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Case Report

Cardiac amyloidosis: A report of two cases

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ABSTRACT

Cardiac amyloidosis is a manifestation of multisystem disorder. The condition is rare, difficult to diagnose and invariably fatal. We report 2 cases of amyloidosis associated with plasma cell dyscrasia. A high index of clinical suspicion, echocardiographic clues (ventricular thickening, diastolic dysfunction, biatrial enlargement) and elevated cardiac biomarkers led to the diagnosis. Early institution of amyloid specific treatment should be the aim. Cardiac treatment is supportive and results are often disappointing.

1. Introduction

Systemic amyloidosis is a relatively rare multisystem disorder caused by extracellular deposits of insoluble fibril proteins in various tissues and organs. Amyloidosis patients may present to any speciality and the diagnosis is frequently delayed. Cardiac amyloidosis refers to involvement of the heart by amyloid deposition whether as a part of systemic amyloidosis (as is most commonly the case) or as a localized phenomenon.[1,2]

We report two cases of this rare disease associated with Waldenstrom's macroglobulinemia in one and uncertain type of plasma cell dyscrasia in the other. The cases highlight the difficulties in recognition and adverse prognosis. Role of echocardiography, cardiac biomarkers and tissue biopsy in diagnosis is emphasized.

2. Case reports

2.1. Case I

A 22 years young male was referred in 2008 with progressive dyspnea, fatigue and episodes of presyncope during the last 6 months. He gave history of receiving antitubercular therapy six months back for a febrile illness and attributed all his symptoms to this therapy.

Clinical examination revealed anemia, blood pressure of 90/70 mmHg, slow atrial fibrillation (AF), elevated jugular venous pressure, cardiomegaly, loud P2, hepatomegaly and clear lung fields. Relevant blood investigations revealed anemia, mildly deranged liver and renal function tests attributable to heart failure. Cardiac troponin (2.35 ng/ml, normal <0.1 ng/ml) and NT pro BNP (1600 pg/ml, normal <125 pg/ml)

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were elevated. Electrocardiogram revealed AF with ventricular rate of 30–35beats/minute and non-specific ST-T changes. Skiagram of chest revealed cardiomegaly (cardiothoracic ratio of 0.65), prominent left and right atrium and signs of pulmonary venous hypertension. Two dimensional echocardiography (Fig. 1A and B) revealed marked thickening and speckled appearance of interventricular septum, left ventricular and right ventricular posterior wall, biatrial dilatation and left ventricular ejection fraction (LVEF) of 70%. Atrioventricular valves were normal. Doppler interrogation revealed restrictive pattern and moderate pulmonary hypertension (PH). Hemodynamic data is shown in Table 1 and revealed elevation of end diastolic pressure in both the ventricles and moderate PH. Left and right ventriculography (Fig. 1C and D) showed hypertrophy of the ventricles, delayed relaxation, normal systolic function and favored a cardiac infiltrative disease.

Abdominal fat pad biopsy demonstrated apple green birefringence on staining with Congo red and provided the diagnosis of amyloidosis. Serum and urine electrophoresis and bone marrow suggested abnormal plasma cells. Serum immunofixation test could not be performed. There was marginal symptomatic benefit with diuretics and a VVI pacemaker implantation. No specific therapy for amyloidosis was initiated. Six months later, patient died at home.

2.2. *Case II*

A 72 years male with Waldenstrom's macroglobulinemia, was admitted to hematology unit of our institute for chemotherapy in September 2012. He had cough, dyspnea and an episode of transient AF. Skiagram chest PA view revealed bilateral basal haziness and computed tomography (CT) of chest showed minimal bilateral pleural effusion and interstitial septal thickening. Bronchoscopy did not reveal any abnormality. Serial electrocardiograms showed sinus rhythm and non-specific ST-T changes. Bedside two dimensional echocardiography revealed normal cardiac valves, normal LVEF, no pericardial effusion and no PH. Chemotherapy using monoclonal antibody rituximab, steroids and supportive treatment was instituted. There was partial response and patient returned to his hometown.

