CARDIAC AMYLOIDOSIS MASQUERADING AS HYPERTROPHIC OBSTRUCTIVE CARDIOMYOPATHY

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Poster Contributions
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Background: Cardiac amyloidosis (CA) remains a diagnostic challenge and is associated with significant morbidity and mortality.

Case: A 67-year old male was referred for septal myomectomy for hypertrophic obstructive cardiomyopathy (HOCM) due to worsening symptoms despite medical therapy. His medical history included chronic diastolic heart failure, and atrial fibrillation post pulmonary vein isolation. His examination revealed mid peaking crescendo-decrescendo systolic murmur, worse on Valsalva. An electrocardiogram revealed normal sinus rhythm and normal voltage. Transthoracic and transesophageal echocardiogram revealed asymmetric septal hypertrophy, chordal systolic anterior motion (SAM), and left ventricular (LV) obstruction with a peak gradient of 40 mmHg at rest. Cardiac Magnetic Resonance Imaging (cMRI) was consistent with hypertrophic cardiomyopathy with chordal SAM and no evidence of late gadolinium enhancement. He underwent successful septal myomectomy with LV gradient of 5 mmHg after surgery and resolution of SAM. The pathology revealed diffuse interstitial thickening of myocardium with mild disarray of skeletal muscle fibers. There were focal areas of brick red staining on Congo red dye and showed apple-green birefringence under polarized light. Further workup lead to the diagnosis of light chain amyloid heart disease.

Decision-making: CA is difficult to diagnose and require a high level of suspicion and multimodality imaging. Approximately 5% of patient with CA can have LVOT obstruction and could be misdiagnosed as HOCM. The standard diagnostic test is cMRI that shows widespread enhancement of thickened myocardium on delayed contrast inversion recovery. The cardiac biopsy is the gold standard with brick red staining on Congo Red dye and apple-green birefringence under polarized light

Conclusion: This case highlights the challenge in the diagnosis of CA. The identification of restrictive diastolic dysfunction and increased right ventricular thickness, are both atypical of HOCM. A high level of suspicion and multimodality imaging might assist in the early recognition of cardiac amyloidosis with chordal SAM and LVOT obstruction.