

CASE REPORT

CARDIAC AMYLOIDOSIS IN A PATIENT WITH MULTIPLE MYELOMA

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ABSTRACT: Cardiac amyloidosis is a rare disorder, which is characterised by the extra cellular deposition of amyloid, a protein polysaccharide complex in the heart. This infiltrative cardiomyopathy presents with restrictive heart failure. We report a 58 year old male who presented with macroglossia, proteinuria and heart failure. His detailed evaluation revealed multiple myeloma with concurrent Primary (AL) amyloidosis. This case report is to highlight the occurrence of cardiac amyloidosis and multiple myeloma which are two separate plasma cell dyscrasias which have presented together.

KEYWORDS: Cardiac amyloidosis, Multiple myeloma, AL Amyloidosis, Macroglossia.

CASE PRESENTATION: A 58 year old male presented with a 10 month history of slowly progressive abdominal distension, swelling of both legs, shortness of breath and orthopnea. He also complained of generalized fatigue, loss of weight and appetite. On general examination, he had an elevated jugular venous pressure, pallor, normal blood pressure, macroglossia (Fig.1) and pitting edema of both lower limbs up-to the level of the knees.



Fig 1: Clinical picture revealing macroglossia

Systemic examination revealed a soft grade 2/6 systolic murmur in the pulmonary area, hepatomegaly and ascites. Investigations revealed a normocytic anemia (Haemoglobin: 8.4mg/dL, Red blood cells: 3.17×10^6 /cu.mm and Mean Corpuscular Volume: 86fl) and elevated ESR (130mm at 1

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hour). The serum albumin level was 2.7g/dL and globulin level was 7.4g/dL. His renal parameters were normal and urinalysis revealed proteinuria (1280mg per day). Liver function tests revealed elevated alkaline phosphatase of 204 U/L (Normal range: 26–120U/L). Serum protein electrophoresis revealed an M spike (Fig.2) and immunofixation electrophoresis was positive for IgG λ (Fig 3).

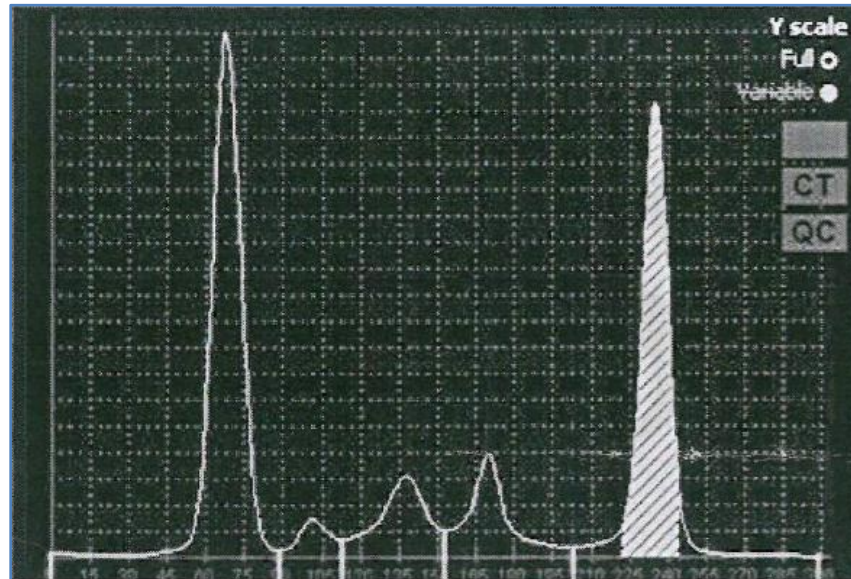


Fig 2. Serum protein electrophoresis with prominent M spike in the gamma region

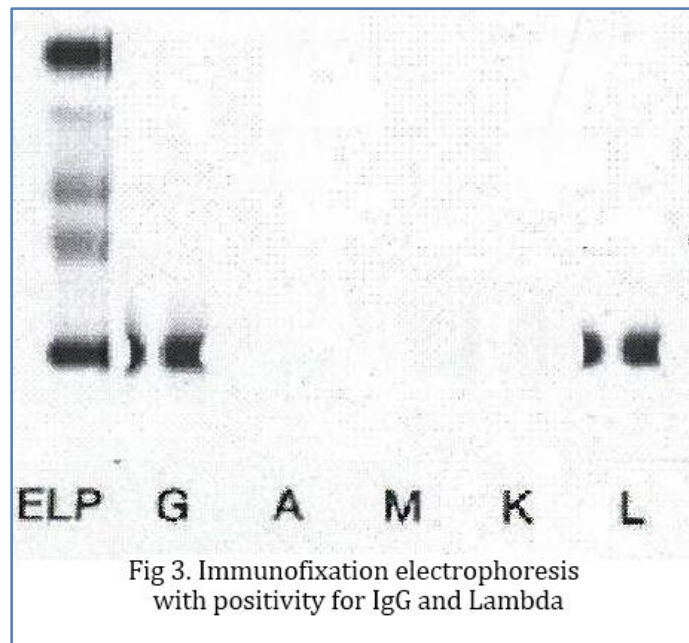


Fig 3. Immunofixation electrophoresis with positivity for IgG and Lambda

Thyroid function tests were normal. The electrocardiogram revealed a 1st degree AV block. Abdominal Ultrasound revealed hepatomegaly with ascites. Echocardiography showed biatrial enlargement with biventricular hypertrophy and a hyperechoic, granular myocardium which was

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suggestive of amyloid infiltration. Radiological investigations revealed cardiomegaly with right sided pleural effusion, punched out lesions were detected in skull and diffuse osteopenia of the spine. (Fig 4a, 4b).

