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Severe atrial fibrosis as a cause of significant intraatrial conduction delay in a patient with scleromyositis

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ABSTRACT

Scleromyositis is an autoimmune disease and an overlap syndrome of scleroderma and poly/dermatomyositis. It is characterized by frequent cardiovascular involvement including heart failure, arrhythmias and conduction disturbances. We present a case of a 73-year old female patient who required an upgrade from a DDD pacemaker to cardiac resynchronization therapy due left ventricular dysfunction and permanent ventricular pacing. Electroanatomical mapping (CARTO 3D) revealed extensive right atrial fibrosis which resulted in significant delay in intraatrial conduction. Interval from atrial paced stimulus to A signal in His bundle was 364 ms, while AH and HV intervals were within normal range.

Introduction

Scleromyositis is an autoimmune disease and an overlap syndrome of scleroderma and poly/dermatomyositis [1]. Cardiovascular involvement including heart failure, arrhythmias and conduction disturbances is frequently observed in patients with connective tissue diseases [2]. Progressive fibrosis of myocardium is believed to be responsible for occurrence of conduction disturbances [3]. Significant fibrosis in atria may result in extremely low voltage of P waves in surface electrocardiogram leading to potential difficulties and mistakes in evaluation of heart rhythm. Furthermore, fibrosis is known to be associated with elevated sensing and pacing thresholds in patients with cardiac implantable electronic devices [4].

Case report

We present a case of a 73-year old female patient with scleromyositis, implanted with dual chamber pacemaker due to bradycardia-tachycardia syndrome (sinus and AV node conduction disturbances and atrial tachyarrhythmias) in 2012. Four years later this patient underwent new atrial lead implantation due to reported loss of sensing and pacing. At the time of reimplantation a patient was in II NYHA class, echocardiography showed left ventricle ejection fraction (LVEF) of 40%. No more detailed data was available based on existing medical history. One year after this procedure this patient was referred to our Department due to aggravated signs of heart failure (III NYHA class) and suspected malfunctioning of a replaced atrial lead based on lack of visible paced P waves in surface ECG. Fig. 1 shows patient's electrocardiogram with DDD pacing. No clearly visible P waves after atrial spikes and effective ventricular pacing after 350 ms atrioventricular delay were observed.

Transthoracic echocardiography revealed worsening of LVEF to 35% and severe enlargement of atria – left atrium volume index 82 cm^3/m^2 and right atrium volume index 104 cm^3/m^2. Preserved atrial activity was observed on echocardiography as documented by mitral flow. Furthermore, high echocardiographic probability of pulmonary hypertension was observed (SPAP 61 mmHg; TR-Vmax = 3.4 m/s; RAP 15 mmHg; PV-AcT = 65 ms). Diastolic dysfunction with restrictive inflow mitral pattern was present (E/A 2.4; TDI e-mean = 4.5 cm/s; E/e′ = 28).

Interrogation of a pacemaker revealed intrinsic junctional rhythm at 45 bpm. During atrial pacing at 50 bpm 1:1 conduction to ventricles with long S—V interval (470 ms) was observed. No intrinsic atrial activity was observed during pacemaker interrogation. Pacemaker memory showed 100% of ventricular pacing over the last 6 months. Electrophysiological study with 3D electroanatomical mapping (CARTO 3D) was performed in order to understand the nature of conduction disturbances and qualify a patient to further electrotherapy. Electroanatomical mapping revealed that extensive first degree atrioventricular block visible in surface ECG resulted from extremely slow conduction through severely fibrotic right atrium. Interestingly, the

