Challenges in pacing in right ventricular endomyocardial fibrosis: A case study

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Introduction

Endomyocardial fibrosis (EMF) is one of the most common forms of restrictive cardiomyopathy worldwide.[1] It is highly prevalent in regions close to the equator and impoverished areas of sub-Saharan Africa.[2] Additionally, its incidence rate is notably higher in Kerala, India.[3] EMF is characterized by apical filling with fibrotic tissue of 1 or both ventricles; it is often associated with thrombus and calcification.[4] Calcification denotes a burnt-out phase of EMF and confirms the malignant nature of the disease.[5]

EMF is often associated with rhythm abnormalities, with atrial fibrillation being the most frequent. Bradyarrhythmias are prevalent in elderly individuals,[6] and subendocardial fibrosis caused by inflammation may impact the conduction system, potentially leading to complete heart block.[7]

KEY TEACHING POINTS

- Endomyocardial fibrosis is sometimes associated with conduction abnormalities and calcification.
- Calcification in endomyocardial fibrosis makes fixation of pacemaker lead challenging.
- Options in severe calcific endomyocardial fibrosis are left ventricular pacing through the posterolateral, anterior interventricular, and middle cardiac veins; another available option is epicardial pacing.
- Surgical epicardial pacing can be challenging owing to right ventricular failure and epicardial or pericardial calcification.

Case report

A 63-year-old woman with type 2 diabetes mellitus experienced presyncope and 1 episode of syncope. Her electrocardiogram revealed a long sinus pause of approximately 3 seconds, followed by atrial fibrillation with a fast heart rate (Figure 1). The diagnosis was tachy-brady syndrome and she was taken up for emergency temporary pacemaker insertion. On fluoroscopy, there was calcification in the right ventricle, and despite multiple attempts, placing the pacemaker lead in the right ventricular apex was not possible. Although the lead position was unstable, the lead was placed in a proximal portion of the right ventricle just beneath the tricuspid valve. Electrical parameters were not favorable; the threshold was 5 volts, the sensitivity was 2 millivolts, but as a last option output was kept at 8 volts for safety margin and pacing was confirmed.

Once stabilized, she was evaluated, and her echocardiography showed dilated right ventricle and right atrium with calcific obliteration of the right ventricular apex and calcific moderator band (Figure 2), suggestive of right ventricular EMF. Her cardiac computed tomography showed a calcific right ventricular apex with a calcific septum extending to the right ventricular outflow tract (Figure 2).

The patient improved symptomatically, but there was an intermittent loss of capture the next day, and she was taken up for permanent pacemaker implantation. Initially, pacing was tried with the right ventricular lead, but the lead could not be screwed into the right ventricle apical septum. The next option was to place the lead in the higher septum for conduction system pacing. Higher septal pacing was tried with a 3830 (69 cm) left bundle lead (Medtronic Inc, Minneapolis, MN) with the help of nondeflectable C315HIS sheath (Medtronic Inc), but unfortunately the lead could not be screwed owing to calcification, and even when screwed, the threshold was more than 8 volts. Multiple positions in the right ventricular septum and right ventricular outflow tract were tried, but they were not favorable.

After careful consideration, left ventricular pacing via coronary sinus was planned. Using an MB2 catheter (Medtronic Inc), coronary sinus angiogram revealed a very small posterolateral vein (Figure 3), which precluded lead placement in the

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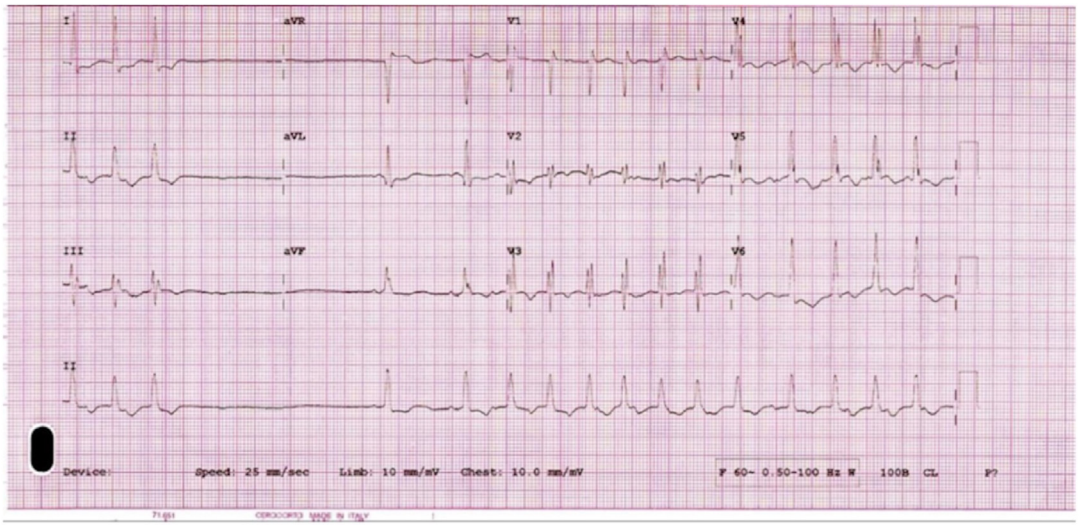

Figure 1 Electrocardiogram shows a long pause followed by atrial fibrillation with a fast ventricular rate.

posterolateral vein. As a last resort, the 4296 (88 cm) bipolar left ventricular lead (Medtronic Inc) was positioned in the superior branch of the middle cardiac vein (Figure 3). The threshold was 0.5 volts at 0.4 milliseconds. However, there was diaphragmatic stimulation at 2.5 volts at 0.4 milliseconds. Hence, the sensed atrioventricular delay was set at 350 milliseconds, which reduced the need for ventricular pacing. The patient was discharged on the following day.

