Bedside to Bench – A Difficult Journey

An Irregular Wide QRS Tachycardia

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Abstract

A 40 year old man presented with palpitations and was found to have a sustained, monomorphic, irregular wide QRS tachycardia. He had a history of sarcoidosis, based on muscle biopsy. Cardiac MRI, PET scan and electrophysiology study were normal. Potential mechanisms of this tachycardia are discussed.

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Key Words

Sarcoidosis • Ventricular tachycardia • Tachycardia mechanism

Introduction

Mechanistic understanding of most clinical arrhythmias are extrapolated based on rigorous bench research and/or established clinical studies. For many arrhythmias, the mechanisms don't change from case to case. For example, atrioventricular reentry, atrioventricular nodal reentry and common atrial flutter are all due to reentry and that fact is invariant. However, for many clinical arrhythmias, the mechanism is not so clear and all the clinical data may not be sufficient to go from "the bedside to the bench" in ascertaining whether the arrhythmia in question is due to reentry, enhanced automaticity or triggered activity. In these cases, the journey from the bedside to the bench may be a difficult, as highlighted by this case.

Case

A 40-year-old Caucasian man with muscle biopsy-proven sarcoidosis presented to the emergency department with new onset palpitations. He denied having had chest pain, dyspnea, lightheadedness or syncope. Past medical history was negative for similar episodes or any cardiovascular history, except hypertriglyceridemia. He had had a skin rash 2 years previously and sarcoidosis was suspected; a muscle biopsy was confirmatory. Physical examination was normal. The initial 12 lead Electrocardiogram (ECG) obtained in the emergency department showed a sustained, monomorphic, irregular wide QRS complex tachycardia (Figure 1).

Subsequent ECG showed normal sinus rhythm (NSR). Ventricular ectopic beats had the same morphology in lead II and V_1 - V_6 , as the wide QRS tachycardia (Figure 2).

The ECG

The irregular wide QRS tachycardia was thought to be ventricular tachycardia (VT). Atrial fibrillation (AF) was ruled out by 1) the fact that retrograde atrial activity (V-A conduction) was present during tachycardia (Figure 3 - arrows) and 2) wide QRS complexes due to RBBB aberrancy was ruled out because of its atypical nature (positive concordance). Positive concordance in a wide QRS tachycardia may be seen in

Figure 1. First 12 Leads ECG. Sharp spikes seen in some leads were due to printer artifact; the patient did not have a pacemaker.

Figure 2. Second 12 lead ECG.

Figure 3. The rhythm strip during episode of palpitations.

the case of a preexcited tachycardia using a posteriorly located accessory pathway (AP) or VT originating in the postero-basal left ventricle. Preexcited AF was ruled out by excluding AF and the absence of slurring of the initial part of the R-waves in V_1 - V_6 . An antidromic reentrant tachycardia was ruled out by the irregularity of the tachycardia.

Initial labs including cardiac biomarkers, thyroid function tests and electrolytes were normal. While the patient was being monitored, he had long episodes of palpitations. Rhythm strips showed the same phenomena as the 12-lead ECGs (Figure 3).

A 2-D echocardiogram was normal. Cardiac magnetic resonance imaging (MRI) showed mildly dilated right ventricle (RV) with RVEF of 45% and LV diameter at the upper limit of normal with normal EF, without late Gadolinium enhancement. A detailed electrophysiology study was normal; ventricular tachycardia could not be induced with vigorous programmed stimulation, even with isoproterenol infusion. A fluorodeoxyglucose positron emission tomography (FDG-PET) scan was normal.

Discussion

Sarcoidosis is a systemic granulomatous disease that can involve various organs such as the lung, skin, eyes, liver and the heart. Sarcoidosis is more common in women and African Americans with incidence of 10.9 per 100,000 in whites and 35.5 per 100,000 in African-Americans [1]. The heart can be involved in 25%

of patients with sarcoidosis [2]. Cardiac involvement may present with heart block, arrhythmias or heart failure. Ventricular tachycardia is a common arrhythmia in patients with cardiac sarcoidosis and the overall prognosis of sarcoidosis is worse in patients with cardiac involvement [3].

Patients with established extra-cardiac sarcoidosis need comprehensive evaluation to look for any features suggestive of cardiac sarcoidosis (CS). This includes screening tests such as a 12-lead ECG and an echocardiogram [4]. Any abnormalities in the primary tests warrant further investigation with more specific tests like cardiac MRI or FDG-PET scanning. Delayed Gadolinium enhancement in the left or right ventricle might be seen in patients with CS. A FDG-PET scan showing patchy uptake in the myocardium may suggest cardiac involvement [3].

The mechanism of VT in CS is most commonly due to reentry involving a granulomatous scar, although automaticity and triggered activity may play a role as well. Immunosuppression may aid antiarrhythmic therapy and catheter ablation may be an option for refractory cases [5]. As per the guidelines, FDG-PET is necessary to look for myocardial inflammation. Patchy uptake on FDG-PET is suggestive of inflammation and using immunosuppression may be helpful in the treatment of VT [6].

Monomorphic Ventricular tachycardia has been reported as an initial presentation for newly diagnosed CS. Some prospective studies have reported the incidence of CS in patients with unexplained VT to be

around 25-28%, suggesting that CS should be considered in the differential diagnosis of unexplained, idiopathic VT [7]. In certain cases, unexplained VT was the first sign of an underlying multisystem involvement in sarcoidosis [8].

Commentary

This patient presented with a markedly irregular, sustained, monomorphic ventricular tachycardia. What is the mechanism of this tachycardia? This is where we run into difficulty because VT was not inducible during electrophysiology study. So, the arrhythmia mechanism cannot be ascertained based on electrophysiologic maneuvers and can only be inferred based on the surface ECG. While monomorphic, reentrant arrhythmias may be slightly irregular, they don't show the degree of irregularity seen in this case. However, reentrant arrhythmias are more likely to be monomorphic, but this is not exclusive. Recently, Panda, et.al., compared VT morphologies between CS and idiopathic VT and found that pleomorphism during VT was more common in CS than in idiopathic

VT [9]. The group from UCLA has proposed a new entity, arrhythmogenic inflammatory cardiomyopathy in patients with occult sarcoidosis [10]. They found evidence of myocardial inflammation (positive FDG-PET scan) in patients with ventricular arrhythmias, suggesting cardiac sarcoidosis as the underlying cause. So, in the final analysis, the journey from the "bedside to the bench," i.e., from the clinical arrhythmia to the specific mechanism that causes it, cannot be made in this case, nor is it possible in every case. To confuse matters more, since the MRI and FDG-PET studies were relatively normal, cardiac involvement with sarcoidosis may be conjectural as well in this case.

Conflict of Interest

The authors have no conflict of interest relevant to this publication.

Comment on this Article or Ask a Question

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Cite this article as: Salehi N, Jhand AS, Satija V, Lohawijarn W, Thakur RK. Bedside to Bench – A Difficult Journey: An Irregular Wide QRS Tachycardia. *Arrhythmia Grand Rounds* 2015;1(1): 38-41. DOI: <http://dx.doi.org/10.12945/j.agr.2015.00021-14>