Clinical Case Report

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Complete heart block in systemic sclerosis

A case report and literature review

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Abstract

Rationale: Complete heart block (CHB) is a common clinical manifestation. Systemic sclerosis (SSc) is a rare etiology of CHB that has not received enough attention. Whether pacemaker implantation is required remains inconclusive, especially when patients have no symptoms or mild symptoms of CHB.

Patient concerns: In this study, we report the case of a 48-year-old Chinese male who suffers from SSc and CHB.

Diagnose: The patient was previously diagnosed with left anterior hemiblock (LAHB) and right bundle block with normal heart function. CHB was observed on a regular follow-up electrocardiogram (ECG) examination 1 month before his hospitalization.

Interventions: A permanent dual chamber pacemaker was implanted.

Outcomes: The patient responded well to pacemaker implantation treatment, and his exertional dyspnea disappeared.

Lessons: The occurrence of heart block associated with SSc often appears concealed. The case highlights the importance of regular follow-up of a patient with SSc. Pacemaker implantation might be unavoidable if CHB is secondary to SSc, even if it is asymptomatic.

Abbreviations: CHB = complete heart block, CI = cardiac involvement, ECG = electrocardiogram, LAHB = left anterior hemiblock, LV = left ventricular, NR = normal range, RBBB = right bundle branch block, SSc = systemic sclerosis.

Keywords: cardiac pacing, complete heart block, complication, systemic sclerosis

1. Introduction

Complete heart block (CHB), a common condition caused by a local lesion of the heart, may be a complication resulting from various etiologies. Systemic sclerosis (SSc), a rare etiology of CHB, has not received enough attention. For patients with SSc, cardiac involvement (CI) is directly caused by myocardial fibrosis or ischemia or is secondary to pulmonary arterial hypertension.[1] According to the EULAR study, the estimated prevalence of CI in patients with SSc is more than 50%.[2] For 26% of the patients who died of SSc, the causes of death were cardiac related.[3] CI has been recognized as a poor prognostic factor for SSc, which is often asymptomatic and difficult to find in the early stage. Signs and symptoms of arrhythmias have been reported in patients with SSc. The incidence of supraventricular and ventricular arrhythmias in SSc patients was approximately 30% and 67%, respectively. However, advanced and CHB occurred rarely (<2%),[4] and right bundle branch block (RBBB) may be an independent predictor of mortality.[5] Whether pacemaker implantation is necessary remains inconclusive when asymptomatic CHB happens. Here, we describe a male patient with diffuse SSc who suffered from asymptomatic CHB.

2. Methods

Approval from the ethics committee of the Peking Union Medical College Hospital was obtained for this case report study. Detailed information about this study has been fully disclosed to the patient, and informed consent has been obtained.

3. Case report

A 48-year-old Chinese male was admitted to our hospital because his heart rate slowed down 1 month ago. He had been diagnosed with diffuse SSc 1 year ago when he suffered from Raynaud's phenomenon, skin pigmentation, sclerodactyly, weight loss, sour regurgitation, heartburn with slight dysphagia and malaise. In a previous electrocardiogram (ECG), normal sinus rhythm with bifascicular intraventricular block [RBBB and left anterior hemiblock (LAHB)] was shown. At that time, his drug regimen included prednisone 5 mg per day, cyclophosphamide 50 mg twice a day, tripterygium glycoside 10 mg twice a day and aspirin

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100 mg per day, but previous medications included methyl prednisolone. Apart from mild exertional dyspnea, the patient did not have edema or other obvious symptoms. His cardiac function was in class II according to the grading standard of the New York Heart Association (NYHA). On admission, after physical examination, we found that the patient had diffuse skin tightening, waxy luster, sausage appearance of the fingers, depressed scar and escharosis of the fingertips (see supplemental content, which showed typical skin manifestations of diffuse SSc, http://links.lww.com/MD/C632). His heart rate fluctuated from 37 to 42 beats per minute, and his blood pressure was 120/60 mmHg. On auscultation, Cannon's sound was audible, and a grade II/VI systolic murmur was heard at the lower left parasternal border. Blood tests were normal, the lymphocyte proportion was moderately reduced (16.3%, normal range (NR) 20.0%-40.0%), and NT-probrain natriuretic peptide was mildly increased (437 pg/mL, NR < 125 pg/mL). The titer of antinuclear antibodies was 1:640, and high-sensitive c-reactive protein was also above the normal level (60 mg/L, NR:0–3 mg/L). The pulmonary function test showed slight pulmonary fibrosis, which was also proved by a computerized tomography check. The resting ECG revealed normal sinus rhythm with complete atrioventricular block associated with bifascicular intraventricular block (RBBB and LAHB) (Fig. 1). A transthoracic echocardiogram showed a slightly enlarged right atrium (42 mm, NR: 28–40 mm), a normal left ventricular (LV) end-diastolic diameter (53 mm) and a normal LV systolic function with an ejection fraction of approximately 70%. There was a degree of tricuspid regurgitation with a maximal speed of 2.4 m/s. Treatment was initiated with prednisone, cyclophosphamide, tripterygium glycoside, and aspirin added to the original drug treatment regimen. In addition, oxygen, potassium, and magnesium were supplemented. The hemodynamics were stable during treatment. Except for SSc, the patient did not have any disease. No clues to other diseases and drugs that could result in arrhythmia were found. On the third day of admission, a dual chamber pacemaker procedure was performed without any complications. The remaining hospital course was uneventful. After 6 months, the patient was able to tolerate mountain climbing exercise. He is in good condition with a good quality of life after 18 months of follow-up.

