



CASE REPORT

Systemic Scleroderma with Complete Heart Block

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Abstract

For patients with Systemic Sclerosis (SSc), cardiac involvement (CI) is directly caused by myocardial fibrosis or ischemia or is secondary to pulmonary arterial hypertension¹ Here we present a similar case of a female patient aged 45 yr old female who presented with complaint of recurrent syncopal attacks, and difficulty in swallowing food and liquids. She had typical CREST Syndrome manifestations with complete heart block (CHB). For which pacemaker implantation was done.

Keywords

Systemic Sclerosis, cardiac involvement (CI), CHB

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). Complete AV block is

a very rare but serious complication in SSc. It has been reported in only five previous case reports. Screening for cardiac involvement of SSc has historically been limited to electrocardiograms (ECG), echocardiograms, and laboratory tests including troponin-I (Tn-I) and B-type natriuretic peptide (BNP) levels (5).

Case report

Here we present a similar case of a female patient aged 45 yr old female who presented with complaint of recurrent syncopal attacks, and difficulty in swallowing food and liquids. Initially she had mild difficulty with eating food and swallowing eatables, then gradually she started experiencing tightening of skin, unable to close wrist properly, consequently her mouth opening also reduced, following which she started having shortening of digits which sometimes had distal digital infarcts too (*Figure 1*). On the day of presentation, she had variable heart rate fluctuating from 24-36bpm (*Figure 2*), her blood

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Figure 1

pressure was 110 systolic over 60 diastolic, she had this complaint of syncopal attacks for past one month, but she ignored her symptoms and gradually the frequency of such symptoms of blackouts increased and started hampering her daily routine activities, for which she sought medical attention.

Limited scleroderma (formerly known as CREST) syndrome is characterized by Calcinosis, Raynaud's phenomenon, Esophageal dysmotility, sclerodactyly, and telangiectasia and our patient had all of these typical manifestations. She was evaluated for systemic sclerosis. Which came out positive for diffuse scleroderma and ECG showed complete heart block, other blood workup came out within normal limits, No clues to other diseases and drugs that could result in arrhythmia were found.

Then she was diagnosed with diffuse systemic sclerosis complicating with complete heart block. A dual chamber pacemaker procedure was performed without any complications. The remaining hospital course was uneventful. She is being followed up in opd on regular basis.

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

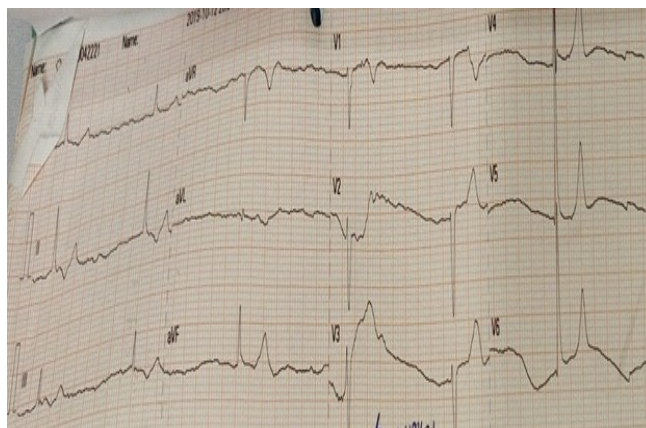


Figure 2

(6). A total of 26% of deaths were cardiac related, 42% of which were attributed to arrhythmia. Fortunately, the incidence of CHB appears to be very low (7). Although cardiac involvement is not uncommon in patients with systemic sclerosis, second- and third-degree AV block is rare, accounting for less than 2% of SSc cases with cardiac involvement (4). Instead, diastolic dysfunction and first-degree AV block are more frequently observed (8).

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 (9). 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 (10). The atrioventricular node, His bundle, or left and right bundle branches could be destroyed with progressive fibrosis, collagen deposition, and degenerative changes (11). The antibodies reacted only with the Purkinje cells, which might play a pathogenetic role in autoimmune diseases and atrioventricular heart block (12).

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.

Conclusion

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 (13), not every patient with CHB secondary to immune diseases needs permanent pacemaker implantation (14). Implantation of a pacemaker is mandatory for patients with advanced second-degree block or CHB who are symptomatic of syncope or congestive heart failure

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