Cardiac Light Chain Amyloidosis in a Patient with Heart Failure with Preserved Ejection Fraction

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CASE BLOG

A 55-year-old man with a history of exertional fatigue presented with dyspnoea since 10 days ago. The electrocardiogram showed low voltage on the limb leads I and aVL, and QS-type waves on the precordial leads V1 to V3. The echocardiography revealed the ejection fraction 59%, left ventricular end-diastolic diameter 36 mm, and interventricular septum 15 mm. The serum B-type natriuretic peptide was 1940 pg/ml and the 24-hour urine protein was 3.69 g. Renal biopsy showed amyloid deposition in glomeruli and mesangial cells (panel A). Both the Congo red staining and potassium permanganate staining were positive (panels B, C). The bone marrow aspiration showed plasma blasts 0.4%, proplasmacytes 6.8% and plasmacytes 17.6%. Flow cytometry showed that there were 24.36% abnormal cells of bone marrow (D, R2, red), positive CD38 expression (E) and the positive expression of kappa light chain (F). The patient was diagnosed with systemic light chain amyloidosis with cardiac and renal involvements and given melphalan and prednisone. His recovery was uneventful.

Figure 1: Hematoxylin and eosin staining (A) and Congo red staining (B) show amyloid deposition in glomeruli and mesangial cells. Potassium permanganate staining (C) is positive. Flow cytometry shows 24.36% abnormal cells of bone marrow (D, R2, red), positive CD38 expression (E) and the positive expression of kappa light chain (F).

Amyloidosis is a protein-folding disorder with more than organ infiltrated by proteinaceous deposits, and the prognosis of the disease is determined both by the organs involved and the type of amyloid. Cardiac amyloid involvement carries the worst prognosis of any involved organ. Early diagnosis is important for improving the patient’s outcome.