Cardiac Sarcoidosis Induced Heart Failure

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Introduction

Sarcoidosis is a granulomatous disorder that typically presents in younger patients with unknown cause. Cardiac involvement has a wide range of involvement reported anywhere from 2 to 75% of patients with systemic sarcoidosis depending on criteria used to diagnose cardiac involvement.

Case History

A 40-year-old with no prior past medical history presented with acute heart failure exacerbation with reduced ejection fraction at 14%. Electrocardiogram showed new onset conduction delay with right bundle branch block and first-degree atrioventricular block (Figure 1). Left heart catheterization showed clean coronary arteries. Cardiac magnetic resonance imaging revealed severe cardiomyopathy with reduced ejection fraction estimated at 14% along with a patchy transmural fibrosis. Cardiac biopsy was consistent with sarcoidosis (Figure 2 & 3). Optical medical management for heart failure with reduced ejection fraction was initiated. Automatic implantable cardioverter defibrillator was placed, and glucocorticoids were started. She did not respond to glucocorticoids so methotrexate and eventually leflunomide were initiated.

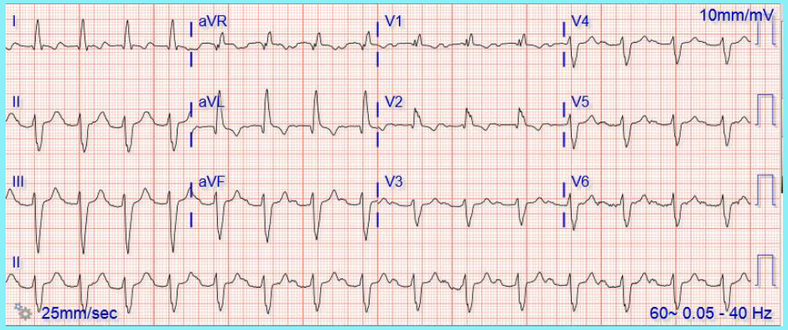

Figure 1: EKG showing first degree AV block with RBBB

Discussion

Cardiac presentations can be the first (and/or an unrecognized) manifestation of sarcoidosis in a variety of circumstances.

Diagnosis:

1. Electrocardiogram can show conduction delays
2. Echocardiograms are typically unremarkable until clinical symptoms are present
3. Cardiac magnetic resonance imaging [sensitivity (93%) and specificity(85%)] Besides the presence of regional wall motion abnormalities and segmental wall thickening or thinning, findings can include localized gadolinium enhancement which is typically patchy and multifocal, representing a combination of noncaseating granulomas and associated inflammation. In addition, the extent of myocardial late gadolinium enhancement is emerging as an important prognostic factor.
4. Endomyocardial biopsy can be used for direct sampling to diagnose cardiac sarcoidosis.

Treatment options:

1. Optimal medical management of heart failure
2. automated implantable cardioverter defibrillator when left ventricular dysfunction is present
3. Prednisone taper
4. Glucocorticoid-sparing agents such as methotrexate, azathioprine, mycophenolate and tumor necrosis factor alpha antagonists

Prognosis depends on severity of cardiac involvement. However, when treatment is initiated early, patients can be stabilized and even show improvement in left ventricular function over time.

Conclusion

This case shows the importance of including cardiac sarcoidosis in the differential diagnosis of a young and otherwise healthy patient that presents with signs and symptoms of new onset heart failure and non-ischemic cardiomyopathy.

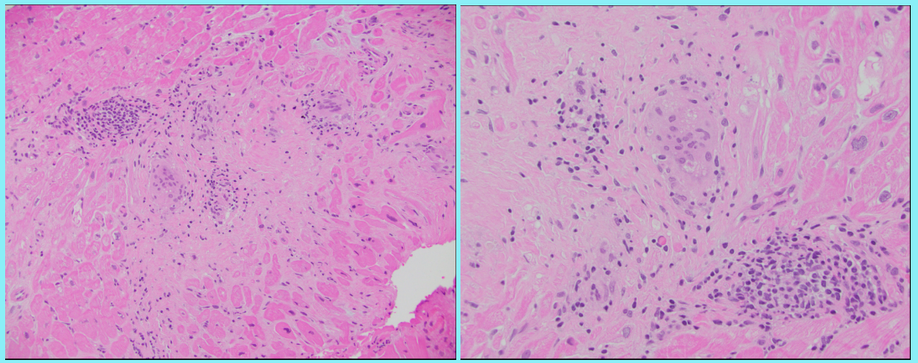

Figure 2 & 3: Cardiac biopsy showed dense interstitial fibrosis with non-necrotizing granulomas with mild inflammation composed of lymphocytes, macrophages and multinucleated giant cells, some containing asteroid bodies consistent with sarcoidosis

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