Cardiac Sarcoidosis Mimicking Arrhythmogenic Right Venticular Dysplasia.

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INTRODUCTION

Arrhythmogenic right ventricular dysplasia (ARVD) and cardiac sarcoidosis (CS) are uncommon causes of VA and increase the risk for sudden cardiac death.[1,2] Both diagnoses are far distinct from each other, but they can be confused if a diagnosis is made and alternative diagnoses are not thoroughly considered. We present the case of a 66 year-old male who presented to our hospital in monomorphic ventricular tachycardia due to CS mistaken as ARVD.

CASE REPORT

A 66 year-old white male presented to our Emergency Department (ED) with abrupt onset of dyspnea, palpitations and lightheadedness. In the ED, he was found to be in monomorphic ventricular tachycardia he had electric cardioversion, which resulted in normal sinus rhythm.

Initial laboratory workup came back normal and cardiac catheterization ruled out ischemia. However, on initial ECG he was noted to have an epsilon wave in V1, a QRS of 140 and T-wave inversions in V1-V5. Subsequently, an echocardiogram showed preserved left ventricular function, no valvular heart disease but a markedly dilated and severely reduced right ventricle systolic function.The patient was treated with intravenous/oral amiodarone and an implantable cardioverter defibrillator (ICD) for secondary prevention of VAs.

Since the patient had known pulmonary nodules, cardiac sarcoidosis was still in our differential diagnosis. Therefore, we obtained a fluorodeoxyglucose (FDG)-positron emission tomography (PET) computer tomography (CT), which noted uptake within multiple, patchy parenchymal pulmonary nodules with concurrent, nearly diffuse left ventricular and patchy right ventricular myocardial uptake. Findings are consistent with cardiac sarcoidosis, so steroids were initiated. To monitor response on steroids, a repeat FDG-PET CT was performed. It was notable for persistent cardiac uptake with improving pulmonary uptake. Given his refractory disease and ICD shock, 500mg twice daily of mycophenolate was added to his 40mg of prednisone daily.

On follow up FDG-PET CT imaging, the patient demonstrated drastic improvement in FDG uptake within the pulmonary parenchyma, as well as left and right ventricular myocardium (Figure 3b and Supplemental Figure 2b). Unfortunately, despite the improvement in active inflammation within the myocardium, the patient had recurrence of his VA requiring multiple shocks within 2 weeks of the scan requiring hospitalization. This event was attributed to scar and he underwent successful ablation. The patient was continued on 2g per day of mycophenolate and prednisone. With 3 months of follow up there has been no VA recurrence.

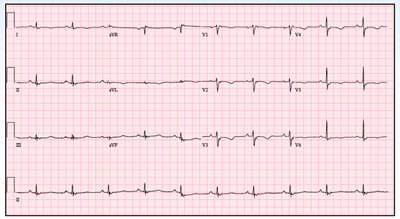

Figure 1: Initial ECG tracing notable for an epsilon wave in V1, a QRS of 140 and T-wave inversions in V1-V5, all consistent with the diagnosis of ARVD.

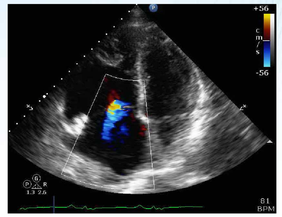

Figure 2: The four-chamber view with color Doppler image shows the marked dilatation of the right atrial and ventricular enlargement. We also noted tricuspid annular dilation with secondary, moderate tricuspid regurgitation.

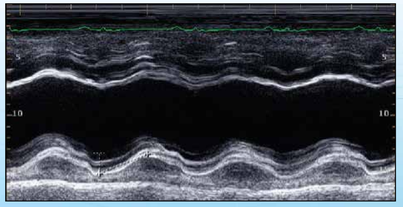

Figure 3: The patient shows reduced tricuspid annulus planar systolic excursion (TAPSE) of 1.6 cm. This is consistent with reduced systolic function of the right ventricle.

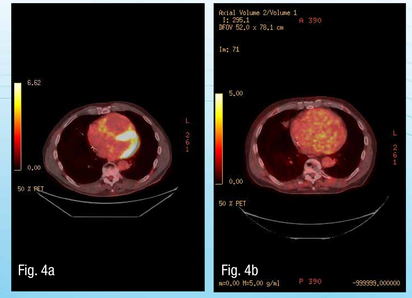

Figure 4a and 4b: Transverse FDG-PET CT images demonstrating nearly complete resolution of FDG uptake within the right and left ventricular myocardium after mycophenolate therapy was initiated.

CONCLUSION

Initial treatment is similar for all causes of VAs, but long-term management of the underlying cause is crucial. First, ischemia should be evaluated, but then other etiologies of VAs, including channelopathies, congenital heart disease, cardiomyopathies (such as hypertrophic cardiomyopathy), drugs, electrolyte imbalance, and infiltrative diseases should then be considered.[3] In this case, FDG PET CT was used to monitor therapy and assist in the diagnosis CS.

CS is a rare but important cause of VA and should be pursued when the etiology of a VA is unclear. As demonstrated in this case and prior reports in the literature, treatable infiltrative disorders can mimic ARVD and should be ruled out with further imaging with cardiac MRI or FDG-PET CT.

References:

1. Tabib A, Loire R, Chalabreysse L, et al. Circumstances of death and gross and microscopic observations in a series of 200 cases of sudden death associated with arrhythmogenic right ventricular cardiomyopathy and/or dysplasia. *Circulation* 2003;108:3000-5.
2. Mehta D, Mori N, Goldbarg SH, Lubitz S, Wisnivesky JP, Teirstein A. Primary prevention of sudden cardiac death in silent cardiac sarcoidosis: role of programmed ventricular stimulation. *Circulation Arrhythmia and electrophysiology* 2011;4:43-8.
3. Koplan BA, Stevenson WG. Ventricular tachycardia and sudden cardiac death. *Mayo Clinic proceedings* 2009;84:289-97.
4. Marcus, Frank I., et al. "Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia." *European heart journal* (2010): ehq025.
5. Dubrey, Simon W., and Rodney H. Falk. "Diagnosis and management of cardiac sarcoidosis." *Progress in cardiovascular diseases* 52.4 (2010): 336-346.
6. Youssef, George, et al. "The use of 18F-FDG PET in the diagnosis of cardiac sarcoidosis: a systematic review and metaanalysis including the Ontario experience." *Journal of Nuclear Medicine* 53.2 (2012): 241-248.