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Rapid communication

Bidirectional ventricular tachycardia in cardiac sarcoidosis ☆

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ABSTRACT

A 73-year-old man with history of pulmonary sarcoidosis was found to have runs of non-sustained bidirectional ventricular tachycardia (BVT) with two different QRS morphologies on a Holter monitor. Cardiac magnetic resonance delayed gadolinium imaging revealed a region of patchy mid-myocardial enhancement within the left ventricular basal inferolateral myocardium. An 18-fluorodeoxyglucose positron emission tomography (FDG-PET) showed increased uptake in the same area, consistent with active sarcoid, with no septal involvement. Follow-up FDG-PET one year later showed disease progression with new septal involvement. Cardiac sarcoidosis, characterized by myocardial inflammation and interstitial fibrosis that can lead to conduction system disturbance and macro re-entrant arrhythmias, should be considered in differential diagnosis of BVT. BVT may indicate septal involvement with sarcoidosis before the lesions are large enough to be detected radiologically.

1. Introduction

Bidirectional ventricular tachycardia (BVT) is defined as a tachycardia showing beat-to-beat alternation in the QRS axis. The rate is typically between 140 and 180 bpm, with a frontal plane axis varying between –20° and 110°. The most common causes of BVT include catecholaminergic polymorphic ventricular tachycardia and cardiac glycoside toxicity. Other previously described etiologies include myocarditis, long QT syndrome type 7, congenital cardiomyopathies, cardiac tumors, and acute cardiac allograft rejection. Cardiac sarcoidosis is characterized by myocardial inflammation and interstitial fibrosis that can lead to slowed conduction and macro re-entrant arrhythmias. We report a case of BVT in a patient with cardiac sarcoidosis and briefly discuss the proposed mechanisms underlying BVT.

2. Case

A 73-year-old man with history of chronic pulmonary sarcoidosis was seen for an annual checkup, during which ventricular bigeminy was identified on a 12-lead electrocardiogram. Subsequent Holter monitor assessment showed multiple premature ventricular beats and several short runs of non-sustained ventricular tachycardia (VT), with two different QRS morphologies (Fig. 1). A cardiac magnetic resonance tomography with delayed gadolinium showed a curvilinear region of patchy mid-myocardial enhancement within the inferolateral left ventricular myocardium near the base, consistent with cardiac sarcoidosis (Fig. 2). The left and right ventricular ejection fractions were 47% and 37%, respectively. A nuclear myocardial perfusion study using single-photon emission computed tomography showed no myocardial perfusion defects on stress or rest imaging, ruling out ischemia. A fasting 18-fluorodeoxyglucose positron emission tomography (PET) demonstrated increased uptake in the same area of the myocardium that had shown late gadolinium enhancement, consistent with active cardiac sarcoid (Fig. 3A). A one-year follow-up PET showed disease progression, with new septal involvement

Abbreviations: BVT, Bidirectional ventricular tachycardia; ICD, Implantable cardioverter defibrillator; PET, Positron emission tomography; PVC, Premature ventricular contraction; VT, Ventricular tachycardia.

☆N. B. The patient was treated at the University of Wisconsin Hospital. Dr Scheinman was consulted as an expert in the field and has reviewed and modified the submission.

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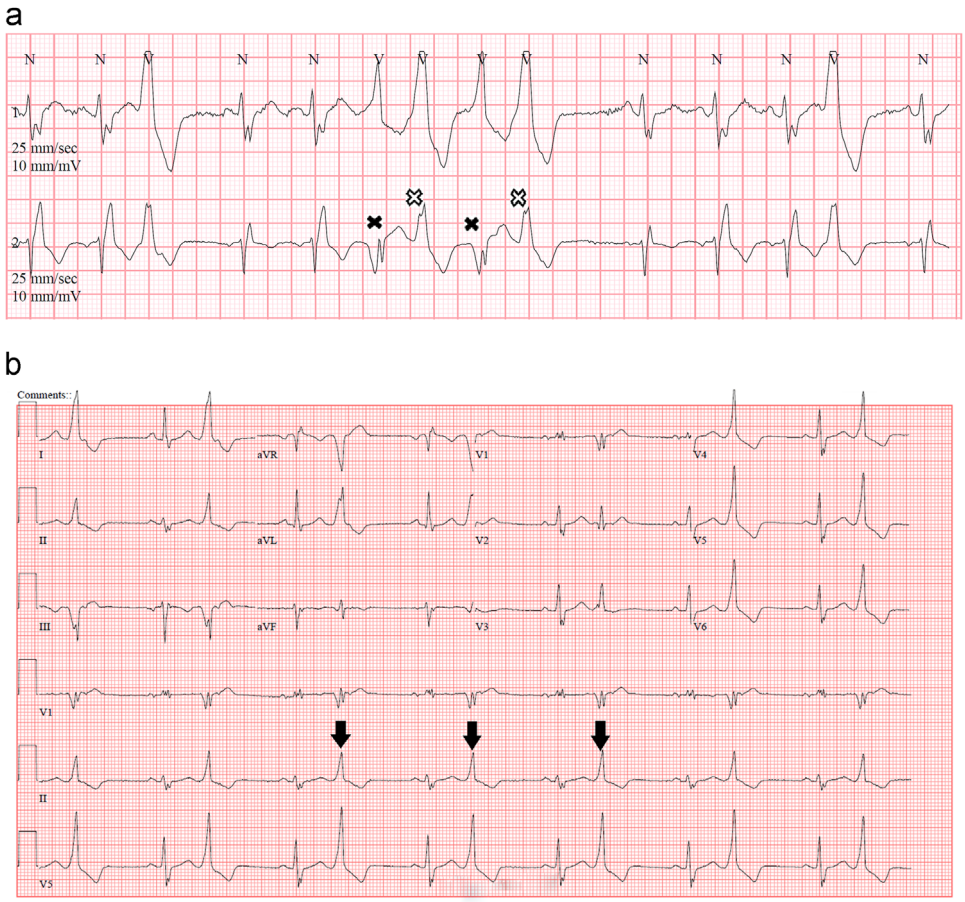

