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Heart Failure: As Initial Presentation of Systemic Amyloidosis in Absence of Renal Features

A Case Report Presented in APICON 2016 in Hyderabad

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Abstract: ***Amyloidosis is the term for a group of protein folding disorders characterized by the extracellular deposition of insoluble polymeric protein fibrils in tissues and organs. Systemic amyloidosis can have variable clinical presentation. Heart is the 2nd most commonly affected organ (50-60% cases), after renal involvement. But cardiac involvement as initial presentation is rare. Amyloid cardiomyopathy is characterized by concentric ventricular hypertrophy and diastolic dysfunction associated with elevation of brain natriuretic peptide and troponin.***

Keywords: amyloidosis, cardiomyopathy, low voltage ECG

1. Introduction

Amyloidosis presents in a variety of ways and can make diagnosis difficult. Systemic amyloidosis implies involvement of visceral organ(s) or multiple tissues. Kidney is the most common organ involved in AL, AA, and most forms of hereditary amyloidosis, except ATTR. Proteinuria is present in 73% of the AL amyloidosis patients, with 30% exhibiting nephrotic syndrome. Renal involvement is nearly universal (97%) in AA amyloidosis. Heart is the next most common organ involved In AL amyloidosis with abnormal echocardiographic findings noted in 65% of patients. It is the predominate feature in senile TTR amyloidosis.

Presentation varies from asymptomatic to a subtle decrease in exercise capacity to fatigue, dyspnea, and lower extremity edema to angina, syncope, ascites, and anasarca, which are associated with more advanced disease. Overt heart failure can be seen in 24%. Low voltage on ECG and concentric thickened ventricles on echocardiogram are classic signs of cardiac involvement by amyloidosis.

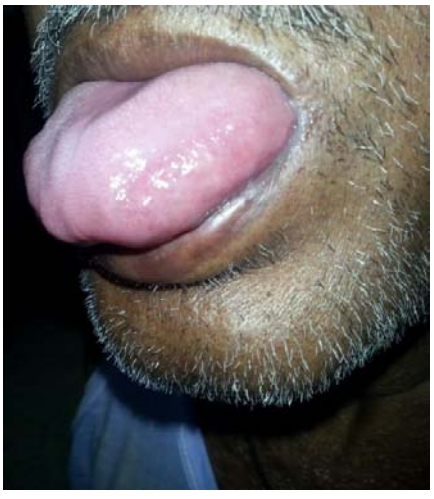
2. Case Report

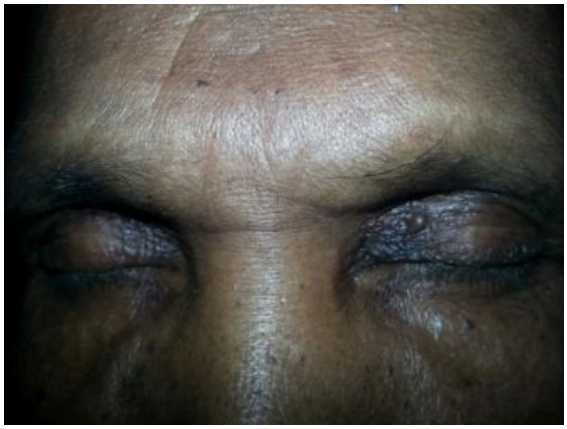
A 58yr old male, presented in emergency department with complains of breathlessness, chest pain and swelling of both lower limb since 2 months. On examination, the patient was anxious and restless. He had pallor, engorged neck veins, bipedal oedema and bilateral decreased breath sound with fine basal crepts. His heart rate was 96 per minute and BP was 108/76 mm of Hg. Cardiac examination was normal. An urgent ECG showed low limb lead voltage with atrial fibrillation. The patient was managed in line of DCMP with heart failure and he was symptomatically relieved.

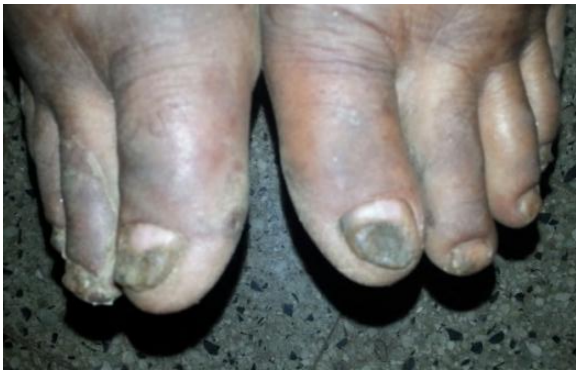
He was a chronic smoker and alcoholic. There was no history of chronic medical illness including HTN, DM type 2, asthma or tuberculosis.

His routine blood tests and urine examinations was normal. His chest x-ray revealed bilateral pleural effusion, more on right side and was transudative in nature. Trans-thoracic echocardiography suggests severe LVH, normal LVEF and type 1 diastolic dysfunction without any evidence of pericardial effusion. His USG whole abdomen was normal.

The presence of severe LVH (in absence of hypertension) in a 58 yr old male and presence of low voltage ECG, particularly in limb leads (in absence of pericardial effusion) was alarming. Only the infiltrative disorders like amyloidosis can solve this paradox. Later on, upon reexamination, there was surprising facts beside us. There was periorbital eccymosis on the medial side of upper eyelid of both eyes, nail dystrophy affecting nails of all four limbs and tongue hyperthrophy without any teeth indentation, which is pathognomonic of systemic amyloidosis. It was confirmed by “apple green” birefringence by polarized light microscopy when abdominal fat aspiration was stained with Congo red dye.







Kidney is the most common organ involved in systemic amyloidosis. But there was no evidence of any renal involvement as his USG whole abdomen, renal function tests, serum and urinary protein was within normal limit.

3. Discussion

Heart failure as the initial presentation in systemic amyloidosis, in absence of renal involvement, is a rare entity and is often difficult to diagnose. Systemic amyloidosis should always be in differential diagnosis in a patient with low limb lead voltage in ECG and ventricular wall hypertrophy in echo, because amyloidosis with initial cardiac involvement can easily be misdiagnosed by ECHO as hypertrophic cardiomyopathy. In fact, combined ECG and echo model would increase the ability to identify 'Amyloid Cardiomyopathy' in clinical practice.

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Author Profile

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