

CASE REPORT

Complete Atrioventricular Block and Cardiopulmonary Involvement in Rapidly Progressive Systemic Sclerosis

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Abstract

The heart and lung are target organs in systemic sclerosis (SSc) and similar symptoms (dyspnea and cough) may make the differential diagnosis between the two lesions difficult. In addition, complete atrioventricular block (CAVB) is a rare complication of this disease. This case report is about a patient with SSc and pulmonary fibrosis who was admitted to the emergency room with CAVB, heart failure (HF) and progressive worsening of the underlying disease.

Introduction

Systemic sclerosis (SSc) is a multifactorial autoimmune connective tissue disease, with high morbidity and mortality rates, whose prevalence in the general population is 5%.^{1,2} It is characterized by vascular injury and fibrosis of the skin and internal organs, the heart and lungs being the most frequently involved organs.^{3,4} It is divided into 2 main subsets based on the extent of cutaneous involvement, limited and diffuse; the latter is associated with more visceral involvement.¹⁻³ Cardiac involvement can affect the pericardium, the myocardium, and the conduction system.^{4,7} Complete atrioventricular block (CAVB) is the least common conduction abnormality.^{4,7} Here, we report a case of rapidly progressive systemic sclerosis complicated by CAVB and heart failure (HF).

Keywords

Heart Failure; Atrioventricular Block; Scleroderma, Systemic; Hypertension, Pulmonary; Pulmonary Fibrosis.

Case Report

We report the case of a 50-year-old black man diagnosed with rapidly progressive diffuse Systemic Sclerosis, confirmed by clinical and serological tests⁸ under treatment with methotrexate, folic acid and monthly pulse therapy with methylprednisolone and cyclophosphamide. He sought care in the emergency room presenting with a clinical picture of HF, with progressive worsening in the last three months, in addition to evening fever. On physical examination he presented tachypnea, with no signs of respiratory effort, cold extremities, slow capillary filling, JVP at 45 degrees and cannon "a" wave in JVP and diffuse skin thickness. Blood pressure 110/70 mmHg, heart rate 42 bpm and respiratory rate 26 bpm. The examination of the thorax revealed left deviation of the ictus cordis, regular heart rate, presence of LV third heart sound, a grade 2/6 systolic murmur and a grade 3/6 tricuspid regurgitation murmur. Lungs with bilateral crackles. Hepatomegaly with pain on palpation. Bilateral lower extremity edema (2+/4+). Electrocardiogram (ECG) showed CAVB (Figure 1).

Therapy with intravenous furosemide, spironolactone and enalapril was initiated upon admission, and the patient was referred to the cardiac intensive care unit. On the second day of hospitalization, a permanent dual-chamber, epicardial pacemaker was implanted (Figure 2).

The transthoracic echocardiography (TTE) revealed increased left cavities, diffuse hypokinesia, LVEF of 38% (Simpson's rule), moderate mitral and tricuspid regurgitation and PSAP 65 mmHg. Thoracic computed tomography (CT) displayed ground-glass opacity distributed diffusely through both lungs, bronchiectasis, inter and intralobular septal thickening and paraseptal emphysema in the upper lobes. Laboratory tests showed

