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Cardiac amyloidosis: A report of 2 cases and review of literature

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ABSTRACT
Cardiac involvement by amyloidosis is a rare finding and many of these case remain undiagnosed for long due to lack of tissue diagnosis. We report two cases one each of primary and secondary cardiac amyloidosis which are confirmed on biopsy and immunohistochemistry.
Keywords*: Amyloid; Cardiac amyloidosis ; Heart failure*

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INTRODUCTION

Cardiac amyloidosis pertains to disposition of amyloid (β-pleated) protein in the cardiac tissue which is resistant to proteolysis and cause oxidative stress in organ.[1]. Cardiac involvement by amyloidosis is a rare finding and many of these case remain undiagnosed for long due to lack of tissue diagnosis. Cardiac involvement is usually seen in primary and hereditary transthyretin amyloidosis and rarely in secondary amyloidosis. [2]. We report two cases one each of primary and secondary cardiac amyloidosis confirmed on biopsy and immunohistochemistry.

MATERIAL & METHODS

We report two cases, one each of primary and secondary amyloidosis causing restrictive cardiomyopathy.

Clinical details:

Case 1 was a 50 year old male who presented with complaints of chest pain and dyspnea on exertion (NYHA II-III) for last 5-6 months. He underwent ECG and Echo (Fig 1) which showed left ventricular hypertrophy with low voltage limb leads in ECG. (Fig 2). LVEF was 35%. Cardiac biopsy from showed deposition of amyloid in interstitium and around individual myocytes. IHC performed Kappa Restriction. The patient was thus diagnosed as Primary Amyloidosis. Further work up of patient was performed based on biopsy findings. Bone marrow biopsy showed features of plasmacytoma with serum protein electrophoresis showed M band. Case 2 was a 46 year old male with sudden onset shortness of breath with no associated chest pain. Patient was a known case of Rheumatoid Arthritis for last 10 years and is on regular medication. Cardiac biopsy from right ventricle showed deposition of amyloid in interstitium and also along the vessel wall. IHC performed is negative for Kappa and Lambda and showed positive staining on Serum Amyloid Associated Protein. The patient was thus diagnosed as Secondary Amyloidosis.

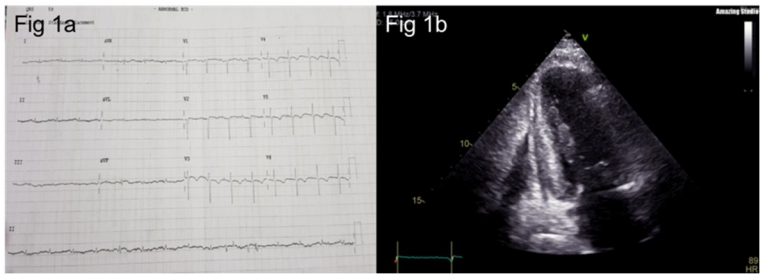

Fig 1a ECG showing low voltage leads Fig 1b-Echo showing LVH

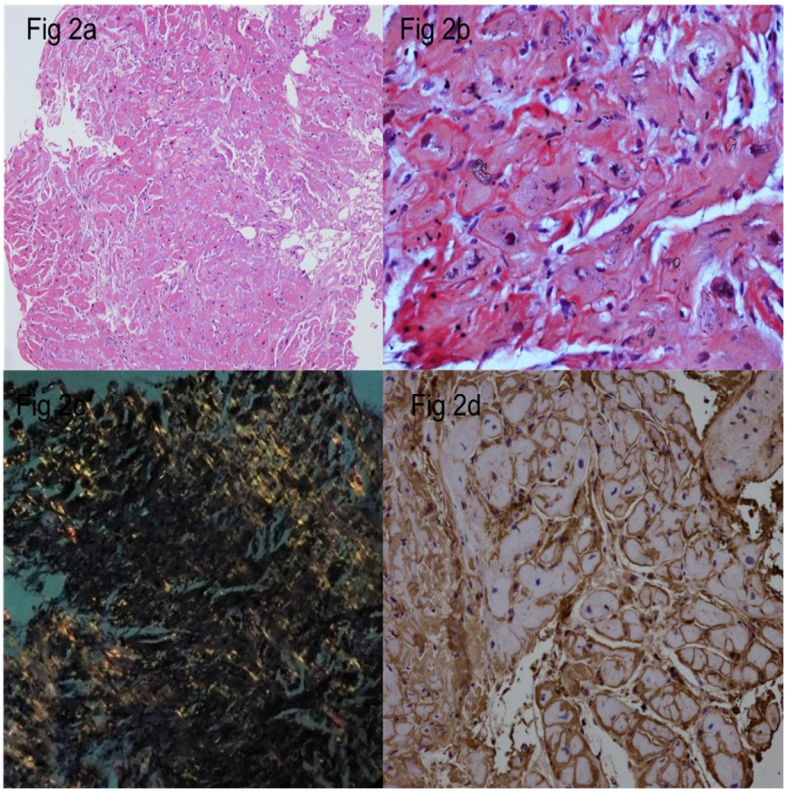

Fig 2a-Cardiac biopsy showing pericellular and interstitial amyloid deposition (H&E X 10) Fig 2b-Congo red stain positive in amyloid (CR X 10)Fig 2c-Polarizer showing apple green birefringence Fig 2d-IHC for SAA showed SAA positivity

