A Novel pacing option in patients with endomyocardial fibrosis: A case series

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A R T I C L E   I N F O

Article history:
Received 28 October 2020
Received in revised form 21 April 2021
Accepted 19 May 2021
Available online 24 May 2021

Keywords:
Endomyocardial fibrosis
Atrial flutter
Complete heart block
Epicardial pacing

A B S T R A C T

Endomyocardial fibrosis (EMF) is characterized by fibrous tissue deposition on the endocardial surface leading to impaired filling of one or both ventricles, resulting in either right or left heart failure or both [1]. Although Sinus node dysfunction and tachyarrhythmia - atrial fibrillation, ventricular tachycardia, have been commonly reported, complete heart block (CHB) necessitating a pacemaker is rare in EMF. Transvenous pacing is technically limited by fibrotic obliteration of the affected ventricle that results in poor lead parameters, and alternative pacing strategy like epicardial pacing may be required in many. We report three cases of EMF, who were treated with an alternative pacing strategy.

1. Introduction

Endomyocardial fibrosis (EMF) is characterized by fibrous tissue deposition on the endocardial surface leading to impaired filling of one or both ventricles, resulting in either right or left heart failure or both [1]. Although Sinus node dysfunction and tachyarrhythmia - atrial fibrillation, ventricular tachycardia, have been commonly reported, complete heart block (CHB) necessitating a pacemaker is rare in EMF [1]. Transvenous pacing is technically limited by fibrotic obliteration of the affected ventricle that results in poor lead parameters, and alternative pacing strategy like epicardial pacing may be required in many. We report three cases of EMF, who were treated with an alternative pacing strategy.

2. Case report

2.1. Case 1

A 75-year-old male presented with frequent syncopal episodes in the last 4 years, last episode being 20 days prior to the admission in our hospital. There was no history of angina or dyspnea on exertion. There was no history suggestive of any familial cardiac illness and sudden cardiac death.

On clinical examination, he was found to have a pulse rate of 40 bpm with a blood pressure of 112/70 mmHg. Cardiomegaly was present with grade 2/6 pansystolic murmur at apex. ECG showed features of atrial flutter with CHB, narrow QRS escape with a rate of 35–40 bpm, and incomplete right bundle branch block (Fig. 1A). Transthoracic echocardiography (TTE) shows dilated right atrium (RA), calcified and obliterated RV apex, partially obliterated RV body with normal RV inflow and outflow, mildly dilated RV outflow tract. Left ventricular (LV) angiogram showed irregularly shaped LV cavity and moderate diastolic mitral regurgitation.

A diagnosis of biventricular EMF with atrial flutter and CHB was made. The patient was planned for permanent pacemaker implantation. Lead parameters at multiple RV positions were suboptimal due to the obliterated RV body apex and body. Pacing threshold at RV outflow tract was better, but stability of the lead is a concern. Hence, coronary sinus (CS) was cannulated and LV active fixation lead (Attain Stability®, Model 4796-88 cm, Medtronic, France) was implanted in the left ventricle with suboptimal CS parameters. One month later, the pacing threshold had improved.
Minneapolis, MN, USA) was positioned at the lateral tributary of CS. Lead parameters (bipolar threshold 1.3 V @ 0.5 ms, impedance 662 Ohms, R wave 17 mV) were found to be satisfactory. Post-procedure fluoroscopy shows good lead position (Fig. 2, B-E). At 4 years of follow-up, patient remains symptom free with no worsening of the lead parameters.

2.2. Case 2

A 54-year-old male was admitted with a history of exertional dyspnea for 18 months. Clinical examination revealed a pulse rate of 40 bpm with a blood pressure of 160/70 mmHg and elevated JVP. Mild cardiomegaly was present with grade 2/6 pan systolic

**Fig. 1.** ECG showing (A) atrial flutter with complete heart block (B) Post pacemaker implantation.

**Fig. 2.** RV angiogram(A) in RAO view showing obliterated RV apex and body with calcification (arrow). The orange line represents the outline of the Right ventricle. Fluoroscopy (B–E) after pacemaker implantation showing lead position in various views.
murmur at apex, but patient was not in heart failure.

ECG (Fig. 3A) revealed atrial flutter with CHB and a ventricular rate of 40 bpm. Echocardiography showed dilated RA, obliterated RV apex, moderate TR and moderate mitral regurgitation. A 24-h Holter showed intermittent CHB.

RV angiography (Fig. 3C and D) shows involvement of the RV inflow with calcified and obliterated RV apex, dilated RV outflow and significant TR. A smooth endocardial border was visualized in the LV angiogram suggestive of LV involvement. A diagnosis of biventricular EMF with atrial flutter and intermittent CHB was made. Electrophysiological study showed high pacing threshold (6V at 0.4 ms) at the RV endocardium along with poor R wave sensing (0.6mV). Coronary sinus was cannulated and LV lead (Corox ProMRI OTW-S, Biotronik) was positioned at the middle cardiac vein distally, as there was no other good tributary. The parameters (LV bipolar vector: R wave-12mV, Threshold-1.8V at 0.5 ms, Impedance-650ohms) were found to be satisfactory. Post-procedure chest X-ray (Fig. 3 E and F) showed optimal lead position. Lead parameters remained stable at 4 years of follow up.

