Bone marrow amyloidosis

A 54-year-old man presented with nephrotic syndrome, renal impairment and restrictive cardiomyopathy. On admission, physical examination revealed pallor, peripheral edema and moderate hepatosplenomegaly. Hematologic values were: Hb 9.7 g/dL, WBC 10.4×10^9/L with a normal differential count, platelets 109×10^9/L. Kappa monoclonal light chains were identified by urine immunofixation. Circulating kappa free light chains were 378 mg/L and lambda 56.3 mg/L. Bone marrow aspirate displayed slightly hypocellular marrow with maturing hematopoietic progenitors and mild dyserythropoiesis. There were 8% morphologically normal plasma cells. Various-sized clumps of pink amorphous material were scattered on the smears (Figure, A and B). These deposits stained with Congo red (Figure, C), that under polarized light produced a characteristic apple-green birefringence (Figure, D). Abdominal fat pad aspirate confirmed the presence of amyloid. Therefore, a diagnosis of AL amyloidosis with renal, cardiac and bone marrow involvement was made. The patient was treated with high-dose dexamethasone with progressive improvement of his condition.

Systemic AL amyloidosis is a plasma cell dyscrasia in which the fibril amyloid protein is produced by monoclonal plasma cells and consists of whole or fragments of immunoglobulin light chains. It is associated with plasma cell myeloma in about 15% of cases. In the other cases a moderate monoclonal increase in plasma cells is usually present in the bone marrow. A monoclonal immunoglobulin is found in the serum or urine in more than 80% of patients. Amyloid deposits are detected in blood vessels and as interstitial foci in bone marrow sections in approximately 60% of patients, but very rarely, and only when there is extensive bone marrow involvement, extracellular amyloid clumps are present in a bone marrow aspirate.

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References