Discussion

Endomyocardial fibrosis is estimated to be the most common and progressive form of restrictive cardiomyopathy worldwide. Jack N. P. Davies described EMF in Uganda. Also known as Davies disease, it is mainly a tropical disease.[1] The right ventricle is the cardiac chamber most frequently affected in EMF, either in isolation or as part of bilateral disease.[2] Apical filling with fibrotic tissue of 1 or both ventricles is the hallmark of EMF, often associated with thrombus, calcification, and atrioventricular valve regurgitation, leading to typical symptoms of restrictive heart failure.[4]

EMF has an insidious onset and an indolent course, and most often affects children and young adults.[3] There is a bimodal incidence peak for EMF at 10 years and 30 years of age in population studies performed in sub-Saharan Africa, without a clearly defined sex predilection.[4] In a study by Muthian and colleagues,[8] the oldest female participant was 85 years old and had mushroom-shaped apical fibrosis in the ventricles in an echocardiogram. However, the youngest participant was only 10 years old.[8] In the 14 years from 2007 to 2020, the Uganda Heart Institute noted declining trends in the incidence rates of EMF.[9] Cases at Uganda Heart Institute from January 2007 to December 2020 were divided into 2 groups, group A (2007–2013) and group B (2014–2020), representing equal time periods. Comparison of the 2 groups showed no notable disparities in age at diagnosis, sex distribution, and EMF phenotype. However, there was a trend toward older mean and median age of patients over the course of the years compared to that at the beginning of the study period.[9]

EMF is usually seen in areas within 15 degrees of the equatorial belt.[1] Its highest prevalence is in poor regions of

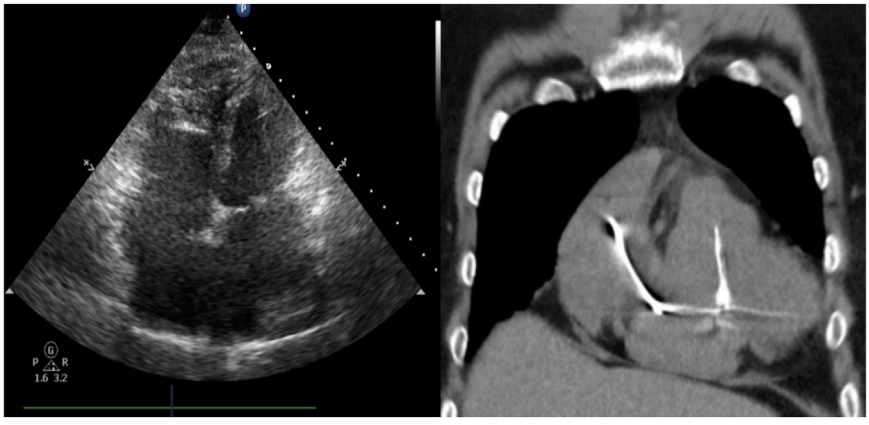

Figure 2 Echocardiography image on left side showing dilated right ventricle and right atrium with calcific apical obliteration and cardiac computed tomography image on right side showing calcification in right ventricle extending to right ventricular outflow tract.

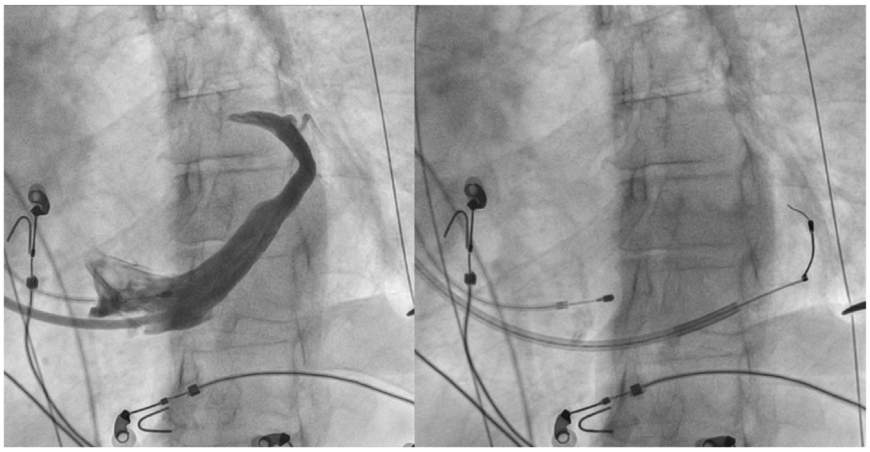

Figure 3 Angiogram of coronary sinus in left anterior oblique 30, caudal 0 view showing very small posterolateral vein; image on right showing left ventricular bipolar lead placed in superior branch of middle cardiac vein.

