

# Cardiac involvement in systemic sclerosis. A case with wide-QRS tachycardia

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Cardiac involvement is one of the most frequent complications in systemic sclerosis but the asymptomatic course of the disease make early diagnosis and treatment difficult. We present the case of a woman diagnosis with systemic sclerosis who presented with an acute episode of wide-QRS tachycardia. [Emergencias 2008;20:285-288]

**Key words:** Systemic sclerosis. Cardiac diseases. Tachycardia, ventricular.

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## CONFLICT OF INTEREST:

None

## Introduction

Cardiac involvement in systemic sclerosis (SS) is one of the most frequent visceral complications and is the main determining factor of the prognosis of the disease<sup>1</sup>. Alterations in rhythm, conduction defects, autonomic dysfunction or pericardial disease may be found<sup>2</sup>. The clinical manifestations are infrequent and unspecific and are often confused with other complications of the disease such as pulmonary or renal events, making early diagnosis as well as prevention of complications even more difficult. We describe the case of a woman diagnosed with systemic sclerosis with cardiac manifestations of wide-QRS tachycardia.

## Clinical case

A 36-year-old female patient diagnosed with systemic sclerosis and secondary Raynaud phenomenon in 2000 was attended. In July 2005 she presented an acute episode of dyspnoea and palpitations after moderate effort for which she came to the Emergency Department. Physical examination on arrival showed arterial pressure of 103/66

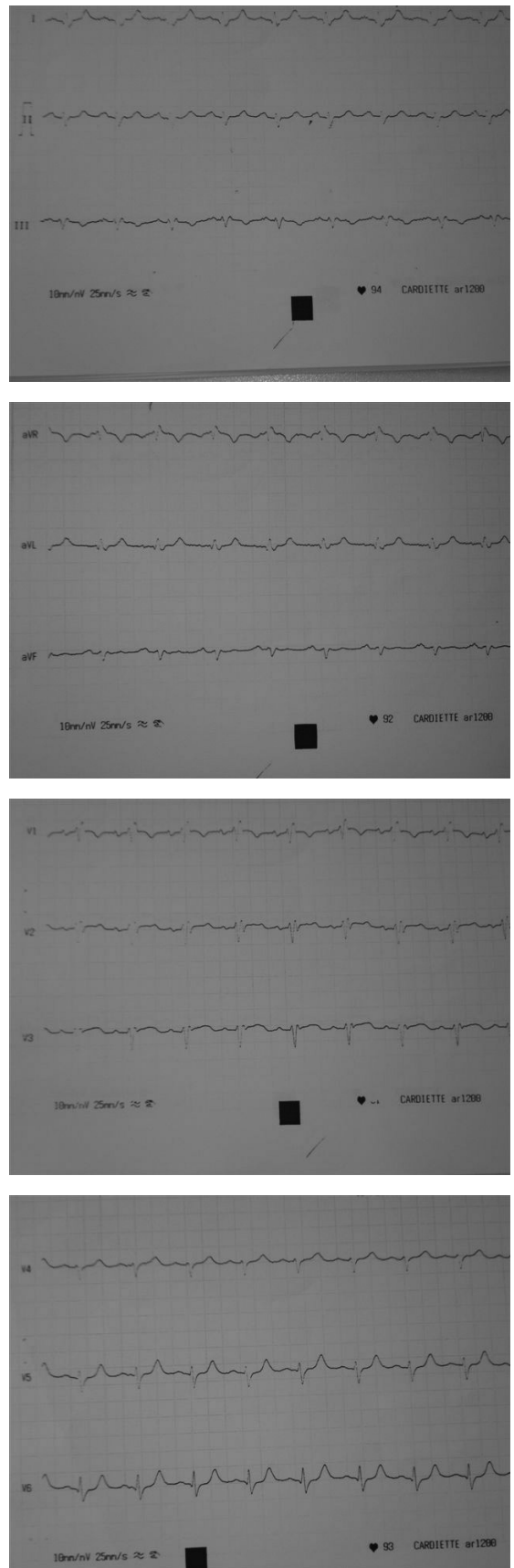
mmHg and cardiac frequency of 220 per minute with arterial oxygen saturation by pulse oximetry of 95%. On cardiac auscultation, the beats were rhythmic with holosystolic murmur in the tricuspid foci. Blood analysis, haemostasis and biochemical analysis for enzymes demonstrating myocardial lesion which were normal were performed. The electrocardiogram (ECG) demonstrated wide-QRS tachycardia (Figure 1). Chest x-ray showed a cardiothoracic index within the upper limit of normality with no other findings of note. After the administration of antiarrhythmic treatment with amiodarone, sinus rhythm reverted with complete blockade of the right branch in the control ECG (Figure 2) and good clinical response. During admission a echocardiographic study was performed showing hypertrophy of the asymmetric septal left ventricle with a left ventricular ejection fraction (FEV1) estimated of 50%, thinned interventricular septum and flattening suggestive of overload, dilatation of the auriculum and right ventricle, severe tricuspid insufficiency and very mild pulmonary hypertension (31 mmHg) (Figures 3 and 4). Treatment with verapamil was initiated and an implantable automatic defibrillator was implanted one month later with symptomatic control.