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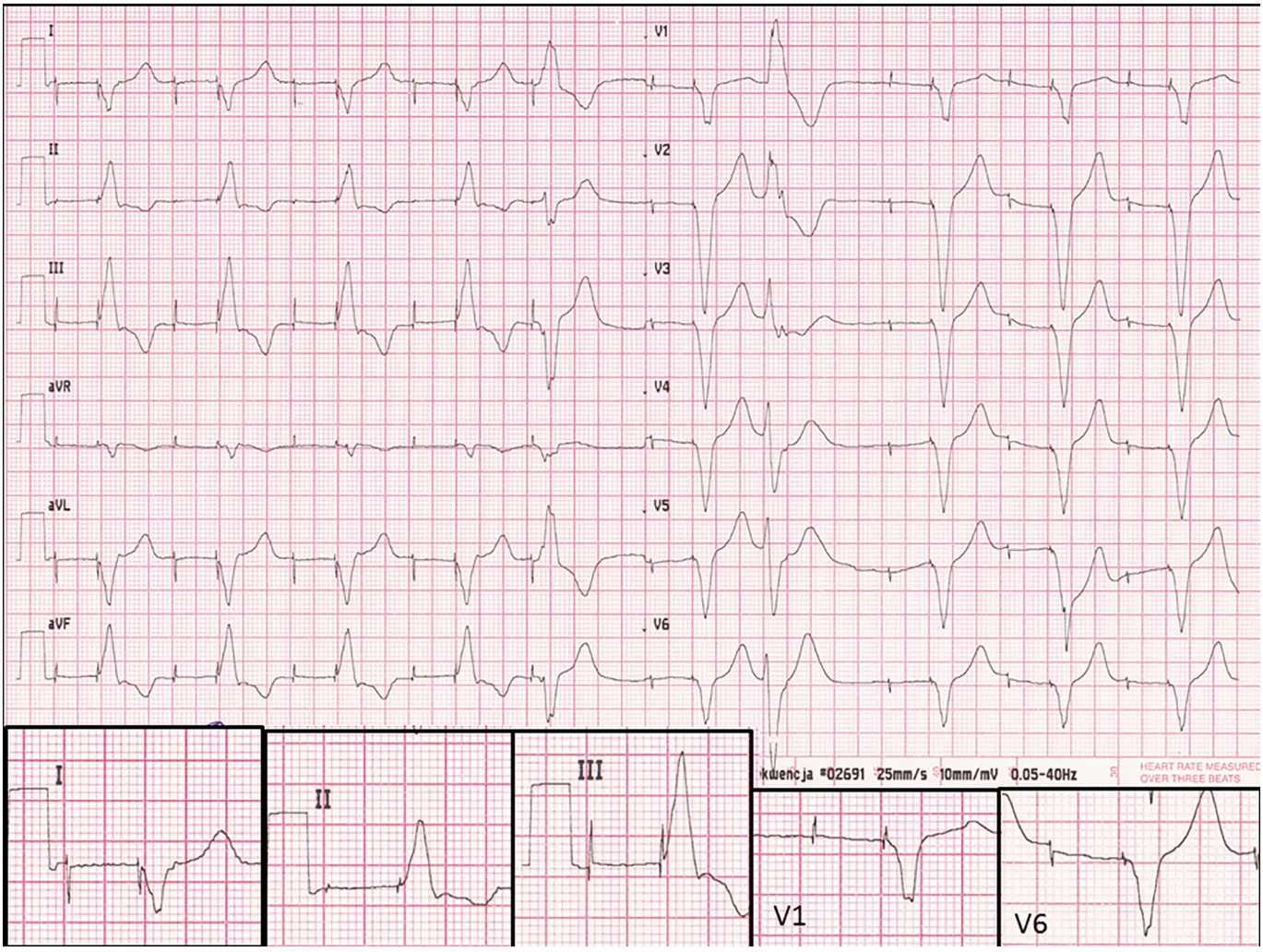

Fig. 1. ECG at admission suggestive of atrial lead failure to pace. Dual chamber Ap-Vp pacing no visible P waves. Pacemaker settings DDD 50 bpm, AVD 350 ms.

area of atrioventricular junction was not affected by fibrosis (Fig. 2). Conduction inside the right atrium, measured as a distance from atrial stimulus to A signal in His bundle region, was significantly prolonged (364 ms), while AH and HV intervals were within normal range (Fig. 3). We managed to identify few points of atrial capture around carotid sinus ostium with conduction to ventricles lasting below 200 ms. Taking into consideration progressive nature of scleromyositis, low LVEF and need of permanent ventricular pacing the pacemaker was upgraded to cardiac resynchronization device. Extensive fibrosis of enlarged right atrium was considered as a predictor of low probability of successful ablation for atrial arrhythmias. Therefore, due to lack of clear clinical indications left atrial catheterization and mapping were abandoned. During one year follow up control ambulatory visits documented 98% of biventricular pacing with improvement in heart failure symptoms and LVEF (47%).

