AL Amyloidosis in a Patient with IgD Myeloma

Sir,

Multiple myeloma associated with IgD monoclonal protein is a well-recognized, but rare entity that affect less than 2% of patients with multiple myeloma.[1]

Many studies including patients with IgD multiple myeloma have been published to date.[2-6] This entity appears to have clinical and evolutionary differences compared with non-IgD myeloma, including a younger age at disease presentation, a higher incidence of kidney injury and a poor prognosis. A 55 years old male, with pulmonary tuberculosis in 2007, presented with lower limbs edema, dyspnea and upper limbs paresthesia for the past 4 months. He did not have bone pain. Physical examination revealed peripheral edema, macroglossia, periorbital petechial and ecchymotic lesions. There was no peripheral lymphadenopathy and no hepatosplenomegaly.

Laboratory results were as follows: serum albumin 27 g/l, total serum proteins 47 g/l, 24 hours urine protein 7 g, hematuria 3 × 104/ml, serum creatinine: 0.6 mg/dl, serum potassium: 3.7 mEq/l, serum calcium 11.3 mg/dl, hemoglobin: 11.3 g/dl, white cell count: 9.2 × 109/l and platelet count: 314 × 109/l. A renal biopsy was performed. It revealed eleven glomeruli. Three of them were sclerosed. Remaining glomeruli revealed acellular, amorphous, pale pink deposits with Congo red positivity and apple-green birefringence under polarized light. Immunofluorescence revealed that staining for lambda light chain was positive and staining for kappa light chain was negative. AL amyloidosis diagnosis was retained.

Serum protein electrophoresis showed hypogammaglobinemia 2.66 g/l that evoked free light chain myeloma [Figure 1]. Therefore, we systematically completed routine serum protein immunofixation by testing for IgD and IgE that revealed IgD lambda monoclonal immunoglobulin. Urine protein immunofixation showed lambda monoclonal free light chains at a high concentration [Figure 2]. The serum levels were IgG: 3.04 g/l, IgA: 0.18 g/l, IgM: 0.11 g/l, kappa free light chain: 0.94 mg/dl, lambda free light chain: 1218.84 mg/dl and kappa to lambda ratio <0.09. Bone marrow aspiration showed a marrow invaded by 60% of dystrophic plasma cells confirming multiple myeloma.

Standard skeletal imaging was done and was found normal. Echocardiography revealed dilated left ventricle, anterolateral hypokinesis, and good systolic function with ejection fraction at 55%. Coronary angiography showed spastic atheroma.

Due to severe cardiac involvement, (Mayo Clinic stage 4: Troponin T: 138 ng/l, N-terminal propeptide of brain natriuretic peptide: 9380 ng/l, serum free light chain difference: 12179 mg/l),[7,8] a bortezomib-cyclophosphamide-dexamethasone regimen was used for the treatment. Unfortunately, after one cycle, the patient died of myocardial infarction.

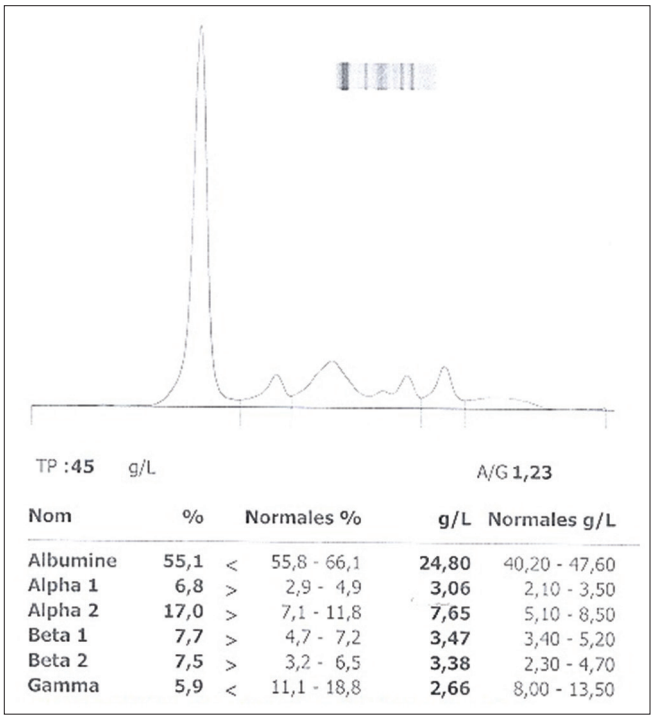

Figure 1: Serum protein electrophosis showed hypogammaglobinema

IgD amyloidosis is rare. Among 3955 patients with AL amyloidosis seen at Mayo Clinic during a period of 41 years, 53 patients (1.3%) had a serum IgD monoclonal protein. Those patients had a lower frequency of renal and cardiac involvement and had similar survival than their non-IgD counterparts.[9]

The diagnosis of IgD amyloidosis can be difficult. In fact, the IgD monoclonal level is often small and immunofixation in search of IgD is not systematically performed. Therefore, many cases could be unnoticed or taken for a light chain myeloma.[10]

Patients, in whom electrophoresis evoke the presence of monoclonal free light chains, should also be tested for the presence of both IgE and IgD.[11]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not

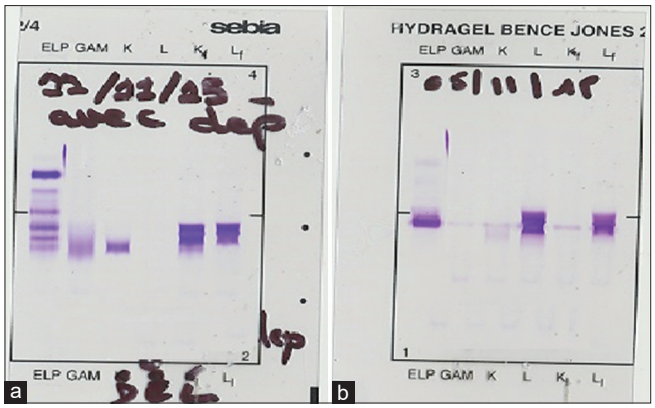

Figure 2: (a) Serum protein immunofixation revealed IgD lambda monoclonal immunoglobulin. (b) Urine protein immunofixation showed lambda monoclonal free light chain

be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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