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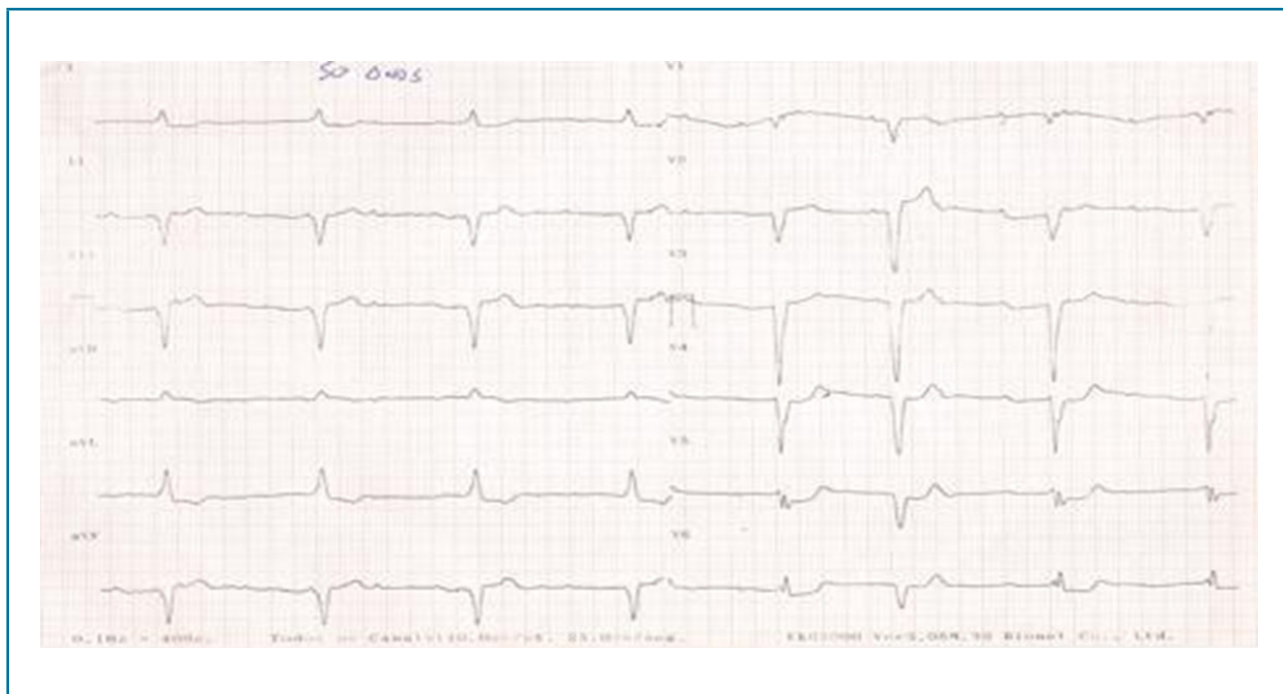


Figure 1 - 12-lead ECG showing CAVB.