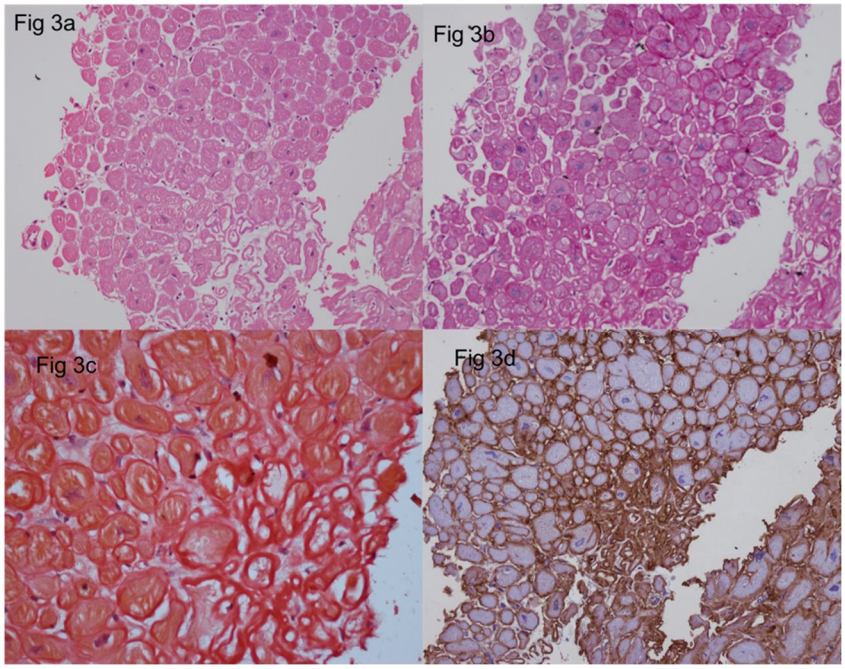

Fig 3a-Cardiac biopsy showing pericellular amyloid deposition (H&E X 10) Fig 2b-PAS stain showing weak PAS positivity in eosinophilic material (PAS X 10) Fig 3c-Congo red stain positive in amyloid (CR X 10) Fig 3d-IHC for Kappa showed Kappa restriction

DISCUSSION

Amyloidosis affects various tissue and organ such as kidney, heart, nerves, liver and gut.[3] Clinical presentation depends upon the type of organ involved and also on the degree of organ dysfunction. In heart, amyloid deposition occurs in interstitial pericellular and endocardial areas.[4] Usually a large amount of

amyloid is already deposited in heart at the time of diagnoses. [5] Cardiac involvement is most common in AL amyloidosis accounting around 50% of cases and rarely occurs in secondary amyloidosis.[6] Most common presentation is rapidly progressing heart failure involving around 60% of cases and is caused by amyloid deposition in between myocytes leading to restricted cardiac filling and causing abnormal diastolic function. It is often manifested as dyspnea. Amyloid deposition in arteries sometimes causes angina and myocardial infarction because of the progressive narrowing of lumen.[6,7] Electrocardiographic (ECG) findings are usually found normal in an already known case but the hallmark ECG finding are low voltage QRS and pseudo-infarction pattern. [8] Echocardiography is important tool for diagnosing amyloidosis but many of the characteristic features are present in later half of disease. Common echocardiography features are thickening of left ventricular wall, granular myocardial appearance, thickened interatrial septum, bilateral atrial enlargement, thickened valves and pericardial effusion. [2] Fine needle aspiration of abdominal fat is helpful in detecting systemic AL amyloidosis (84% sensitivity) [9]. Definitive diagnosis is established on abdominal fat biopsy (67% diagnostic) or on bone marrow biopsy (72% diagnostic) or on endomyocardial biopsy in case of localised cardiac amyloidosis.[6,5] Amyloid is confirmed by staining with Congo red which under polarised light gives green birefringence (sensitivity 57–85% and specificity 92–100%) or can be stained with sulphated Alcian blue which has higher specificity. [6] Immunohistochemistry or mass spectrophotometry can be employed to determine the specific type of amyloid protein involved for appropriate treatment. Immunohistochemistry commonly employed are anti-kappa, anti-lambda, anti-amyloid A and anti-transthyretin serums antibodies. [6] The prognosis of cardiac amyloidosis mainly depends upon the extent of cardiac involvement. Median survival rate in patients with cardiac AL amyloidosis is 6 months from the onset of heart failure symptoms. The major causes of death are heart failure and pulseless electronic activity causing sudden death. [6]

CONCLUSION

Cardiac amyloidosis is rare and because of nonspecific features it diagnosed late and often requires high level of suspicion. Histopathological examination is gold standard for diagnosing amyloidosis along with use of special stains such as Congo red and immunohistochemistry. Immunohistochemistry is particularly important in developing nation for knowing the specific amyloid protein involved in affected organ for proper treatment.

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