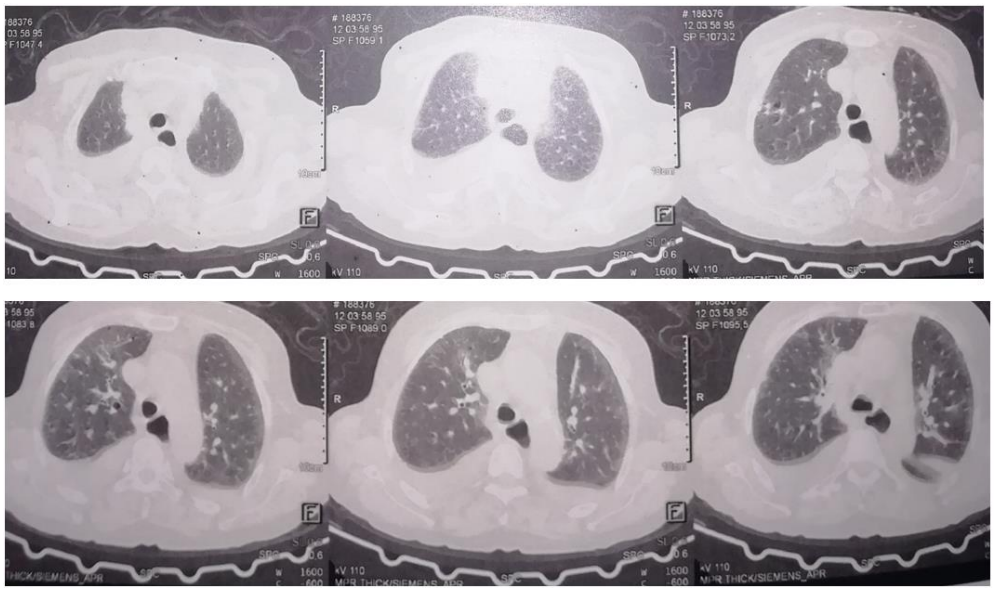

Figure 2: Interstitial damage with foci of dilations of sequella branches and a bilateral pleural effusion.

The thoracic tomodensitometry finds a minimal interstitial damage with foci of dilations of sequella branches, no signs in favor of a Covid 19 pneumopathy and a bilateral pleural effusion. MRI finds dilated cardiomyopathy with severe tricuspid leakage, absence of ischemic sequelae or acute myocarditis (Figure 3).

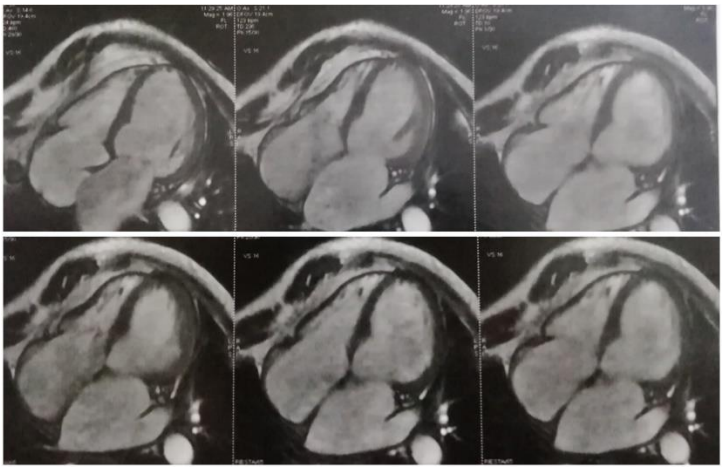

Figure 3: Appearance of dilated cardiomyopathy without ischemic sequelae.

24 hours ECG holter is in favor; permanent regular sinus rhythm with normal constant PR, low density atrial extrasystoles with passage to paroxysmal AF, 33 isolated monomorphic long-coupled ventricular extrasystoles without VT, absence of paroxysmal high degree conduction disturbances (Figure 4).

