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A Rare Case of Diastolic Heart Failure

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Abstract:
Cardiac amyloidosis results in a restrictive cardiomyopathy caused by extracellular deposition of proteins in the myocardium. The proteins have an unstable structure that causes them to misfold, aggregate, and deposit as amyloid fibrils This deposition causes diastolic dysfunction and can lead to diastolic heart failure
Key Word*: Amyloidosis; diastolic dysfunction; diastolic hear failure; multiple myeloma; restrictive cardiomyopathy.*

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I. Introduction

Amyloidosis refers to a large group of disorders caused by extracellular deposition of insoluble abnormal fibrils composed of misfolded proteins, which can alter tissue structure and impair function of multiple organs including the heart. Cardiac amyloidosis is primary determinant of prognosis. Cardiac involvement is primarily encountered in AL amyloidosis and transthyretin-associated amyloidosis.

II. Case Report

A 65year old male presented with extreme fatigue, weakness, loss of weight, exertional breathlessness, h/o orthopnea, PND and bilateral pitting pedal edema of 1month duration. No risk factors and no significant family history. History of surgery for carpel tunnel syndrome for both wrists. On Examination: He is pale, bilateral pitting pedal edema is present. JVP is raised. Pulse and BP are normal. Heart sounds were normal and no murmurs.

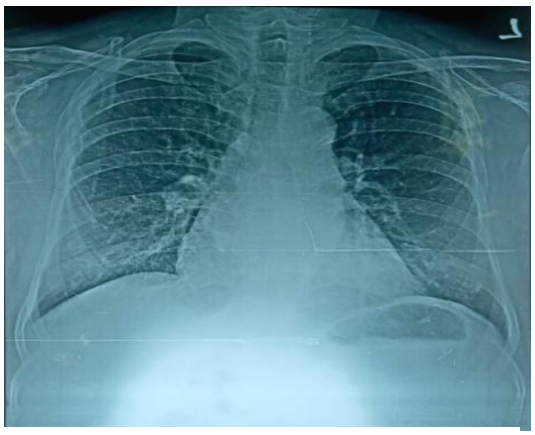

Fig 1: Chest X-ray

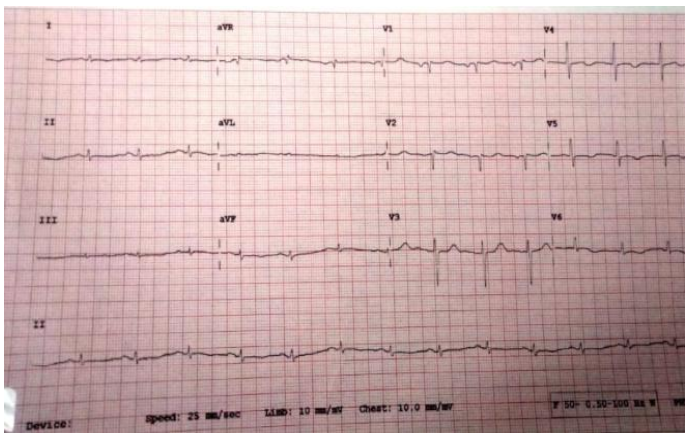

Fig 2: ECG showed low voltage complexes

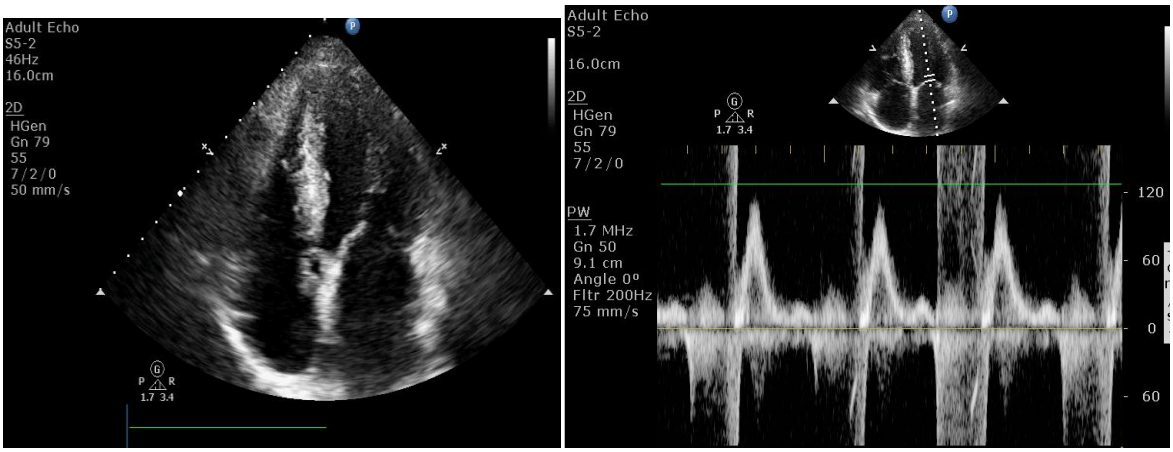

Fig 3: 2D Echo showed bi-atrial enlargement, thickened and glistening inter-atrial septum grade III diastolic dysfunction.

