Case study

Complete heart block revealing a systemic sclerosis disease, a case report

ABSTRACT

Complete atrioventricular block represents the major conductive disorder in cardiology, it is a diagnostic and therapeutic emergency

Systemic scleroderma (SSc) is an autoimmune disease with fibrotic tropism of connective tissues in its pulmonary, skin and vascular location. It is often associated with cardiopulmonary manifestations increased by pulmonary hypertension, with complications of a rhythmic nature especially

Cardiac conductive disorders in the context of scleroderma is a rare entity with a poorly understood physiopathological mechanism.

We report the case of a 47 years old patient with complete syncopal atrioventricular block revealing a systemic sclerosis.

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INTRODUCTION

Systemic sclerosis represents connective tissue damage characterized by vasomotor disorders due to fiborsis preferentially affecting the lungs, skin and blood vessels.

Cardiac involvement is often marked by the repercussions of pulmonary hypertension, conduction disorders are rarely described, their pathophysiological mechanism is not clearly established.

We report the observation of a syncopal emergency in a young patient revealing systemic sclerosis

CASE PRESENTATION:

47 old man, was admitted in the cardiology department for an episode of syncope associated with dizziness and fainting, without chest pain or dyspnea

The patient reported the presence of Raynaud's phenomenon for 10 years, and progressive skinthickening and hypopigmentation.

On admission examination: Blood pressure was 140 over 65 mm, cardiac pulse was 45 bpm, without signs of heart failure. Cardiac and pulmonary auscultation were unremarkable. The mucocutaneous examination revealed diffuse skin sclerosis, erasing of facial wrinkles, a tapering nose with telangiectasias, pursed lips with limited mouth opening and sclerodactyly with pulp ulcerations (Figure 1).

The electrocardiogram (ECG) showed complete left bundle branch block and complete atrio-ventriuclar block (Figure 2).

A transthoracic echocardiography showed a normal left ventricle with an ejection fraction of 57%, an undilated atriums, with a normal right ventricle.

The laboratory tests revealed no abnormalities except an inflammatory syndrome. The titer of antinuclear antibodies and Antitopoisomerase-1 antibodies were positive which led us to confirm the diagnosis of diffuse SSc.

Temporary endocavitary cardiac stimulation was placed(Figure 3) in the hope of regression of atrioventricular block. SSc treatment was initiated with prednisone, cyclophosphamide. During his hospitalization,a permanent double chamber pacemaker was implanted a few days later (Figure 4).

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DISCUSSION

Complete atrioventricular block is a heart emergency due to a complete disruption of conduction between the atria and the ventricles. [1]

The most frequent aetiologies are represented by the degenerative, ischemic, metabolic and congenital causes, rarely an autoimmune origin such as systemic lupus erythematosus; systemic sclerosis is a very rare entity. The literature reports a few orphan cases[2, 3].

Systemic sclerosis is an autoimmune pathology due to connective tissue damage manifested by several organ fibrosis damage, such as vascularitis, gastrointestinal, pulmonary and skin locations; cardiac involvement is directly caused by myocardial fibrosis or ischemia or is secondary to pulmonary arterial hypertension, 26% of scleroderma deaths were of cardiac origin, of which 42% were attributed to arrhythmia[4, 5].

> The physiopathology of cardiac conduction disturbances in patients with systemic sclerosis is not clearly defined[4].Coronary microvascular damage is often implicated in which ischemia could increase the incidence of conduction disorders. The installation of myocardial fibrosis is a second track that could affect the prognosis, the different parts of the conductive system could be destroyed with progressive fibrosis, collagen deposition and degenerative changes[6, 7].

Post-mortem histological examination revealed in some cases that the sinoatrial node and AV node as well as its initial par were unharmed, with greater involvement at the distal level of the atrio-ventricular node and the bundles of His [8].

The standard treatment for symptomatic complete atrioventricular block is the permanent implantation of a pace maker as was done for this patient, corticosteroids are often ineffective, the various cases reported in the literature have objected to a persistence of CAB despite the medical treatment, this could be explained by the irreversible installation of fibrosis[3, 9].