sub-Saharan Africa, where it is generally found in children and adolescents.[2]

Several hypotheses exist regarding the etiopathogenesis of EMF, indicating the role of filarial infections, helminths, eosinophilia, and malnutrition. However, the geographical distribution of EMF is not related to the prevalence of filariasis and eosinophilia. The endemic variety of EMF can be related to high cerium and low magnesium levels. The coexistence of high incidence of EMF and deposits of monazite element cerium in the soil in 2 coastal talukas in Kollam and Alappuzha districts in south Kerala in India support the geochemical hypothesis. EMF is postulated to be related to prolonged tapioca (tuber) ingestion, accumulating cerium from the surrounding sand. Food contaminated by sand could also lead to cerium accumulation in the body.[3]

In 1967, Cockshott and colleagues[5] described the presence of diffuse calcific endocardial deposits in the ventricles in EMF. Calcification denotes a burnt-out phase of EMF and confirms the malignant nature of the disease. The inflammatory response occurring in the younger age group could manifest as calcification later. The presence of interstitial fibrosis, myohypertrophy, and calcification suggest the role of cytokines in its genesis.[7] Delayed-enhancement imaging shows the typical pattern is the "double V" sign, consisting of a 3-layered pattern of normal myocardium, thickened enhanced endomyocardium, and an overlying thrombus at the ventricular apex, with or without calcifications.[4]

Ventricular stiffness and atrioventricular valvular regurgitation results in atrial enlargement, resulting in atrial arrhythmias such as atrial fibrillation. Fibrosis provides a substrate for wave and break and reentry by reducing conduction velocity and impairing activation pattern.[8] According to reports, atrial fibrillation is the predominant arrhythmia in EMF, accounting for 30% of all cases, followed by junctional rhythm, heart blocks, and intraventricular conduction delay.[8] Although EMF can result in sinus node dysfunction and tachyarrhythmias, such as atrial fibrillation and ventricular tachycardia, complete heart block necessitating a pacemaker is rare.[6] However, among patients with EMF who experienced sudden cardiac death, 15%–20% had documented advanced atrioventricular block or asystole.[10]

There have been significant debate and conflicting views regarding the etiology of heart block in EMF.[6] The incidence of cardiac arrhythmias increases with advancing age. In the United States, the median age of individuals receiving pacemakers for bradyarrhythmias is 75 years, with over 80% of pacemaker recipients being 65 years or older. Owing to reduced normal automaticity and delayed conduction, bradyarrhythmias are common in older adults, even without apparent heart disease, and are further exacerbated by comorbidities or the use of medications resulting in symptoms that require pacemaker implantation.[10] Subendocardial fibrosis owing to the inflammatory reaction may affect the conduction system traversing within, which could be the reason for complete heart block.

Fibrosis in the targeted ventricle often poses a challenge for transvenous pacing, as it can result in poor lead parameters. Moreover, it is challenging to secure the lead passively if the trabeculations in the right ventricle are lost. Furthermore, tricuspid regurgitation can lead to instability of the lead. In these patients, the capture threshold is likely to be high, and local sensing of R-wave amplitude is unacceptably low. Large right atrium and likely septal involvement on the endocardial aspect of the right ventricular side could be a concern for conduction system pacing.[6]

In our report, we attempted to insert the pacemaker lead into the right ventricular apex and tried conduction system pacing, but unfortunately, both were unsuccessful. We also encountered difficulties in placing the lead in the posterolateral vein owing to its small size. Eventually, the lead was placed in the superior branch of the middle cardiac vein, which led to diaphragmatic stimulation. Despite adjusting the electrical parameters, the patient continued to experience diaphragmatic stimulation. It is worth noting that her current electrocardiogram shows a first-degree heart block, which may require ventricular pacing in the future. In situations like these, alternative pacing strategies, such as epicardial pacing, may be necessary. Although epicardial lead

placement is a possible solution, it is important to note that patients with EMF often have heart failure, and possible epicardial or pericardial calcification makes the surgical epicardial approach a difficult option. Placing the lead in the anterior interventricular vein is an alternative option for coronary sinus lead. However, there was an easily accessible middle cardiac vein, which would pace on the left ventricular epicardium. Here, the middle cardiac vein looked relatively stable, with consistent electrical parameters.

Conclusion

Endomyocardial fibrosis is one of the most common forms of restrictive cardiomyopathy. Arrhythmias are common in EMF, but complete heart block requiring a pacemaker is rare. EMF can present technical difficulties when placing the right ventricular lead, particularly in cases of fibrotic obliteration of the apex and calcification. Physiologic pacing may also prove challenging in such situations owing to calcification. Since there was no suitable posterolateral vein, the lead was placed in the middle cardiac vein, which caused diaphragmatic stimulation. Epicardial pacing is an option, but this comes with challenges owing to right ventricular failure and calcification in the epicardium or myocardium.

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