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The Role of Imaging in Diagnosing Transthyretin Cardiac Amyloidosis

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Introduction: Cardiac amyloidosis (CA) is a systemic disease involving various organs in the body. It is the result of the deposition of misfolded proteins, especially light chain (AL) amyloidosis and transthyretin (ATTR) amyloidosis. The diagnosis of cardiac amyloidosis is challenging and requires a high clinical suspicion. This case highlighted cardiac amyloidosis and emphasized the role of imaging in its diagnosis.

Case Report: A 65-year-old white male with a previous history of hypertension was referred for evaluation of shortness of breath with exertion and leg swelling over a one-month period. A 2D echocardiogram showed markedly increased left ventricular wall thickness without left ventricular outflow tract obstruction, mild mitral regurgitation, mild left atrial dilatation, and mildly reduced left ventricular systolic dysfunction with an estimated ejection fraction of 45-50%.

On initial physical examination, blood pressure was 110/70 mmHg with heart rate (HR) of 77 beats per minute. There was jugular venous distention of 8 cm. Cardiac auscultation showed 2/6 left upper sternal murmur, normal S1S2, and regular rhythm. Lung auscultation was symmetrical, clear bilaterally. There was 2+ bilateral lower extremity edema with intact pulses. Electrocardiogram showed normal sinus rhythm with left ventricular hypertrophy.

Due to ongoing symptoms and high clinical suspicion of cardiac amyloidosis, contrast-enhanced cardiac MRI was performed. It showed cardiomegaly with left ventricular hypertrophy. There was a diffuse transmural enhancement in the septal, inferior, and lateral left ventricular walls along with enhancement of the lateral wall of the right ventricle. There was global hypokinesia of the left ventricle with a measured ejection fraction of 42% and trace pericardial fluid.

Nuclear imaging of 99mTechnetium pyrophosphate (99mTc-PYP) planar scintigraphy showed intense radiotracer uptake by the heart that was greater than normal bone uptake (visual score of Grade 3) and a quantitative heart to the contralateral ratio of 1.9 (>1.5 considered positive); both strongly suggestive of ATTR amyloidosis. Serum and urine protein electrophoresis were unremarkable.

A right ventricle biopsy was performed. Microscopic examination showed peri-myocytic and nodular deposits of eosinophilic amorphous material in several of the fragments. The histologic and immunostaining patterns were consistent with the ATTR type of amyloidosis.

The patient was started on tafamidis 61 mg capsule daily. He noted improved symptoms of shortness of breath and edema on regular clinic follow-up.

Conclusion: This case demonstrated the advanced utility of current imaging modalities to better diagnose CA so that treatment options can be introduced at an earlier stage. In the end, the early detection of cardiac amyloidosis is critical since treatment options are currently available with the goal of improving the quality of life and survival in affected patients. Current research has shown promise that the aforementioned imaging modalities may also help assess response to therapy.

Biography

Chaaban earned the undergraduate degree in Biology with a minor in Education from the American University of Beirut (AUB). She attained her M.D. Degree from the University of Balamand, Lebanon, and then completed an internship in internal medicine at Saint Georges Hospital in Beirut. After that, she joined the research arena in Nephrology at the University of Kentucky and was also part of the Post Discharge Acute Kidney Injury Clinic. Today, she is delighted to start my residency training at the University of Kansas School of Medicine-Wichita program