HIGH-GRADE AV BLOCK AS THE INITIAL FINDING IN ISOLATED CARDIAC SARCOIDOSIS

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Background: AV block in young patients may be related to a primary disease process for which treatment options are available. Cardiac sarcoidosis is one such entity that can have additional consequences.

Case: A 49-year-old woman with no prior medical history presented with syncope. A 12-lead ECG showed bifascicular block and long PR interval, and heart monitoring showed intermittent Mobitz 2 AV block. Laboratory data were benign, with a negative Lyme titer. Echocardiogram was benign, with LVEF of 60%. A dual chamber pacemaker was implanted. A subsequent FDG-PET scan showed foci of uptake in the LV myocardium, including the septum, with no other organ involvement.

Decision Making: The possibility of isolated cardiac sarcoidosis was considered, with options of low-yield myocardial biopsy, empiric prednisone, or clinical observation. After much discussion, a plan of close observation was adopted. The patient initially did well with no CHF or other symptoms. Heart block progressed and she became pacemaker dependent. A screening echo 12 months later revealed LVEF 35%, without symptoms. RV pacing-induced cardiomyopathy vs. cardiac sarcoid were considered. Options included upgrade to CRT-D or CRT-P, empiric steroids, or myocardial biopsy. The patient was scheduled for a RV voltage map-guided biopsy. Just prior to the planned biopsy, the patient presented in VT storm, with LVEF 5-10%. Pace-termination of VT, amiodarone, and lidocaine failed to suppress the VT. High-dose IV steroid was associated with cessation of VT. IV inotropes were needed for hemodynamic support. Cardiac cath showed no CAD. A CRT-D system was implanted. RV biopsy showed dense scar and giant cells, consistent with cardiac sarcoid. 6 months later, CHF, complex ventricular ectopy, and steroid side effects remained chronic issues, with the LVEF improving to 20%.

Conclusion: This case illustrates the importance of looking for an etiology of heart block in young patients and demonstrates the entity of isolated cardiac sarcoidosis. There are no current guidelines for this clinical situation. In retrospect, a more aggressive strategy after the positive PET scan finding might have mitigated the patient's subsequent severe clinical course.