A month later, he was readmitted with altered sensorium and progressive dyspnea. Clinical assessment revealed tachypnea, irregular pulse, blood pressure of 100/70 mmHg, bilateral basal crepts and no murmur or S3 gallop. Serial electrocardiograms during the next one week revealed varying rhythms (AF, atrioventricular dissociation, bradyarrhythmias and non-specific ST-T changes). Serial skiagrams of chest revealed hilar haziness, signs of pulmonary venous congestion and suggested pulmonary edema. Relevant blood investigations revealed anemia (Hb – 9.5 g%, normal – 13–17 g %), hyponatremia (Serum Na – 113 mEq/L, normal – 137–150 mEq/L), markedly elevated NT Pro BNP (5606 pg/ml, normal <125 pg/ml). Other biochemistry tests were normal. Repeat two dimensional echocardiography revealed massively thickened interatrial septum (39 mm, normal – 6 ± 2 mm) and thickening of left and right ventricular posterior wall (Fig. 2A and B). There was biatrial dilatation, normal cardiac valves and LVEF of 60%. Doppler interrogation revealed evidence of diastolic dysfunction (E/E′ – 39).

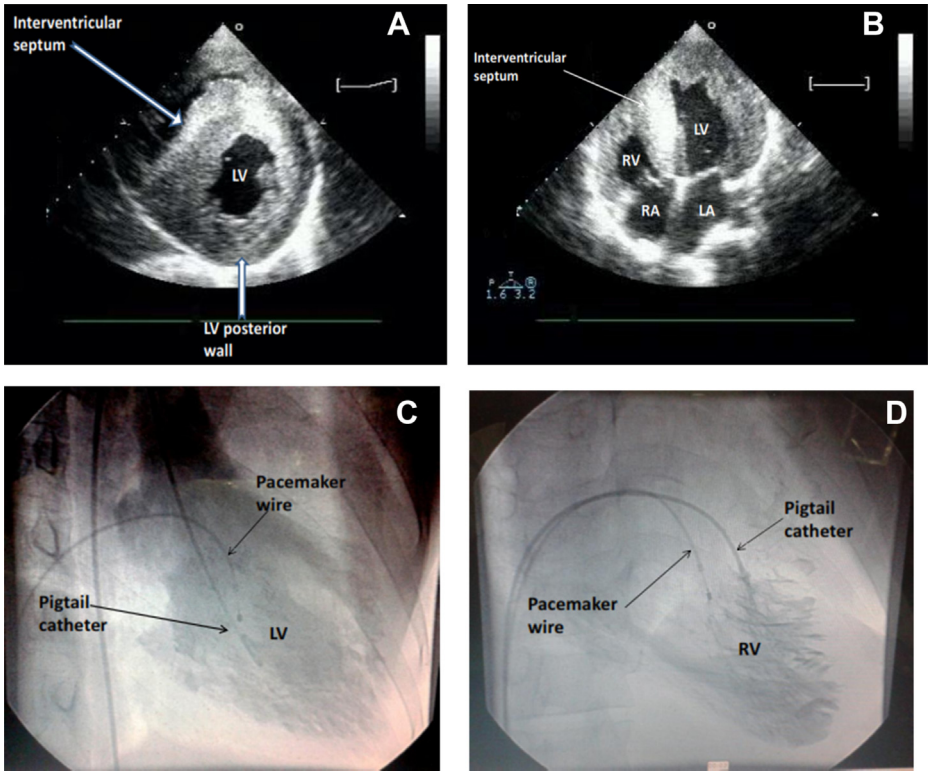

Fig. 1 – Two dimensional echocardiography in short axis (A) and apical 4 chamber view (B) shows speckled appearance and thickening of interventricular septum, LV and RV posterior wall. LV angiogram (C) and RV angiogram (D) showing hypertrophied ventricles. LA – left atrium, RA – right atrium, LV – left ventricle, RV – right ventricle.