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Severe left ventricular systolic dysfunction after permanent pacemaker implantation: Should we pause before upgrading to biventricular pacing?

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ABSTRACT

Left ventricular (LV) systolic dysfunction leading to heart failure (HF) is known to occur after permanent pacemaker implantation (PPI) in a subset of patients. They are often treated by upgradation of the pacemaker to cardiac resynchronisation therapy (CRT). We report a case of progressive LV dysfunction and HF after PPI. Cardiac ^{18}FDG-PET-CT scan revealed abnormal myocardial FDG uptake suggestive of cardiac sarcoidosis (CS). Biopsy from FDG avid lymph node demonstrated non-caseating granuloma. Therapy with steroids resulted in resolution of HF symptoms accompanied by a significant improvement in LV function.

1. Introduction

Following PPI approximately 9% of patients develop significant LV dysfunction [1]. Right Ventricular (RV) pacing-induced cardiomyopathy (PICM) is defined as a 10% or more reduction in the LV ejection fraction (EF) after PPI, resulting in an EF of less than 45% [1,2]. Usually, new onset HF and LV dysfunction after implantation of a pacemaker is attributed to RV pacing induced dyssynchrony. In most instances, coronary angiogram is performed to exclude coronary artery disease and the device is upgraded to CRT. Majority improve following this intervention but approximately one-third remain in persistent HF [2].

Varying degree of LV dysfunction or AV block can be the initial presentation of occult CS [3,4]. It is a progressive disease and what presented as AV block may be followed by LV systolic dysfunction due to chronic inflammatory myocarditis in due course [5]. Unrecognized, such patients usually have upgradation to cardiac resynchronisation therapy (CRT) and some eventually have cardiac transplantation. Studies have shown that, worsening LV function and ventricular arrhythmia (VA) are preventable and might be reversed, if CS is treated early [6].

We report a case which illustrates that delay in diagnosis of CS in a patient with AV block resulted in severe LV dysfunction due to disease progression. Appropriate initiation of immunosuppression led in improvement of LV function and symptoms.

2. Case report

A 61 year-old gentleman underwent PPI for symptomatic AV block [complete heart block with a wide QRS [right bundle branch block (RBBB) morphology escape] [Fig. 1A]. He had undergone aortic valve replacement (AVR) for severe calcific aortic stenosis 3 years back. He had normal LV systolic function on echocardiography (LVEF = 56%) during PPI.

Six months after PPI, he presented with progressive shortness of breath and HF (NYHA class IV). He had bilateral pedal oedema, elevated jugular venous pressure; with bi-basal crepitations. NT-pro BNP was elevated (1432 pg/ml). Echocardiography revealed severe LV dysfunction (LVEF had dropped to 37%) with increase in LV dimensions {LV internal diameter in diastole (LVIDd) 48→63 mm, LV internal diameter in systole (LVIDs) 40→52mm}, grade III