Fig. 1. (A) Rhythm strip with leads II and V (nonspecific ventricular), top and bottom, respectively, showing sinus rhythm and a run of ectopic ventricular beats with two alternating QRS morphologies; the sixth and eighth beats show a left bundle branch block morphology and inferior axis (black X), while the seventh and ninth show a right bundle branch block morphology and inferior axis (white X). This pattern was seen repeatedly on ambulatory monitoring. Note that the degree of the right bundle branch block increases during sinus rhythm; the QRS width in the fourth beat is about 120 ms, compared to approximately 160 ms in the fifth beat. A similar phenomenon is also seen between the tenth and eleventh beats even during a relatively long atrial cycle length of 600 ms. This suggests severe impairment of the conduction system. (B) A 12-lead electrocardiogram showing sinus rhythm with intraventricular conduction delay, with PVCs in a pattern of ventricular bigeminy. The PVCs resembles a left bundle branch block pattern with an inferior axis (arrows).

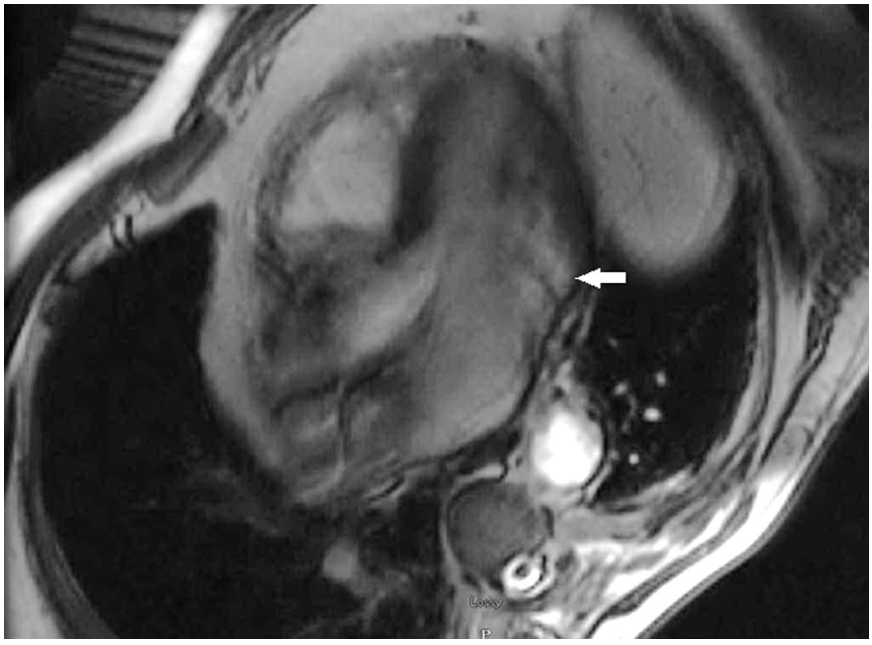

Fig. 2. Delayed enhancement cardiac magnetic resonance tomography with patchy mid-myocardial enhancement in the basal inferolateral left ventricular myocardium (arrow).