4. Discussion

SSc is a connective tissue disease of unknown etiology, characterized by fibrosis of multiple target organs, such as skin and internal organs (lung, gastrointestinal tract, kidney, and heart). Alone or in association with pulmonary arterial hypertension or interstitial lung fibrosis, CI is one of the main determinants of the overall prognosis of SSc.[6] A total of 26% of deaths were cardiac related, 42% of which were attributed to arrhythmia.[3] Fortunately, the incidence of CHB appears to be very low.[7,8]Table 1 shows the summary literature found by using (CHB) and SSc) as search terms in the PubMed database in the last decade. Only 4 cases were reported, and most were female (75%), with an average age of 60.5 ± 9.9 years. The length of time

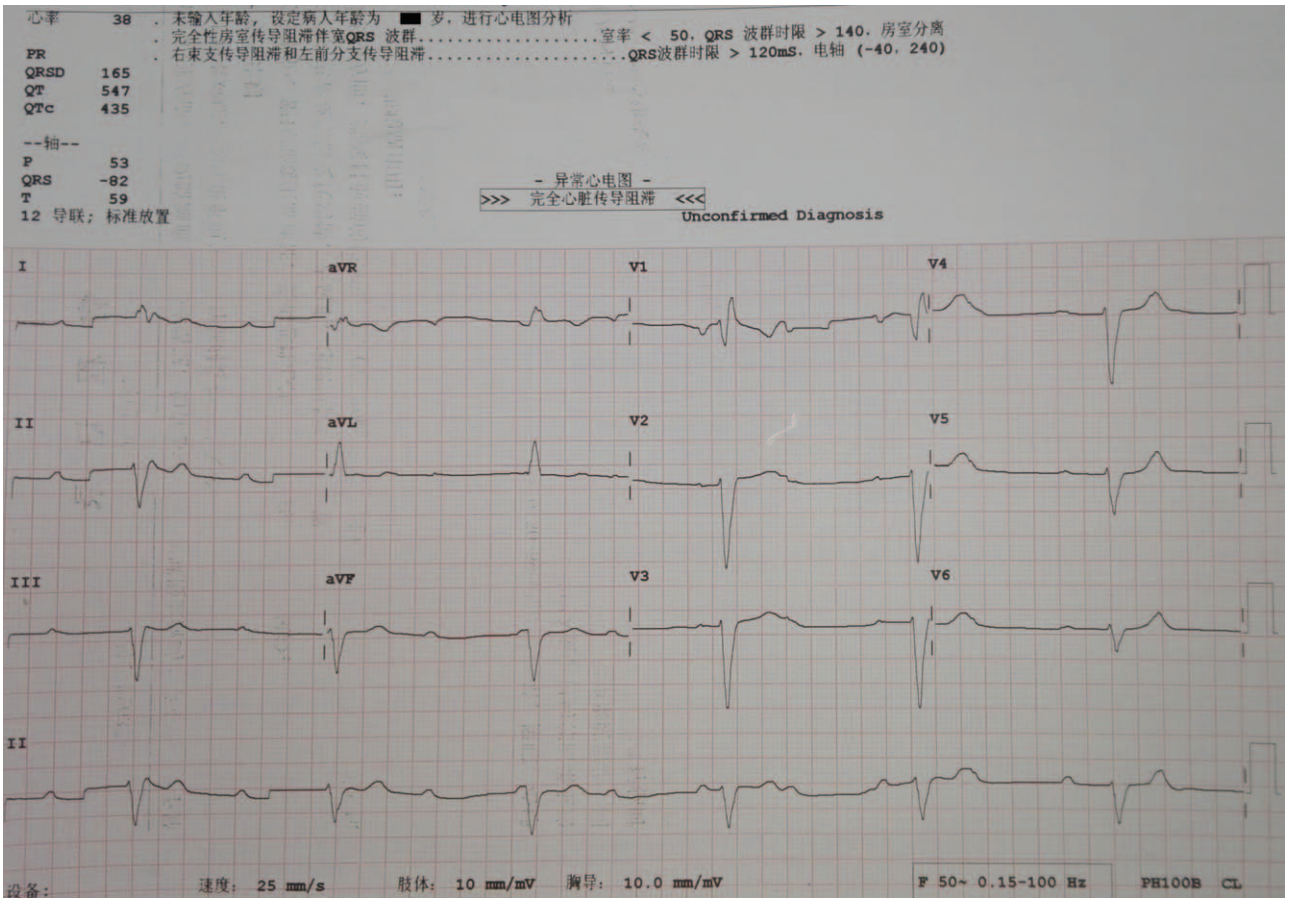

Figure 1. The electrocardiogram revealed normal sinus rhythm with complete atrioventricular block at admission.