Fig. 3. ECG showing (A) atrial flutter with complete heart block (B) Post pacemaker implantation. RV angiogram in RAO view showing (C) endocardial calcification (arrows) (D) obliterated RV apex (asterisk). Chest X-ray after pacemaker implantation in (E) PA view (F) lateral view.
2.3. Case 3

A 66-year-old male, known case of biventricular EMF with severe TR and tachy-brady syndrome, underwent electrophysiological study in 2008 for paroxysmal palpitations and diagnosed as atypical atrial flutter. Patient was started on antiarrhythmics and rate lowering drugs. Now, he presented to the hospital with one episode of syncope 2 days before. On examination, he had a pulse rate of 94 bpm with a blood pressure of 126/84 mmHg. There were no other significant findings on cardiac examination.

ECG revealed atrial flutter with variable conduction and a ventricular rate of 100 bpm [Fig. 4A]. Echocardiography showed good LV function, moderate RV dysfunction, hugely dilated RA, obliterated RV apex, calcification and fibrotic infiltration of the RV endocardium and moderate TR. Holter showed a maximum pause of 3.3 seconds.

RV angiogram (Fig. 4C) showed obliterated and calcified RV apex, RV outflow tract calcification, dilated RA, severe TR and mild RV dysfunction. LV angiogram showed balding of LV apical segments, moderate MR and good LV function [Fig. 4D]. Cardiac MRI showed calcified and obliterated RV apex. Thickening of LV apex with delayed gadolinium enhancement was seen, suggestive of LV EMF [Fig. 4B].

Patient was taken up for the Electrophysiological study which revealed significant scarring of the inflow and body of the RV along with elevated capture thresholds while that of the RV outflow was acceptable. The above and the progressive nature of the disease, it was decided to implant an epicardial LV lead through the coronary sinus. Coronary sinus was cannulated and LV lead (Medtronic Attain Stability MRI SureScan, 4796-88cm) was positioned at the posterolateral vein distally. Lead parameters were found to be satisfactory (R wave 9 mV, Impedance-989 ohms, Threshold 1V at 0.5 ms) Post-procedure chest X-ray (Fig. 4E) showed optimal lead position. At 4 months of follow up, patient remains stable with normal lead parameters.

3. Discussion

Obliteration of the apices of the affected ventricles due to fibrosis is the hallmark of EMF. Endocardial calcification marks the burnt-out phase of EMF [2]. Surgery for EMF is considered only when the patient develops heart failure, as it does not alter the progressive nature of the disease. Since none of the above-described patients had heart failure, surgery was not a first option. Atrial fibrillation has been reported in more than 30% of patients with EMF. Though conduction abnormalities are common, pacing is rarely required in such patients [1]. Atrophicventricular block is not commonly seen in EMF. Earlier attempts at pacing in EMF has been limited to case reports. One case series describes 3 patients with RV EMF and CHB who required pacing. 2 patients had epicardial pacing after failure to obtain stable pacing in the RV due to lack of trabeculae. Third patient had endocardial active fixation lead with acceptable pacing threshold [3].

The etiology of heart block in EMF has been much debated. Subendocardial fibrosis due to the inflammatory reaction may affect the conduction system traversing within which could be the reason for CHB.

The first two described patients had atrial arrhythmia and CHB, last patient had tachy-brady syndrome with significant sinus pause in Holter, which demanded to pace. Atrial arrhythmia in RV restrictive physiology suggested significant underlying RA fibrosis, hence VVI was the only viable option and atrial-based pacemakers like AAI or DDD were not appropriate in these cases. For conventional RV endocardial pacing, obliteration of the RV cavity due to EMF is a major concern. In these patients, the capture threshold is likely to be high, and moreover, local sensing of R wave amplitude is likely to be unacceptably low. In addition, loss of trabeculations in RV would not allow a passive lead fixation. Tricuspid regurgitation can also lead on to lead instability. Large RA and likely septal involvement on the endocardial aspect of the RV side could be a concern for conduction system (left bundle or His bundle) pacing. Epicardial lead placement is an option; however, the associated heart failure commonly seen in patients with EMF makes surgical epicardial approach a risky procedure. Transvenous pacing through coronary sinus tributaries offer many advantages in this case.

There is ample experience with the LV pacing, and long-term stability of the CS lead has already been established [4]. In this approach, the endovascular lead not crossing the tricuspid valve gives an advantage of not worsening the pre-existing TR. Dilated CS tributaries make procedure relatively easier in these cases. However, the long-term issue with CS lead includes diaphragmatic stimulation, CS dissection, lead dislodgement and elevated pacing threshold. Long term stability may demand anchoring of the lead in a large tributary and the use of an active fixation lead a safer option as we did in the first case.

4. Conclusion

EMF provides unique challenges to endocardial pacing.
Transvenous epicardial pacing through CS tributary using an LV lead can provide a safe and effective alternative mode of pacing with optimal long-term pacing outcome.

**Financial support**

None.

**Declaration of interest**

There are no conflicts of interest to declare.

**Disclosures**

The authors have no competing interests, funding or financial relationships to disclose.

**Declaration of competing interest**

This manuscript is not under simultaneous consideration elsewhere and has not been previously published in similar form. There is no source of funding or conflicts of interest.

**Acknowledgment**

None.

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