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The implantation of a pacemaker is mandatory in cases of high degree symptomatic AVB (syncope or congestive heart failure.) As well as patients whose activity levels are altered due to a decrease in tolerance to the exercise. prophylactic stimulation in asymptomatic patients reduced the incidence of serious complications such as syncope and sudden death[10]

CONCLUSION

Atrioventricular block in the context of systemic scleroderma is a rare entity, often irreversible in spite of the etiological treatment, requiring implantation of a pacemaker.

The autoimmune origin of conduction disturbances should include screening for systemic sclerosis in the presence of orienting signs, especially in young patients.

Disclaimer regarding Consent and Ethical Approval:

As per university standard guideline, participant consent and ethical approval have been collected and preserved by the authors

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FIGURES

Figure 1: Sclerodactyly

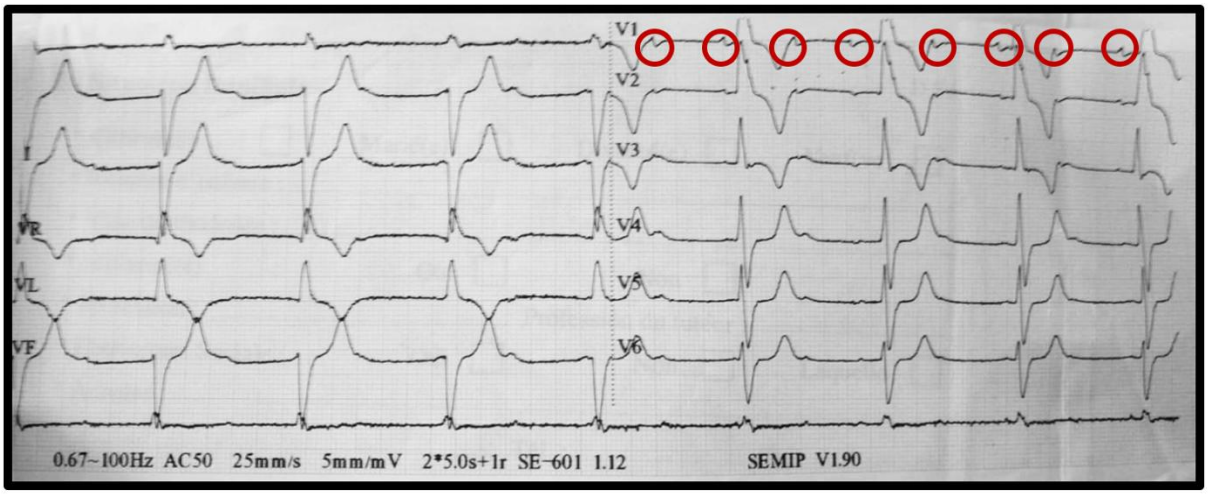

Figure 2: EKG showing a complete heart block

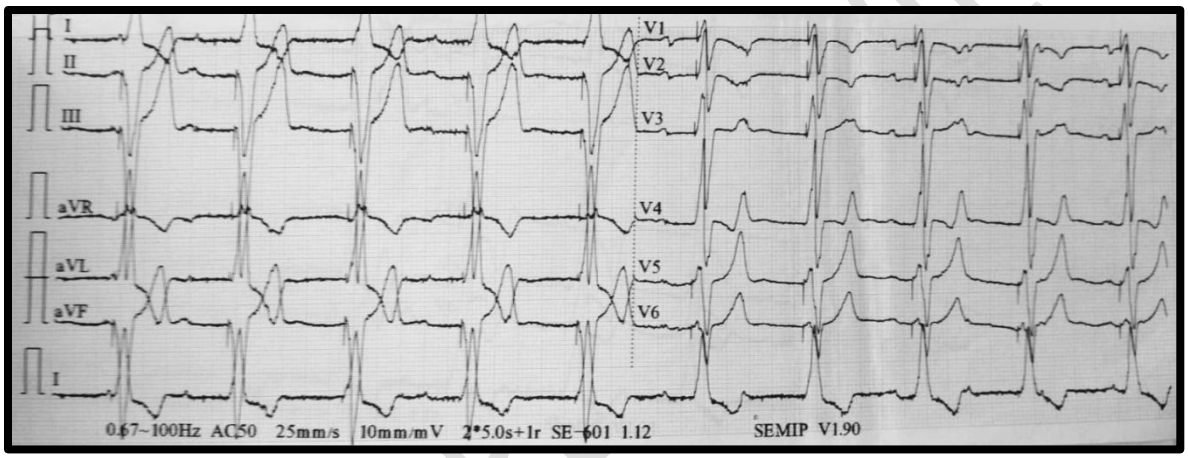

Figure 3: EKG after pacemaker implantation

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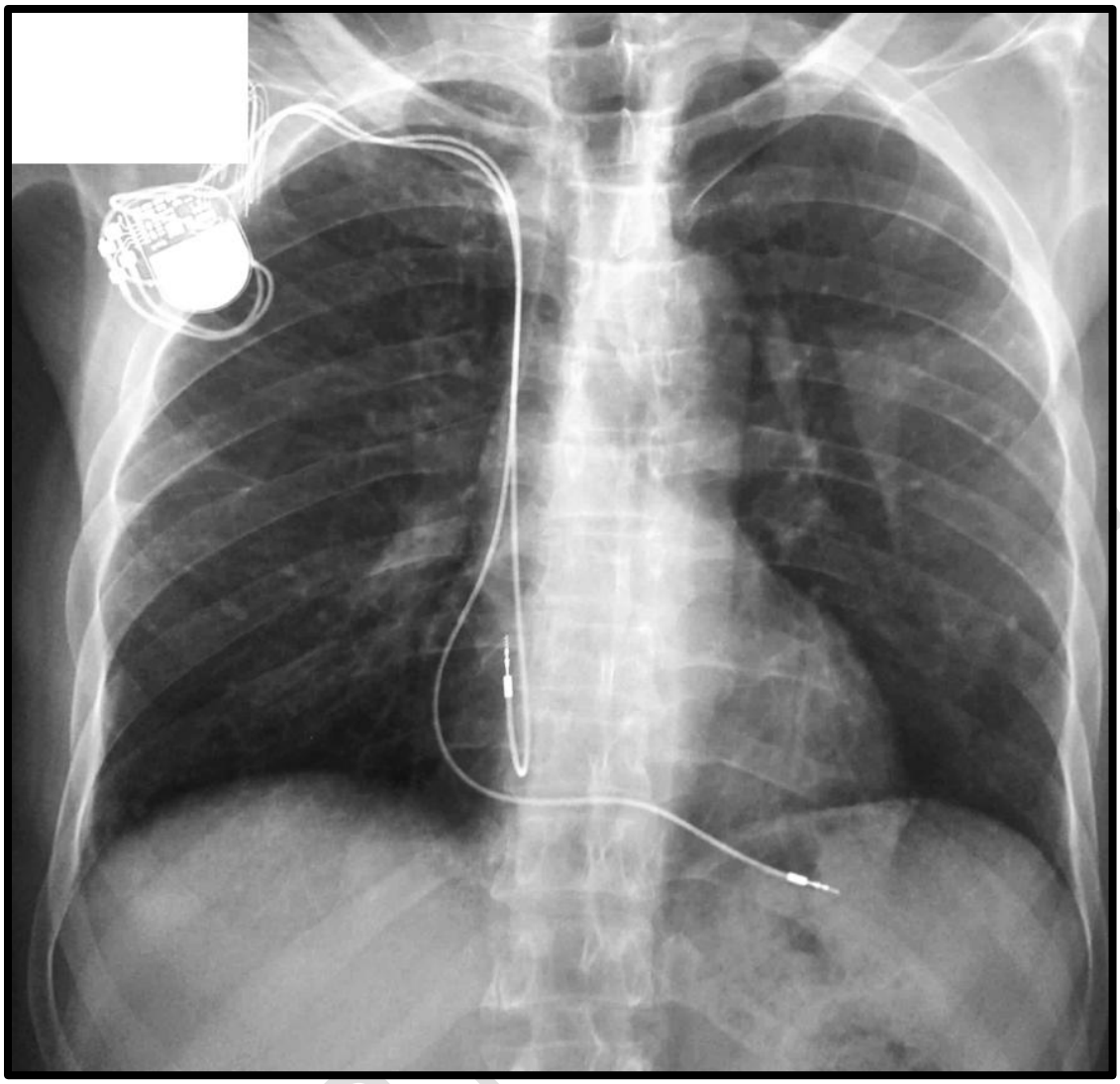

Figure 4:Thoracic X Ray Image showing a double chamber Pacemaker

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