Discussion

Scleromyositis is a rare mixed connective tissue disease. Patients with this overlap syndrome have significantly worse prognosis than those with systemic sclerosis (SSc). Cardiopulmonary involvement was reported at the most common cause of death in this population [5]. Cardiovascular complications in patients with scleromyositis are believed to be similar to those observed in patients with SSc. Microvascular disease, diastolic dysfunction, tachyarrhythmias and conduction disturbances are observed in over 60% of patients. Subclinical involvement due to progressive interstitial fibrosis related to inflammatory process is even higher. Heart failure due to diastolic and less frequently systolic dysfunction is major clinical consequence of myocardial fibrosis which may be complicated by arrhythmias and is associated with increased mortality. “Patchy” distribution of fibrosis is considered as a pathognomonic feature of SSc [6]. In our patient progressive heart failure could be related with natural history of a disease but unfavourable consequence of permanent ventricular pacing should be taken into account as well. In patients with heart failure, decreased LVEF and permanent pacing, CRT therapy can be considered as therapeutical approach [7]. In our case decision on upgrade from pacemaker to CRT therapy was based on progressive nature of scleromyositis, low LVEF and need of constant ventricular pacing due to conduction disturbances. Diffuse fibrosis is considered as one of the mechanism of conduction disturbances in patients with connective tissue disease. Due to pre-existing pacemaker we could not perform magnetic cardiac resonance in our patient. Nevertheless, severe fibrosis of atria reflected as large areas of low voltage was confirmed by electroanatomical mapping. Interestingly, first degree AVB visible in surface ECG in case of our patient was related to severe right atrial fibrosis documented by significantly prolonged conduction time within atrium. Atrioventricular junction was not affected by fibrosis and AV