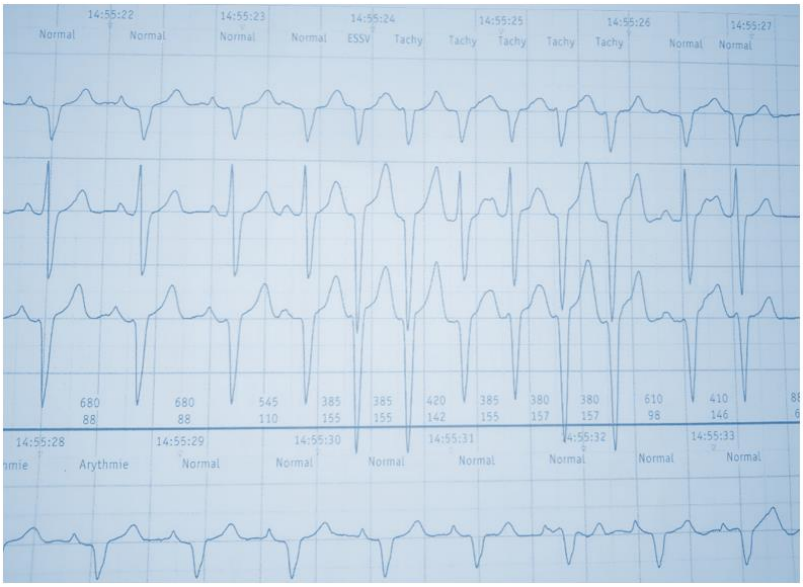

Figure 4: Atrial tachycardia 153/'.

3. TREATMENT AND EVOLUTION

For the outbreak of scleroderma disease, the patient received; Bosentan 125 mg/day and Methotrexate 15 mg/week and Adalat 20 mg/day, or heart failure the patient received bisoprolol 2.5 mg/day, furosemide 250 mg 2 */day, captopril 25 mg/day, Ertapenem 1 g/day, heparin calcium in curative dose. An electrical reduction is planned given the high probability of rhythmic heart disease added to this area of scleroderma with cardiac involvement, but the evolution is marked by sudden death of the patient.

4. DISCUSSION

Left ventricular systolic dysfunction is not an uncommon finding in advanced scleroderma, but the time course and susceptibility for this is not well understood. Systolic and/or diastolic dysfunction can occur as a result of

myocardial fibrosis but the role of ongoing low-grade myocarditis in this process is less well characterized. Anecdotally, patients with reduced ejection fraction and normal coronary arteries may benefit from increasing the patient's immunosuppression. It has been observed that patients with scleroderma with reduced ejection fraction and normal LV chamber size may improve their LV function with an increase in their immunosuppression regimen and a concomitant institution of appropriate drugs to treat heart failure (ACE/ARB, beta blockade, aldosterone antagonist). Overt congestive heart failure occurs in more advanced disease, but systolic dysfunction is often clinically occult [3,4]. As would be expected, there is a marked difference in symptoms and hemodynamics with exertion and, in one study 46% of patients had a reduced LVEF with exercise while only 15% of this group had reduced function at rest19 [5], Right heart failure is most commonly the result of pulmonary hypertension. Pulmonary hypertension is a common manifestation of scleroderma and a poor prognostic sign. It is the ability of the right ventricle (RV) to function under this increased load that determines both the severity of symptoms and survival [6,7]. Signs and symptoms of right heart failure by history, echocardiogram, and catheterization are associated with a significantly increased risk of death. Several echocardiography-derived parameters have been reported to correlate with poor outcome particularly with scleroderma [8]. Right atrial area index, the diastolic eccentricity index and the presence of a pericardial effusion were all predictors of the combined endpoint of death or transplantation; right atrial area index and pericardial effusion were also independent predictors of mortality. However. The use of TAPSE (Tricuspid Annular Plane Systolic Excursion) has been correlated to mortality [9]. Arrhythmias are seen frequently in scleroderma patients and are thought to be a result of fibrosis or ischemia [10]. That scleroderma patients had a higher mean heart rate. Depending on the underlying cardiac involvement, increased numbers and frequency of ventricular ectopic beats, as well as episodes of ventricular tachycardia, can be seen in scleroderma [11]. Cardiac involvement with a cardiomyopathy and ventricular arrhythmias is cause for great concern in scleroderma given the increased likelihood for sudden death. Electrophysiologic studies are recommended in this patient population and implantable automatic defibrillator is recommended in patients with inducible ventricular tachycardia or reduced LVEF.

5. CONCLUSION

Cardiac involvement in scleroderma can be fatal with new and more refined imaging modalities as well as more frequent use of invasive hemodynamics, we will be able to better assess patients for subclinical disease and acquire new knowledge about the long-term prognosis in patients with scleroderma. In addition, early detection will allow us to improve the quality of life and longevity of heart patients with scleroderma and prevent the risk of sudden death.

6. CONFLICT OF INTEREST

The authors declare that they have no competing interest.

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