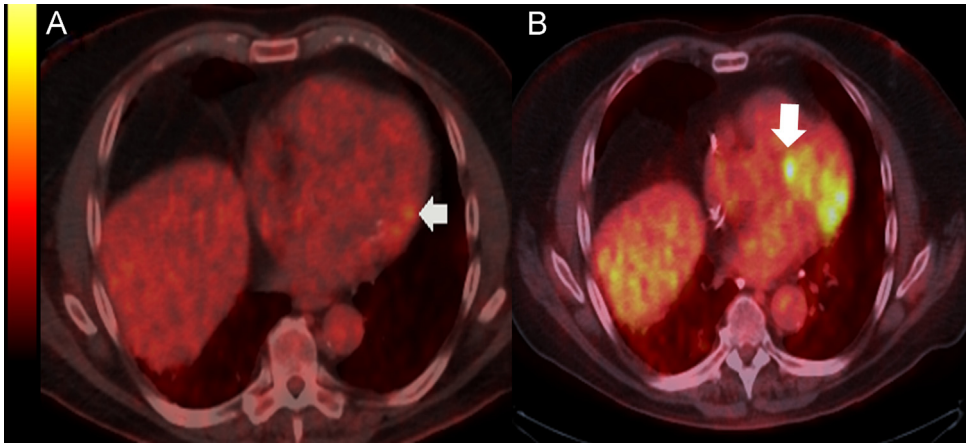

Fig. 3. Fasting 18-fluorodeoxyglucose (FDG) positron emission tomography at baseline (A) showing increased uptake, most pronounced in the basal inferolateral area of the myocardium (arrow), consistent with active cardiac sarcoid, and at follow-up (B), showing increased disease progress with FDG uptake in the basal to mid segments of the anterolateral and basal lateral wall segments as well as new FDG uptake in the basal anteroseptal region.

(Fig. 3B). The patient was started on immunosuppression therapy consisting of prednisone and mycophenolate. The patient was elected to have an implantable cardioverter defibrillator (ICD) placed for prevention of sudden cardiac death. He had not experienced any VT episodes during the one-year follow-up.

3. Discussion

We describe an additional diagnosis to consider when BVT is identified. One of the proposed mechanisms of BVT include elevated intracellular calcium, causing delay after depolarization in anatomically separate parts of the conducting system. Baher et al. [1] proposed that two separate foci, with different rate thresholds for delayed after depolarization–induced ventricular bigeminy, were present in a rabbit model. When the ventricular rate exceeded the lower threshold, bigeminy would develop. This would effectively double the heart rate, increasing the overall ventricular rate above the second threshold. Once this had developed, the two competing sites would simply alternate on a beat-to-beat basis. This is likely the mechanism underlying BVT observed with digitalis toxicity and catecholaminergic polymorphic VT. The other proposed mechanisms for BVT include an alternating bundle branch block related to bifocal automaticity and inscribed in opposite directions, or scar-mediated reentry around a circuit with two alternating exit sites. This latter is likely the underlying mechanism in our case. More recently, Sung et al. described a mechanism including retrograde conduction over the mid septal fascicular pathway, with alternating block in the left anterior or posterior fascicles, to explain polymorphic fascicular VT patterns [2]. Cardiac sarcoidosis causes ventricular inflammation and scarring due to focal non-caseating granulomas. Scarring is typically patchy, with a predilection for the basal septum, anterior wall, and perivalvular regions of the left ventricle. It may also be confluent, affecting the right ventricular epicardium or endocardium. In sarcoidosis, it is plausible that multiform or bidirectional premature ventricular contractions (PVCs) are due to multiple exits from the areas of inflammation and/or scarring. Conduction disturbances in cardiac sarcoidosis are not uncommon, and often affect the His-Purkinje system [3]. In our case, the patient had an intraventricular conduction delay at baseline, with prolonged QRS complexes at relatively low atrial rates (Fig. 1A). The majority of the retrospective data suggest that immunosuppression reduces the burden of arrhythmias, especially in the early phases of the disease [4].

Previous case series have reported various success rates with VT ablation in patients with cardiac sarcoidosis, likely due to the small number of patients in each series, with varying degrees of disease burden. The most common circuit for VT in one report was reentry in the peritricuspid area, which can be safely ablated [5]. In another study, abolishing all inducible tachycardias was not always feasible because of septal intramural circuits, extensive right ventricular scarring, or sites of origin in close proximity to the left anterior descending, the ramus intermedius arteries, or the para-Hisian region, which prohibit safe ablation [6]. In the current case, it is likely that the presence of a septal focus leads to alternate exits into the right or left ventricle which shows the observed electrocardiogram pattern. Although the septal involvement was not visible by imaging until a year after diagnosis, it might have been present initially, but was microscopic in nature. This is consistent with previous autopsy studies that have shown a heterogeneous distribution of sarcoid granulomas in the myocardium [7]. The recent expert consensus document is an excellent resource for management and risk stratification for cardiac sarcoidosis patients [8]. ICD implantation for primary prevention is commonly performed due to the high burden of VT events (estimated incidence rate of 15% per year) in patients with cardiac sarcoidosis [9]. In the current case, ICD implantation was recommended due to the high burden of nonsustained VT episodes, left and right ventricular dysfunction, and the presence of fibrosis and active inflammation by imaging studies.

Conflict of interest

All authors declare no conflict of interest related to this study.

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