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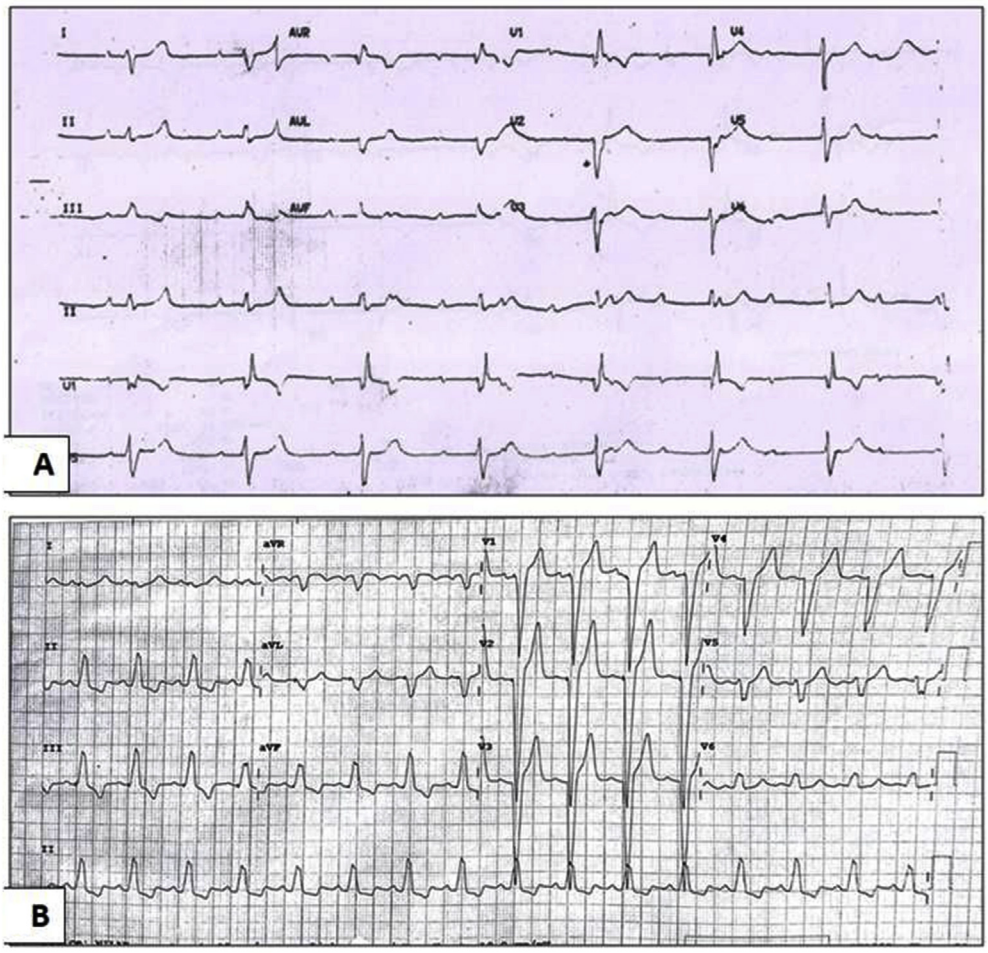

Fig. 1. A- ECG showing complete heart block and wide complex (RBBB) escape beats. 1B- Atrial sense -Ventricular paced rhythm.

mitral regurgitation (MR) and elevated pulmonary artery systolic pressure (PASP- 47 mm Hg). Device interrogation and electrocardiography (ECG) suggested 100% V pacing (As - Vp: 96%, Ap -Vp 4%) [Fig. 1B]. Medical therapy was optimized with adequate doses of angiotensin converting enzyme (ACE) inhibitor (ACEI), beta-blocker and mineralocorticoid receptor antagonist. He continued to remain symptomatic. He was considered for upgradation of device to biventricular pacing.

Over next one month, the LV function deteriorated further (LVIDd- 70 mm, LVIDs- 57mm, LVEF 32%, PASP- 60 mm Hg). Such rapid deterioration of LV function over a short period (of 4 months) was unusual. Few cervical and axillary lymph nodes were palpable on physical examination. His ESR and hs-CRP were elevated. ESR was 78 mm/1st hour (haemoglobin was normal) and CRP was 60.6 mg/dL at the baseline. This was suggestive of something *beyond RV pacing-induced cardiomyopathy*. A cardiac ^{18}FDG PET-CT scan was performed considering possibility of an inflammatory cardiomyopathy. It revealed significant abnormal myocardial (SUV max 8.1) as well as cervical, axillary and mediastinal lymph node FDG uptake (SUV max 7.5) highly suggestive of inflammatory cardiomyopathy (Fig. 2A,B,C). The lymph node (LN) biopsy revealed non-caseating granuloma. Mantoux skin test, TB (Tuberculosis)-PCR and TB culture from the biopsy samples were negative. A diagnosis of cardiac sarcoidosis was made and he was started on oral steroid. His functional status improved to NYHA class II within 3 weeks of starting immunosuppression. Repeat PET-CT scan after 4 months showed significant reduction in FDG uptake (SUV max 2.0 in pretracheal LNs, no myocardial uptake). After 6 months of steroid therapy, his HF symptoms had completely resolved and LV function had improved (EF 50%). [Table 1].

At 1-year follow-up, he was asymptomatic, along with a near complete recovery of LV function (LVEF = 54%) and reduction of MR to grade I (without CRT). The third PET scan at 1 year showed completely absent FDG uptake (Fig. 2D,E,F).

