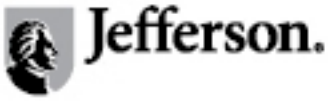

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Senile Cardiac Amyloidosis: An Undervalued Cause of Heart Failure.

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To the Editor

Despite recurrent hospitalizations for heart failure exacerbations the underlying etiology can be missed. Senile amyloidosis is a disease prevalent in the elderly African American population. Senile amyloidosis is underdiagnosed as a cause of heart failure due to its occurrence with more common comorbidities such as long standing systemic hypertension. It is important to maintain a high index of suspicion for senile amyloidosis in patients such as this case, who presented with multiple exacerbations of heart failure combined with characteristic electrocardiographic and echocardiographic findings.

Clinical Presentation

This is the case of a 71-year-old African American male with non-ischemic cardiomyopathy presumed secondary to hypertensive heart disease that presented with heart failure symptoms. He had previous hospitalizations for heart failure exacerbations in the past 2 years.

The patient's medical regimen included an angiotensin converting enzyme inhibitor (ACE-I), beta-blocker and a thiazide diuretic. EKG showed low voltage QRS complexes in all leads. The brain natriuretic peptide (BNP) was elevated. Chest x-ray revealed bilateral pleural effusions with an enlarged heart. Transthoracic echocardiography revealed marked biventricular hypertrophy, global hypokinesis, severe diastolic dysfunction and dilated atria; suggestive of infiltrative cardiomyopathy (Figure 1). Echocardiogram from 6 months earlier revealed the same findings. Serum and urine protein electrophoresis, serum free light chains and immunoglobulins, and a fat pad biopsy were unremarkable. Bone marrow biopsy demonstrated hypercellular marrow and polyclonal plasmacytosis which ruled out plasma cell dyscrasias.

Discussion

There are significant findings in this case that leads to the correct but previously missed diagnosis. The ECG showed low voltage limb leads. The echocardiogram revealed

biventricular hypertrophy, global hypokinesis, dilated atria, and a restrictive pattern suggestive of infiltrative cardiomyopathy. This combination of EKG and echocardiographic findings should raise clinical suspicion for amyloidosis, which is the most common prototype of infiltrative heart disease with increased wall thickness.[1] The endomyocardial biopsy showed amyloid deposition, specifically wild type transthyretrein protein deposition.

Amyloidosis is an infiltrative multisystem disease with the severity of specific organ involvement (including the heart) dependent on the precursor protein.[2] There are different types, but the case presented is that of the senile cardiac amyloidosis. Senile cardiac amyloidosis almost always affects men in their seventies as a slow progressive disease with only cardiac involvement.[3] The first signs of senile cardiac amyloidosis are usually leg swelling or shortness of breath, both due to congestive heart failure.[4]

Therapy for senile cardiac amyloidosis is, at present, purely symptomatic relief. Medications for heart failure, such as ACE-I, beta-blocker and diuretics are still the mainstay for treatment. Patients with senile cardiac amyloidosis however, not uncommonly progress to complete heart block, and permanent pacing becomes necessary.[5] Prompt referral to advanced heart failure physicians may improve quality of life and reduce readmissions due to heart failure exacerbations in such patients.

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Figure Legend

Figure 1. Parasternal long axis view on transthoracic echocardiogram demonstrating biventricular hypertrophy and left atrial enlargement