Polymorphic VT-Not Just Ischemia: Polymorphic VT as an initial presentation of cardiac amyloidosis

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BACKGROUND

Cardiac amyloidosis is an underdiagnosed cause of hypertrophic cardiomyopathy and HFpEF. It has an estimated prevalence of >8-17/100,000. Unfortunately, it can be difficult to make a definitive diagnosis without multiple forms of testing. CMR, pyrophosphate (PYP) scans have afforded us with increased diagnostic capabilities. With the increased prevalence of cardiac amyloidosis and limited treatment options for end stage disease, we aim to understand early signs of disease in order to diagnose patients as early as possible. We describe a case of cardiac amyloidosis that initially presented as recurrent polymorphic ventricular tachycardia.

CASE

A 79-year-old male with history of atrial fibrillation and diastolic dysfunction presented with shortness of breath. Labs on admission showed AKI and transaminitis, lactate 3.1, troponin 0.04 ng/mL, BNP 1017. Initial ECG was sinus rhythm with first degree AV block, low voltage (figure 1). In the ED, the patient had an episode of polymorphic VT and became unresponsive and pulseless. ACLS protocol was initiated however, he spontaneously converted to sinus rhythm.

DECISION MAKING

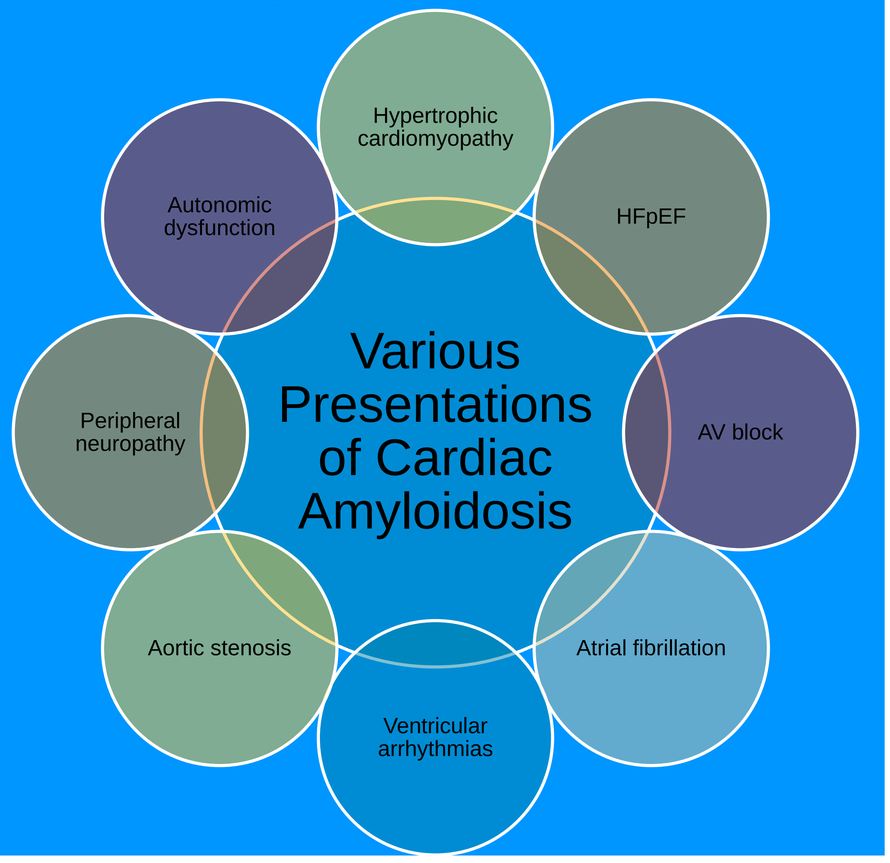
He was started on amiodarone infusion and underwent emergent cardiac catheterization which demonstrated 80% stenosis of mid-LAD and proximal diagonal for which he received DES x 2. Echo demonstrated EF 45-50% with no regional wall motion abnormalities, mild concentric left ventricular hypertrophy, grade 3 diastolic dysfunction. Despite revascularization, he continued to have multiple episodes of polymorphic VT. He underwent PYP scan which was weakly positive. Cardiac MRI demonstrated EF 52%, mild global hypokinesis and patchy mid myocardial late gadolinium enhancement in basal LV segments along with severe biatrial enlargement and inability to null the myocardium consistent with cardiac amyloidosis. An AICD was placed and he was discharged on amiodarone and diuretic therapy. He was started on Tafamidis therapy as an outpatient. He remains with good functional status 6 months later.

DISCLOSURES

No Disclosures

Cardiac amyloidosis is an underdiagnosed cause of heart failure with an estimated prevalence of >9% among patients with HCM. It is associated with increased risk of adverse outcomes including arrythmia, particularly ventricular, and HF.

It is important to consider cardiac amyloidosis as an underlying cause of polymorphic VT.



DISCUSSION

•Cardiac amyloidosis is an infiltrative cardiomyopathy with an increasing prevalence given new diagnostic tools and treatment options. It is an under-recognized cause of HFpEF.

•Cardiac amyloidosis can have various presentations including: CHF exacerbations, hypertrophic CM on echo and even arrhythmias, particularly ventricular in later stages of the disease

•Early ECG findings of cardiac amyloidosis can include low voltage, AV block, left ventricular hypertrophy, pseudo-infarct patterns, atrial fibrillation

•Diagnosis involves multimodal studies including TTE, CMR, PYP scan, lab studies to assess for AL amyloidosis and even endomyocardial biopsy.

•Cardiac amyloidosis has been linked to increased risk of arrythmias, particularly ventricular, heart failure and even sudden cardiac death

•It is important to consider cardiac amyloidosis as an underlying cause of arrhythmias and hypertrophic cardiomyopathies

FIGURE 1

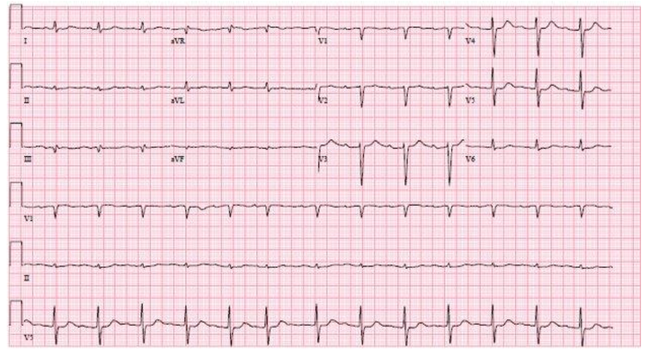


Figure 1. ECG on presentation with sinus rhythm with first degree AV block

FIGURE 2

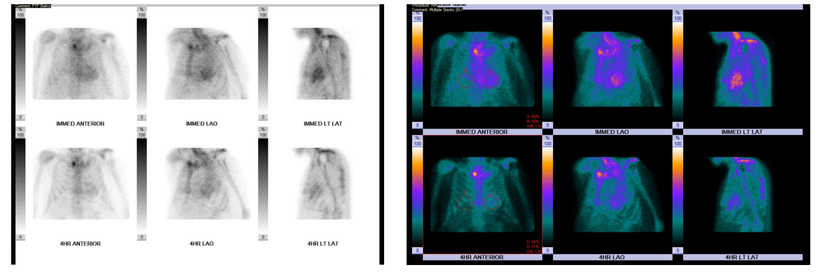


Figure 2. PYP scan with visual semi-quantitative score of 2 on the immediate imaging and a quantitative ratio of 1.51:1 suggestive of amyloid

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