3. Discussion

Sarcoidosis is a multisystem disease of unknown aetiology. Among patients with sarcoidosis, 5% have overt cardiac involvement, whereas additional 20% patients have unrecognized cardiac involvement [7]. Cardiac tuberculosis (CTB) is a close mimicker of CS and at times, it is difficult to differentiate these two entities [8,9]. The common presentations of CS/CTB is VA, LV dysfunction and AV block. They can present in isolation or in combination [8–10]. With different manifestations in the same patient, there may be a temporal separation among them like AV block preceding LV dysfunction or vice versa [4]. As they usually do not have systemic symptoms, the underlying granulomatous myocarditis (GM) is often missed and the disease keeps smoldering.

Our case highlights that a subset of patients who develop HF symptoms and/or LV dysfunction after PPI, may actually have underlying GM/CS even though RV-PICM is the commonest cause for

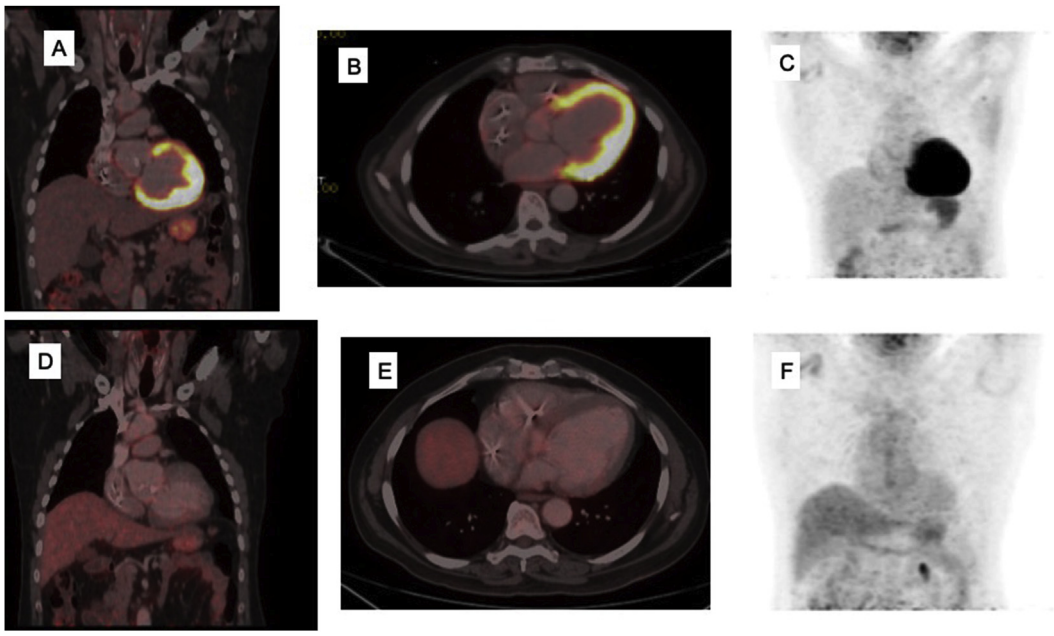

Fig. 2. A, B, C- Significant myocardial and lymph node uptake at presentation. D, E, F- Complete resolution of FDG uptake after 1 year.

Table 1
Showing the serial echocardiographic parameters.

Timeline	LVIDd (mm)	LVIDs (mm)	LVEF (Modified Simpsons) (%)	Mitral regurgitation grade	PASP (mm of Hg)
At pacemaker Implantation	48	40	56	I	30
After 4 months of PPI	63	52	37	III	47
After another 1 month (When steroid was finally started)	70	57	32	II	60
After 6 Months of starting steroid	53	43	50	I	32

the same. Early recognition and treatment of underlying CS can prevent progressive LV dysfunction, reduce morbidity and mortality as well as prevent unnecessary device upgradation or cardiac transplantation. Some authors [5,11,12] have also reported similar cases, where underlying CS rather than PICM which was responsible for progressive LV dysfunction. It is indeed difficult to suspect CS among patients diagnosed of PICM as majority of do not have systemic involvement. Detailed clinical examination (like enlarged lymph nodes) and monitoring acute phase reactants can provide clues to the underlying aetiology.

Conflicts of interest

None.

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