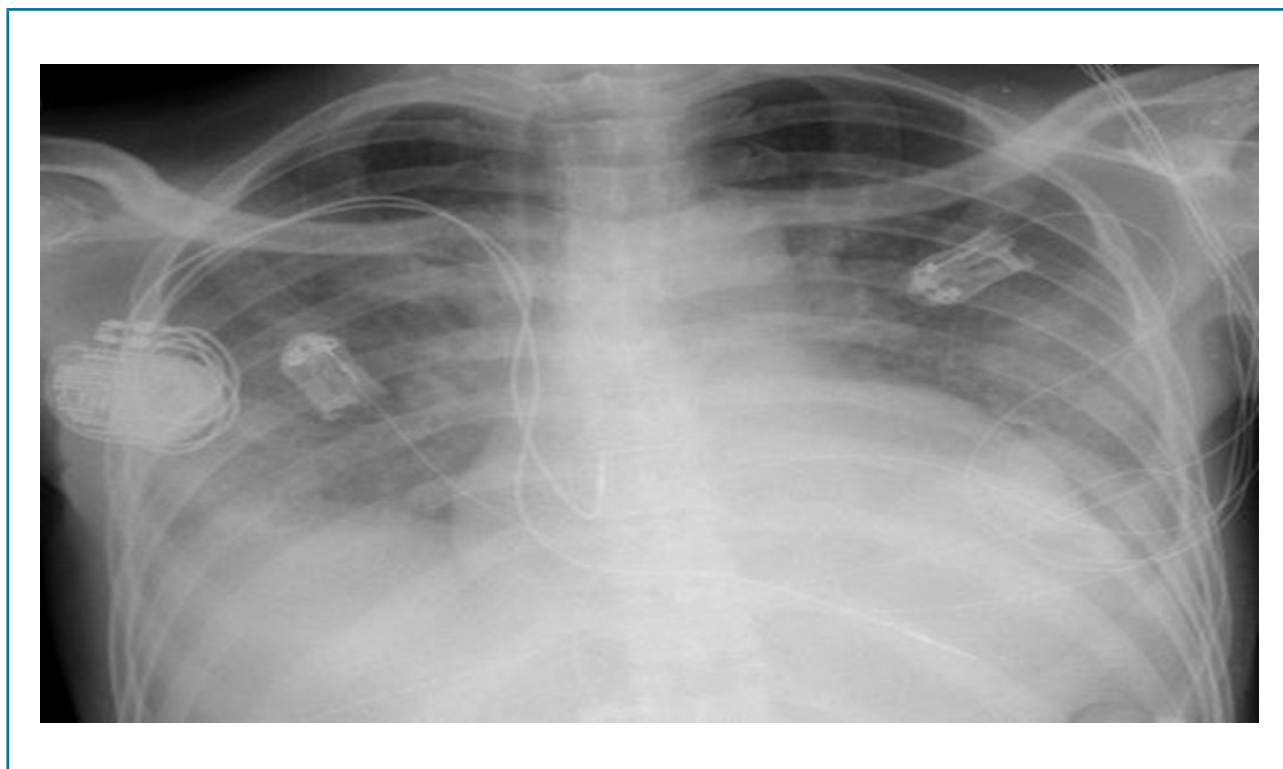


Figure 2 - Bedside PA chest x-ray showing cardiomegaly and pulmonary congestion. In addition, a right-sided dual-chamber pacemaker can be observed.

the presence of anemia, increased CRP and increased NT-proBNP levels (Table 1).

Despite correction of the conduction disorder, there was clinical worsening of HF and of the underlying disease activity with the ongoing treatment, thus a decision was made to start rituximab as rescue therapy⁸ of SSc. There was progressive worsening of HF symptoms despite optimal medical therapy and the patient evolved to death due to refractory cardiac shock.

Discussion

Cardiac involvement in SSc includes pericarditis, myocardial disease and conduction abnormalities. Between 25% and 75% of the patients have electrocardiographic abnormalities, such as ST segment changes, ventricular or supraventricular arrhythmias and conduction disorders.² The most frequent conduction abnormality in SSc patients are left branch block, first-degree AV block, left anterior fascicular block and right branch block. CAVB is a rare complication that affects less than 2 percent of patients.^{4-7,9} In a large international series of 3656 SSc patients, conduction disorders were observed in 12.7% of them.¹⁰

Lung disease is seen in 61% of SSc patients,¹¹ and the most prevalent clinical manifestations are pulmonary fibrosis and pulmonary vascular disease, which cause arterial pulmonary hypertension (APH).^{1-4,8} The prevalence of APH varies according with the diagnostic

method used: between 8 and 12%, by right cardiac catheterization, and about 38% by TTE.^{1,2,4} APH is often diagnosed late in the evolution of the disease, as observed in our patient.

Patients who have clinically evident cardiopulmonary involvement evolve with a worse prognosis;^{1-4,8} therefore screening of subclinical involvement in patients with SSc should be considered. Nevertheless, data on the best screening method are not well-defined yet. Initial investigation of cardiac involvement must include ECG, TTE and cardiac biomarker testing, such as brain natriuretic peptide (BNP) or its inactive form NT-proBNP.² In asymptomatic patients, without confirmed diagnosis by initial examination, the 24h Holter and cardiac imaging exams, such as magnetic resonance imaging, may be indicated.^{8,11}

The rapid progression of the underlying disease, refractoriness to HF treatment and the significant presence of APH contributed to the patient's death.

Conclusion

The reported case reinforces the importance of early diagnosis of cardiac and pulmonary involvement in SSc, aiming at better therapeutic approaches and the reduction of morbidity and mortality.

Author contributions

Conception and design of the research: Nani ES, Mocarzel LOC, Gismondi RA. Acquisition of data: Viegas EC, Ávila DX. Writing of the manuscript: Viegas EC, Ávila DX. Critical revision of the manuscript for intellectual content: Nani ES, Mocarzel LOC, Gismondi RA.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

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Table 1 - Patient laboratory data

	Admission	D14
Hemoglobin (g/dL)	10.8	9.8
Hematocrit (%)	34.2	30.9
Leukocytes (mm ³)	10,700	10,600
Bands (%)	1	2
CRP (mg/dL)	8.91	13.61
INR	1.15	
Urea (mg/dL)	73	65
Creatinine (mg/dL)	1.06	0.86
Sodium (mEq/L)	133	130
Potassium (mEq/dL)	4.2	4.4
Nt-proBNP (pg/mL)	40,643	101,821

Ethics approval and consent to participate

This article does not contain any studies with human participants or animals performed by any of the authors.

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