Fig. 4a: CT imaging of skull showing well defined punched out lytic lesions in skull bones

Fig. 4b: Sagittal CT sections of lumbar spine showing thickened marrow trabeculations with ill-defined lytic areas in vertebral bodies of lumbar spine.

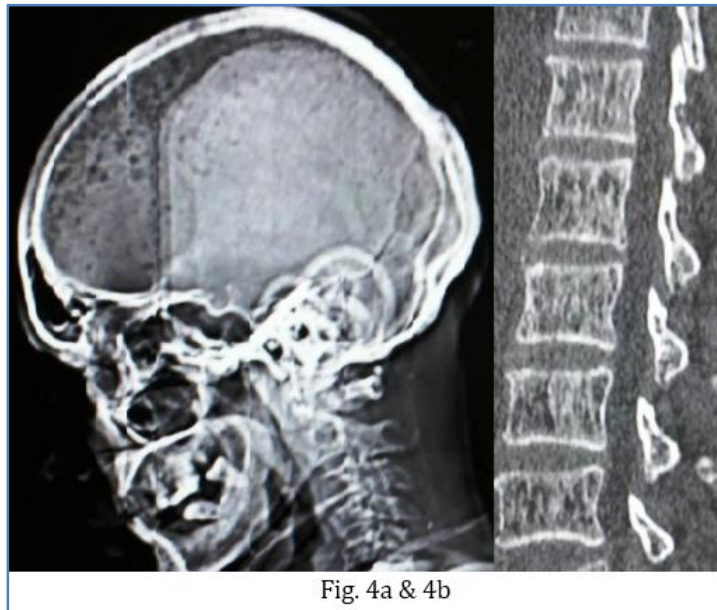


Fig. 4a & 4b

Peripheral smear study showed typical rouleaux formation. Bone marrow examination revealed plasma cells with cart wheel chromatin suggestive of multiple myeloma. (Fig 5)

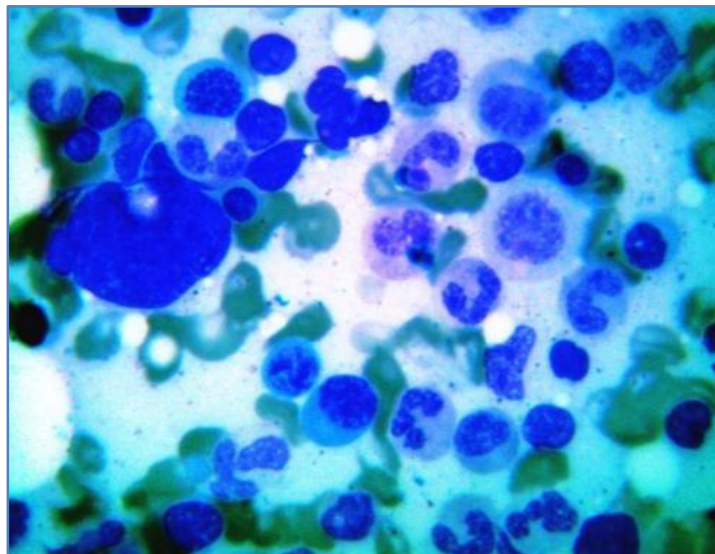


Fig 5: Bone marrow aspiration smear (100X) shows plasma cells with cartwheel chromatin

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He received treatment for heart failure with furosemide and spironolactone and his condition improved. He was started on cyclophosphamide, thalidomide and dexamethasone and is under our follow-up.

DISCUSSION: Multiple myeloma (MM) is a common haematological malignancy characterised by the proliferation of abnormal clonal terminally differentiated plasma cells.¹ Routine use of comprehensive laboratory panels has helped to diagnose MM in many asymptomatic patients, who constitute up to 1/3rd of all patients.²

Our patient fulfilled all 3 diagnostic criteria (Values in parentheses) for multiple myeloma.³ Hepatomegaly is more commonly associated with it than splenomegaly and about 10% of the patients will exhibit macroglossia.⁴ Infrequently, it manifests as a local disorder, restricted to a single organ.² Macroglossia with indented tongue is considered pathognomonic of AL amyloidosis.

Cardiac involvement, in the form of restrictive cardiomyopathy is present in up to 50% of patients with AL amyloidosis. The presence of ventricular hypertrophy in the absence of systemic hypertension along with the presence of granular & hyperechoic myocardium in our case leads to the suspicion of cardiac amyloidosis⁵.

The 24 hour urinary protein of 1280mg/day denotes the renal involvement in amyloidosis. The definite diagnosis of AL amyloidosis requires histopathological confirmation but the patient refused biopsy. But the presence of involvement of multiple organs, in a characteristic manner, helps us make a diagnosis of AL amyloidosis with reasonable certainty.

Multiple myeloma and AL amyloidosis are both clonal plasma cell proliferative disorder which present with different phenotypes. Among the two, multiple myeloma is more common, with a 10 fold higher prevalence.⁶ The progression of AL amyloidosis to multiple myeloma is very rare but a diagnosis of AL amyloidosis is made frequently in patients with pre-existing multiple myeloma.⁷ This represents the coexistence of two separate but overlapping diseases of the plasma cell and shouldn't be wrongly labelled as multiple myeloma with secondary amyloidosis.⁸

Our patient has features of cardiac amyloidosis and macroglossia which favour a diagnosis of AL amyloidosis. Even though the percentage of plasmacytes in the marrow is less than 30%, the presence of lytic lesions on radiology confirms the diagnosis of an overt myeloma.⁹

CONCLUSION: In any case of refractory cardiac failure with multiple organ involvement, the clinician should have a high index of suspicion of amyloidosis. This case is presented to highlight the association of two separate plasma cell dyscrasias, namely multiple myeloma and primary amyloidosis.

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