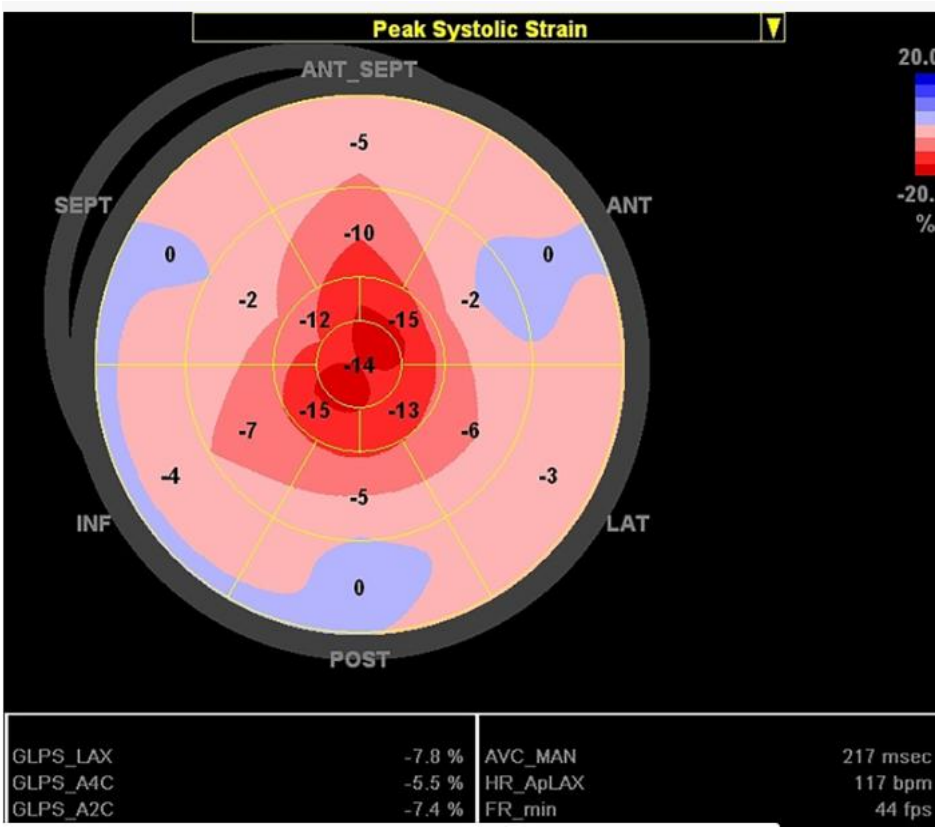

Fig 4: Strain imaging- cherry on top appearance

Cardiac MRI showed features suggestive of infiltrative cardiomyopathy.
Abdominal skin and subcutaneous adipose tissue are negative for amyloid.

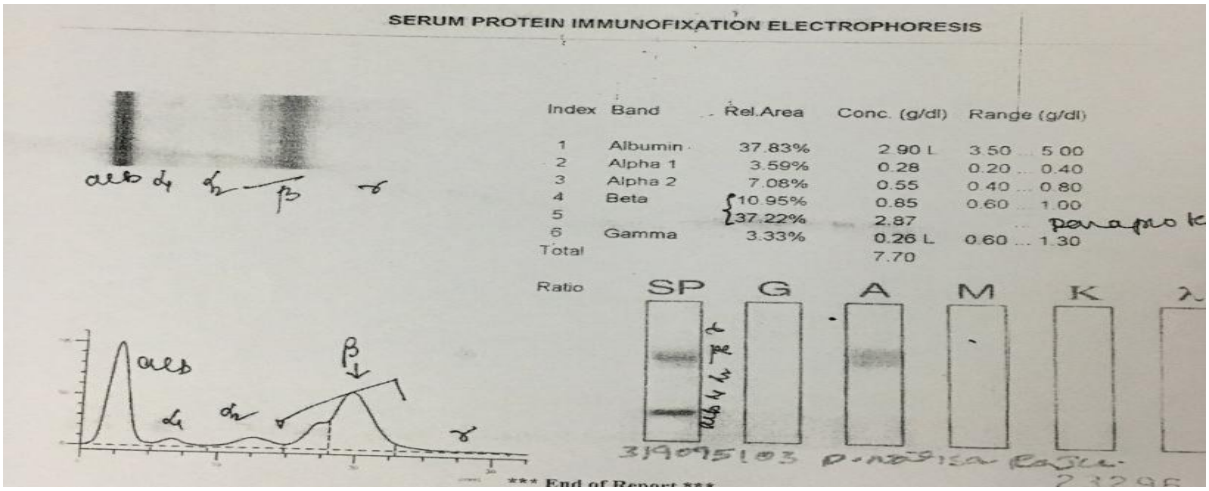

Fig 5: Serum protein immunofixation electrophoresis showed IgA Lambda monoclonal gammopathy.