Table 1

Systemic Sclerosis complicated with complete heart block in the last decade in PubMed database.

Literature	Sex	Age (year)	Disease course (year)	Clinical manifestation	Diagnosis	Pacemaker implantation
Canpolat, 2012[9]	Female	68	2	Generalized weakness, presyncopal episodes, and exertional dyspnea (NYHA class III)	Diffuse SSc	Yes
Femenia, 2010[8]	Female	47	NA	Syncope	SSc	Yes
Ciurzynski, 2007[10]	Male	59	22	Decreasing exercise tolerance with dyspnea	Diffuse SSc	Yes
Moyssakis, 2006[7]	Female	68	18	Generalized weakness, presyncopal episodes, and deterioration of dyspnea class IV (NYHA)	Diffuse SSc	Yes

NA = not applicable, NYHA = New York Heart Association, SSc = systemic sclerosis.

from onset to CHB in SSC patients was from 2 to 22 years, and all patients were administered pacemaker implantation.

The pathogenetic mechanism of heart conduction abnormalities in patients with SSc is not clearly understood. Coronary microvascular lesions are one of the hallmarks of CI.[11] Focal ischemia of the microvascular structure and functional abnormalities might increase the incidence of conduction block. Myocardial fibrosis is another pathophysiologic symbol of CI that could affect the prognosis of individuals with SSc.[12] The atrioventricular node, His bundle, or left and right bundle branches could be destroyed with progressive fibrosis, collagen deposition, and degenerative changes.[13] In cases of CHB resulting from SSc, instead of the normal appearance of a pear-shaped proximal atrioventricular node, the proximal part of the node was slender and nearly the same diameter as the distal part revealed by the postmortem histological examination.[14] However, morphologic abnormalities of the conduction system in some patients were difficult to attribute to progressive SSc per se. For patients without progressive SSc, myocardial lesions also had fibrous atrophy of a portion of the proximal left bundle branch. In addition, atrioventricular conduction tissue was intact in some patients with permanent CHB.[15] The antibodies reacted only with the Purkinje cells, which might play a pathogenetic role in autoimmune diseases and atrioventricular heart block.[16] Cardiac Purkinje cell antibodies did not seem to be the only epiphenomenon of Purkinje cell damage. They could be correlated with idiopathic atrioventricular blocks, which could explain why some medical phenomena could not be caused by morphological structure only.

Because it is a sporadic disease and there is the lack of large cohort study data, CHB in SSc is limited to case reports. Nevertheless, we remained vigilant because the consequences are serious once it happens. Early diagnosis, timely and reasonable therapy are associated with a better prognosis. Noninvasive evaluation with ECG and echocardiogram is useful in detecting the early CI. Moreover, 24 h-Holter, 2-dimensional speckle tracking imaging, nuclear angiography, and cardiovascular magnetic resonance could provide more valuable information for discovering early CI in patients with SSc.[13,17,18] We did not schedule too many medical assessments. Rapid targeted treatment (a pacemaker implantation) was administered after a definitive diagnosis to prevent adverse cardiovascular events.

Understanding the etiopathogenesis is of great importance to the management of arrhythmias secondary to SSc. Although the number of patients treated with pacemaker implantation increases with progression of the disease during long-term follow-up,[19] not every patient with CHB secondary to immune diseases needs permanent pacemaker implantation.[20] Implantation of a pacemaker is mandatory for patients with advanced second-degree block or CHB who are symptomatic of syncope or congestive heart failure. It is also recommended to pace in patients whose activity levels are impaired due to decreased exercise tolerance. In patients with CHB, prophylactic pacing has significantly decreased the incidence of previously reported severe complications such as syncope and sudden death.[21]

The patient was a middle-aged male with a chronic course. CHB might have shown a gradual progression because of his normal heart mechanical function and mild discomfort. We considered the possibility of returning to normal conduction function to be negligible. It was necessary to implant an artificial pacemaker, which might improve the prognosis. Even though myocardial biopsy may be helpful to ascertain the cause of SSc complicated with CHB, implantation of a pacemaker may be unavoidable. Recognition of the etiology may eventually affect the patient's quality of life and prognosis. The follow-up results confirm our initial judgment and treatment strategy.

Author contributions

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