Case report

Cardiac amyloidosis presenting with paroxysmal atrial fibrillation

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Learning Point for Clinicians

We strongly recommend that when patients present with recurrent atrial fibrillation, despite medical and ablation therapy, low voltage on electrocardiogram and left ventricular hypertrophy on echocardiogram, restrictive cardiomyopathy (RCM) should be included in the differential diagnosis, especially amyloidosis, which is known as the most common cause of RCM in USA.

Introduction

Amyloidosis is a rare disease with an incidence of approximately 6–10 cases per million person years.1 It is defined as tissue deposition of fibrils consisting of different types and weight of normal serum proteins’ subunits. Amyloidosis disease can involve different organs depending on the subtype. Amyloidosis frequently affects the heart, kidneys, gastrointestinal organs and nervous system. Amyloid heart disease is an uncommon involvement that can occur as part as systemic primary (AL) or secondary (AA) amyloidosis.

Case presentation

A 69-year-old woman with a history of hyperlipidemia and recently diagnosed paroxysmal atrial fibrillation presented with worsening shortness of breath over the past 6 months despite medical and ablation therapy. She experienced associated generalized weakness and fatigue with an incremental decline in her baseline functional status whereby minimal exertion elicited symptoms. On presentation, her vital signs were stable and physical exam was remarkable for jugular venous distention (JVD). The rest of her examination was unremarkable. Electrocardiogram (EKG) was notable for low voltage, atrial fibrillation and left anterior fascicular block. On chest X-ray, there was a moderate-sized right-sided pleural effusion. Echocardiogram demonstrated preserved left ventricular systolic function and left ventricular hypertrophy (LVH). Cardiac-MR imaging demonstrated diffuse heterogeneous enhancement of thickened endocardial myocardium on late gadolinium enhancement images consistent with cardiac amyloidosis (Figure 1a). Right ventricular biopsy was negative for amyloid deposition by congo red stain under polarized light microscope; however, it showed random, non-branching fibrils ranging in size, from 8 to 10 μm in diameter consistent with amyloid deposition under the electronic microscope (Figure 1b). Patient also had lab findings of serum Lambda light chains of 651.17 and Serum Kappa light chains of 4.20, consistent with (AL) amyloidosis. Bortezomib and dexamethasone treatment were
initiated. Prior to her completion of the treatment she unfortunately passed from sudden cardiac death.

Discussion

Cardiac amyloidosis is associated with atrial arrhythmias in 10–15% of patients. The disease also presents with symptoms and signs consistent with diastolic dysfunction, which is the earliest and most significant finding on echocardiography. Left ventricular thickening due to amyloid infiltration can be missed on echocardiography and frequently read as LVH. However, unlike true LVH, left ventricular thickening in cardiac amyloidosis is associated with a decrease in electrocardiographic voltage on EKG. This combination is unique to infiltrative cardiomyopathies. The prognosis of amyloid cardiomyopathy varies depending on the type of amyloidosis, with high mortality rates in (AL) amyloidosis. Amyloidosis should be considered in patients with atrial fibrillation who fail to respond to conventional treatment and have low voltage on EKG and LVH on echocardiogram.

Conflict of interest: None declared.

References