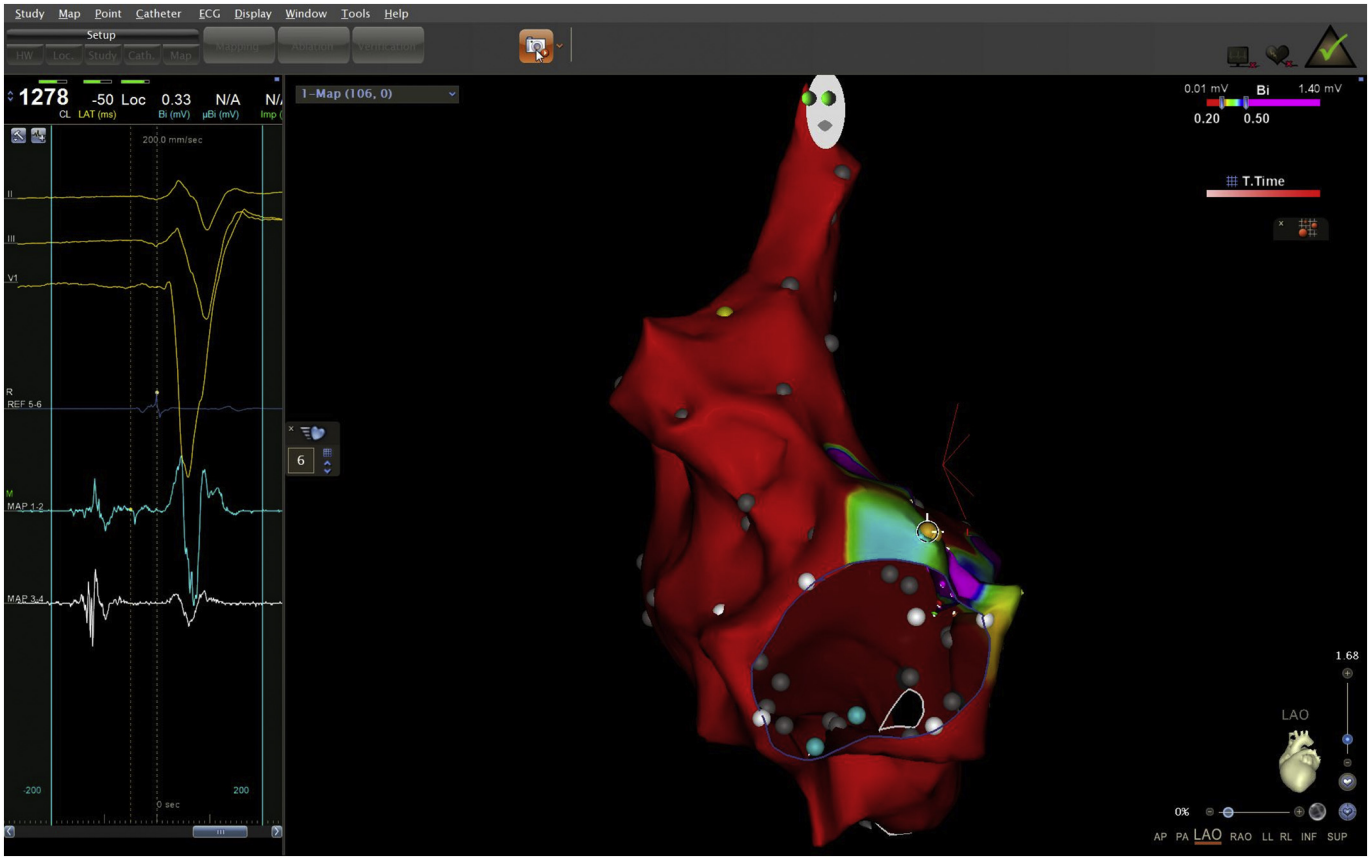

Fig. 2. 3-D electroanatomical map (CARTO 3D) illustrating extremely severe fibrosis of right atrium. Left Anterior Oblique projection.

conduction intervals were within normal range. The exact mechanism of this phenomenon is not clear. Unfortunately, we were not able to assess ventricular fibrosis by MRI due to previously implanted pacemaker. Left atrial mapping was abandoned due to lack of clinical indications.

Some authors reported congenital, genetically determined electrical disorders of atrial activity [8]. In our case it seems that connective tissue disease plays a major role. It is worth emphasizing that extensive fibrosis of atria may result in invisible paced P waves which in our opinion

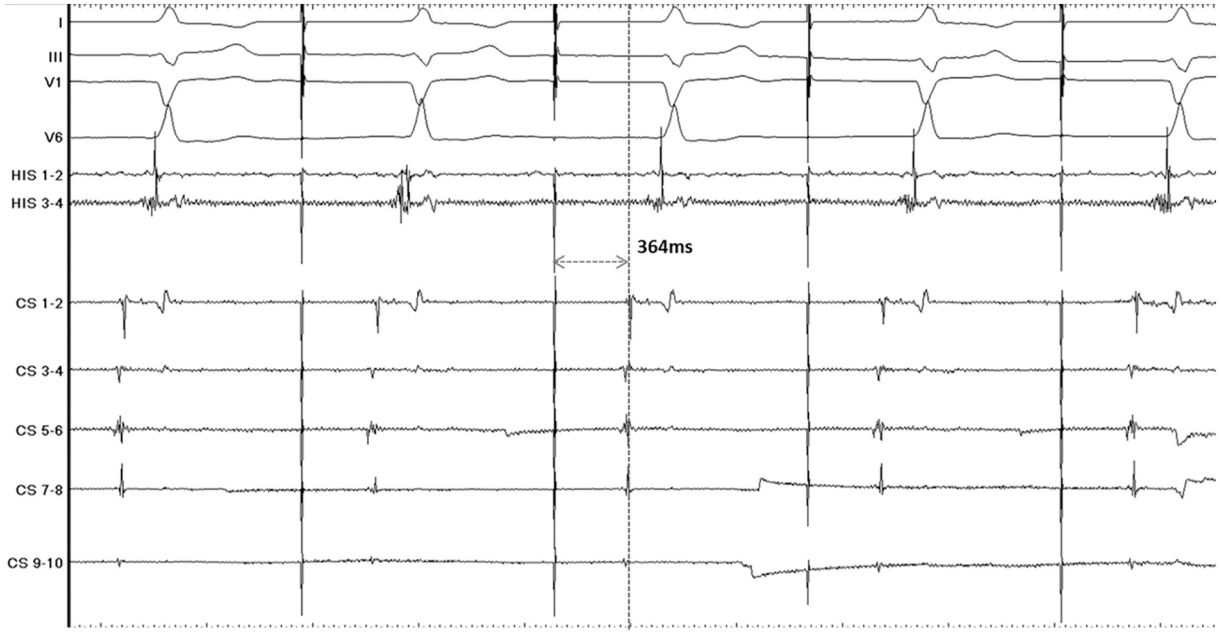

Fig. 3. Intracardiac electrograms. (CS 1–2 = His bundle), the interval from atrial stimulus to A signal in His bundle region is prolonged -364 ms.

was a reason of misdiagnosed atrial dysfunction at the time of the first reimplantation. This diagnostic problem affects not only pacemaker patients. Small baseline atrial activity seen in surface ECGs may be interpreted as atrial standstill, artifacts or low voltage atrial fibrillation/flutter.

Declarations of Competing Interest

None.

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