Amyloid Heart in Senile Amyloidosis

Jasmine Kamboj, Ranadeep Mandhadi, Rahul Kamboj,
Vamsi Kodumuri and Daniela Kovacs

Mt Sinai Medical Center, Chicago, USA

Introduction

Amyloidosis in heart consists of deposition of the insoluble amyloid protein in the heart leading to the replacement of the contractile elements with non compliant rubbery tissue, resulting in restrictive cardiomyopathy and congestive heart failure. Deposition in the conduction pathways, including the SA Node, AV node and infra-His pathways, can lead to arrhythmias and blocks. Cardiovascular amyloidosis could be primary or secondary to systemic illness. The etiology is important in determining the prognosis and median time for survival, with worst prognosis in primary amyloid heart and best in senile amyloidosis.

Here we have a case of amyloid cardiomyopathy secondary to senile amyloidosis with congestive heart failure and atrial fibrillation.

Case

A 67 year old African American female came with acute shortness of breath from the nursing home for 6 weeks. She had worsening shortness of breath on lying down and swelling of feet bilaterally.
She was admitted at another facility 2 months back for cardiogenic shock secondary to acute CHF decompensation and flash pulmonary edema.

She gave a history of 30 pound weight gain since diagnosis of congestive heart failure, around 2 years back. There was no history of bone pains, no fever, chills or night sweats.

Past Medical History was positive for hypertension, diabetes mellitus, atrial fibrillation and congestive heart failure. No history of multiple myeloma, any cancer or any hematologic abnormalities.

Family History – was positive for heart disease of unknown type. No FH for cancer or hematologic disorders.

Vitals : BP 98/70;; HR 70s;; RR 18;; Oxygen saturation 96% on 4litres NC;; Temp 98.8F. Physical Examination: she was alert, awake, oriented in time place and person, looked distressed, positive Jugular venous distension of 9 cm, no lymphadenopathy or masses in neck. On auscultation chest had good air entry, bilateral crepitations in the lower lobes of the lungs; irregular heart rate with audible S3, no murmur/rubs. Abdomen was distended, with good bowel sounds, positive abdomen wall edema, positive hepatomegaly with + hepatojugular reflex. She had bilateral pitting edema of 2+ on both lower extremities.

Labs and Imaging:BUN and creatinine of 50 (h)and 2.5(h), GFR 17(l). BNP1849(h). Rest of the CBC-D, CMP, LFTs, TSH were WNL. Troponins : 0.00, 0.04, 0.01

Chest X Ray showed pulmonary vascular congestion with bilateral pleural effusions.

EKG showed atrial fibrillation, with ventricular rate of 78 per minute, low voltage complexes, non specific intraventricular conduction block with anterolateral infarct.

Echocardiogram showed: LV - concentric left ventricular hypertrophy, global hypokinesis, Ejection fraction of 20%, restrictive LV filling, stage 3 diastolic dysfunction. LA – mildly dilated. RV – Global wall thickness increased (5-10mm). RA – mildly enlarged. Aortic valve did nt show any stenosis or regurgitation. Mitral valve had mild to moderate regurgitation. Mild pulmonary regurgitation. Mild to moderate pulmonary hypertension with RVSP 56 mm Hg. Moderate generalized pericardial effusion. Moderate pleural effusion. IVC dilated with poor inspirational collapse, consistent with elevated right atrial pressures.
Course: The patient being in acute CHF decompensation with atrial fibrillation, acute kidney injury and anasarca, was admitted in the step down unit. On reviewing the echocardiogram findings, restrictive cardiomyopathy was suggested by the cardiology team. Amyloid Heart was one of the differentials. General surgery was consulted and abdominal fat biopsy was congo red stain positive. Oncology service was called in, and they ruled out multiple myeloma, by negative (for monoclonal protein) serum and urine protein electrophoresis. Considering the fact that the CHF was diagnosed few years back, patient’s African American race, and age > 60 years, senile systemic amyloidosis was the suggested diagnosis. Genetic testing for trasthyretin gene mutation was sent (to rule out familial amyloidosis) and came out to be negative. The patient was put on dobutamine drip for its inotropic action and was started on high dose lasiks for anasarca by the renal team. She received amiodarone for atrial fibrillation rate control and was put on anticoagulation. Over the next 48 hours, the patient showed improvement in terms of her shortness of breath, edema and anasarca and was transferred from the step down unit to telemetry floors. The long term options of cardiac transplant, autologous stem cell therapy and possibility of the need for pacemaker were discussed with the patient, but she opted out for all these modalities.

Discussion

Senile systemic amyloidosis has a rate of 25% in patient population > 60 years, with increased prevalence in African American population. It typically involves the atria (91%) and sometimes aorta (2). It has a better prognosis (median survival of 5 years) (1) as compared with primary cardiac amyloidosis but is not completely benign. It results in heart failure, atrial fibrillation and conduction disturbances. Presentation of CHF will occur months to years before the actual cause if discovered.

Restrictive Cardiomyopathy, is the primary manifestation of amyloid heart. The progression occurs as: restrictive pattern → right heart failure → elevated JVP → right sided gallop → hepatomegaly → peripheral edema.

Involvement of the conduction system → can cause arrhythmias and heart blocks.

Involvement of the pulmonary vasculature → can cause pulmonary hypertension and cor pulmonale. (4)
Depending on the site of protein accumulation, ECG may reveal low voltage complexes, poor R wave progression, conduction disturbances, +/- infarct patterns.

Echocardiogram will show abnormal relaxation (early stage), thickened ventricular walls, abnormal texture (granular sparkling), atrial dilatation, valvular thickening or regurgitation, pericardial effusion, and restrictive pattern with elevated filling pressure (late stage).

Diagnosis: Clinical suspicion and echocardiographic findings of restrictive cardiomyopathy are suggestive, of amyloid heart. Biopsy of endomyocardium is the gold standard for diagnosis. However, biopsy of extracardiac tissue (tongue, subcutaneous fat pad, kidney, bone marrow) in background of clinical presentation and echo findings is appropriate. Congo red stain, producing apple green birefringence, under polarized light, is the most specific stain for amyloid.

Treatment and Prognosis: In the event of acute decompensation of CHF, an inotropic agent like dobutamine is appropriate. Digoxin can cause toxicity by binding to amyloid fibrils and hence is not recommended. CCB and BB can worsen cardiac function due to their negative inotropic effect. For atrial fibrillation, amiodarone or ibutilide are effective and anticoagulation is needed. Pacemaker may be needed for symptomatic bradycardia, sick sinus syndrome or high grade conduction disturbances.

Autologous stem cell transplantation, cardiac transplantation, +/- liver transplantation for familial amyloidosis) have been advocated, with better remission and improvement in median survival.

Troponin elevation, involvement of SA and AV node, and infra-HIS conduction prolongation are independent predictors of SCD sec to CHF decompensation in amyloid heart. Left ventricular hypertrophy, right ventricle dilatation are inversely related to median survival period. Clinically, syncope/ dizziness are important in establishing prognosis.

References
Amyloid heart in senile amyloidosis

2. Pathol Int 1995;45:335-42
4. Chest 2001;120:1735-8
5. American heart journal 1946;32:419-37

Received: May, 2012