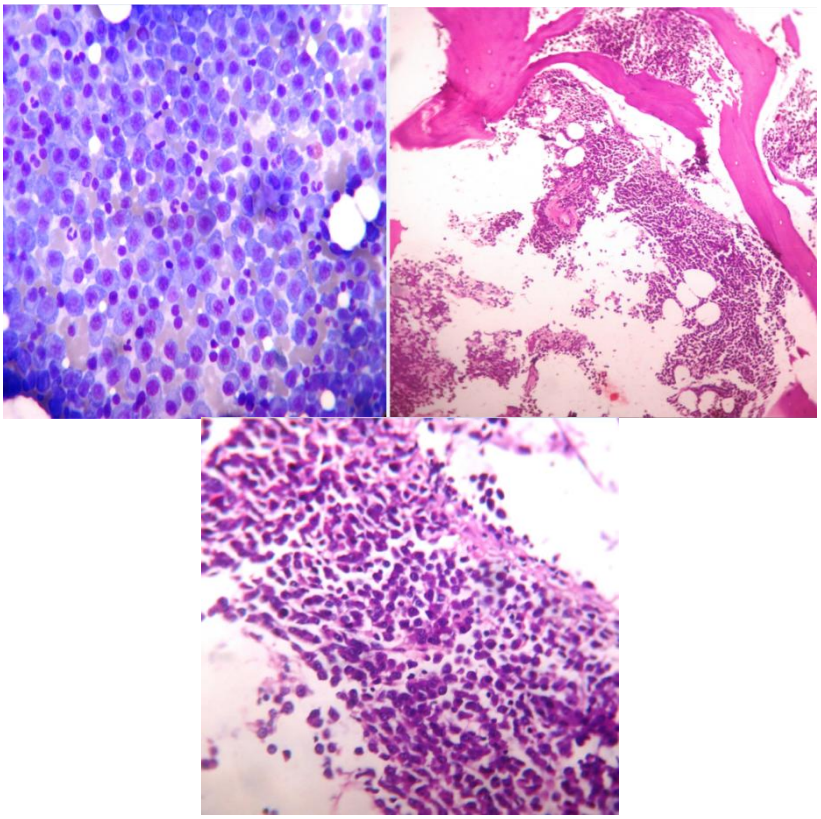

Fig 6: Bone marrow aspiration and biopsy is suggestive of multiple myeloma.

Diagnosis: This is a case of restrictive cardiomyopathy with diastolic heart failure secondary to cardiac amyloidosis due to multiple myeloma.

IV. Discussion

Amyloidosis is a systemic disorder characterized pathologically by the extracellular deposition of insoluble fibrils composed of misfolded proteins in various organs including the heart with resulting alteration in structure and function[1]. The amyloid fibrils are rigid, proteolytic resistant structures, typically less than 10 nanometers in diameter with a characteristic apple-green birefringence with Congo-red staining under polarized light microscopy[2].

Amyloidosis may be hereditary or acquired and the nature of the precursor protein of the fibrils forms the basis of classification[3]. More than 30 such precursor proteins have been identified and of these, 2 types are responsible for about 95% of cases of cardiac amyloidosis: Immunoglobulin light chain amyloidosis (AL) and transthyretin amyloidosis (ATTR)[4].

AL, although overall a rare condition, is the most frequently diagnosed form with about 0.3 cases per 100000 population in a large referral center[5]. AL occurs secondary to a plasma cell dyscrasia resulting in excessive production of immunoglobulin light chain units, with associated misfolding and deposition in extracellular tissue. In addition to cardiac manifestations, hepatic, renal and neurologic complications may be seen in AL. Early diagnosis and initiation of treatment in AL may alter disease outcome although mortality remains high[6].

The diffuse deposition of amyloid fibrils in the cardiac interstitial space produces significant thickening of both ventricles with associated stiffness which results in impaired diastolic relaxation and the characteristic restrictive physiology[2,7].

The classic clinical presentation of cardiac amyloidosis is with symptoms of congestive heart failure including exertional dyspnea and orthopnea. Fatigue and weakness are also common, resulting from compromised CO. Symptoms of right ventricular failure including abdominal distension and lower extremity edema may be more prominent in certain patients[2,8].

The treatment of cardiac amyloidosis is two pronged – treatment of symptoms arising from cardiac dysfunction and treatment of the underlying amyloidosis.

V. Conclusion

Early diagnosis of cardiac amyloidosis may improve the outcomes. This requires heightened suspicion and a systemic clinical approach for evaluation.

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