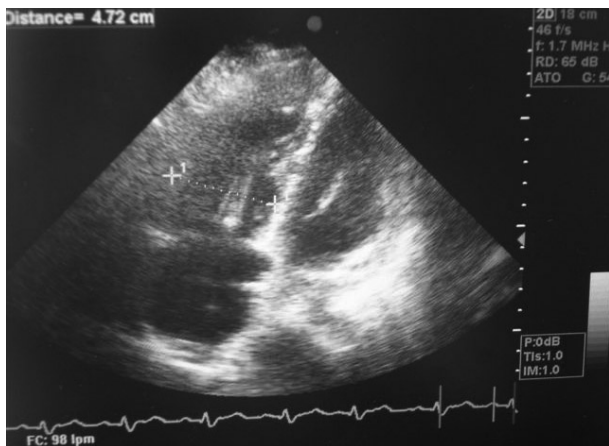
**Figure 1.** 12-lead electrocardiogram showing tachycardia with wide QRS.

## Discussion

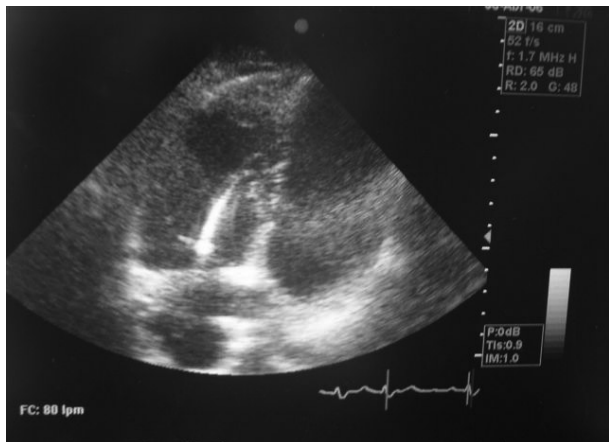
Different types of cardiac arrhythmia have been described in patients with SS. Several studies have demonstrated that up to 50% of the patients with SS present electrocardiographic alterations such as auricular arrhythmias (61%



**Figure 2.** 12-lead electrocardiogram showing sinus rhythm.



**Figure 3.** Left ventricle hypertrophy and thinning of the interventricular septum.



**Figure 4.** Severe tricuspid insufficiency.

supraventricular ectopic beats, 21% series of supraventricular tachycardia) ventricular (67% ventricular ectopic, 7% ventricular tachycardia) or alterations in conduction<sup>3</sup>. In the limited forms of SS the values range from 25%-38% of patients according to the series of branch blockages, auricular-ventricular blockages, alterations in systolic pressure and ventricular hypertrophy<sup>4</sup>.

The mechanisms implicated in the pathogenesis of SS are a selective primary involvement of the conduction system and/or diffuse myocardial fibrosis (up to 81% of the patients with SS)<sup>5</sup> possibly secondary to necrosis in contraction bands as a sequela of intermittent microvascular ischaemia (Raynaud microcoronary)<sup>1,3,5,6</sup>. Fibrosis in SS may be histologically distinguished from that observed in the atherosclerotic coronary arteries. The areas of fibrosis in SS do not correspond with the regional distribution of any coronary artery. There are no myocardial haemosiderin deposits and it is usually located in the subendocardial region<sup>2</sup>.

Ventricular hypertrophy and septal asymmetry have been observed in patients with SS in the absence of arterial hypertension in a greater percentage than in the general population as well as diastolic dysfunction and a reduction in the left ventricular ejection fraction on resting (15.3%) and particularly during exercise (46%).

The clinical manifestations are not specific and are infrequent and may be confounded with symptoms due to other disorders such as vascular or interstitial pulmonary complications found in SS. It has been calculated that cardiac involvement in SS is clinically evident in only 20%-25% of the cases with a mortality of 70% at 5 years<sup>1</sup>. The most frequent symptoms are tiredness and dyspnoea which may overlap with those due to fibrosis and/or pulmonary hypertension, anaemia or musculoskeletal involvement<sup>2,4</sup>. Other symptoms include palpitations and tachycardia as a consequence of alterations in rhythm of autonomic dysfunction and chest pain in cases of pericarditis. On physical examination ventricular galloping, sinus tachycardia, signs of congestive heart failure and pericardic rubbing. Presentation as ischaemic heart disease is infrequent<sup>1,2,5</sup>.

Myocardial involvement is the main factor determining survival in SS<sup>5</sup>. There is a clear relationship between ventricular arrhythmias (ventricular ectopic and ventricular tachycardia) and total mortality by sudden death<sup>2,5</sup>.

Non invasive techniques such as ECG, echocardiography and Holter monitoring are useful for evaluation. The prevalence of conduction alterations significantly increases with the Holter study in comparison with the electrocardiogram (from 32% to 62%)<sup>2</sup>.

With regard to treatment, angiotensive enzyme converter inhibitors (AECI) and calcioantagonists have been used for the treatment of ventricular dysfunction, antiarrhythmics and pacemakers for rhythm disorders and defibrillators or radiofrequency ablation have been implemented in cases of conduction defects<sup>2,6</sup>.

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## **Afectación cardiaca de la esclerosis sistémica. Un caso de taquicardia de QRS ancho**

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La afectación cardiaca de la esclerosis sistémica es una de las complicaciones más frecuentes de la enfermedad pero suele cursar de forma asintomática lo que dificulta su diagnóstico y tratamiento precoz. Presentamos el caso de una mujer con diagnóstico de esclerosis sistémica que presenta un episodio agudo de taquicardia de QRS ancho. [*Emergencias* 2008;20:285-288]

**Palabras clave:** Esclerosis sistémica. Afectación cardiaca. Taquicardia de QRS ancho.