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IMAGES IN HEMATOLOGY

# Primary AL amyloidosis presenting with systemic lymphadenopathy with calcification

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A 72-year-old man presented with systemic lymphadenopathy, including cervical, axillary, and inguinal bilateral lymph nodes. He had noticed slowly progressive lymphadenopathy for more than 10 years, while edema of the right calf, impotence, and modest dyspnea on exertion had developed, more recently. His inguinal lymph nodes were mobile, non-tender, and very large (hen's egg size). Further physical examination revealed a modest decrease in bilateral grip strength, modest hypotonia of bilateral quadriceps, and distal tingling of the left hand on percussion. There were no apparent signs of heart failure or arrhythmia. Laboratory data showed a modest decrease in serum IgG (8.21 g/L), elevation of IgM (9.93 g/L), and deviation of light chain κ/λ ratio (0.161; Ig-κ 23.2 mg/L and Ig-λ 144 mg/L). Serum protein immunoelectrophoresis detected increased IgM-λ monoclonal protein. Neither renal dysfunction nor electrolyte disturbance was observed. Bone marrow was normocellular with no increases in plasma cells, dysplasia, or any abnormal cells. Computed tomography (Fig. 1a–d) revealed enlarged systemic lymph nodes, with conspicuous swelling of the obturator lymph nodes. In addition, diffuse and granular calcification was observed in these nodes. A biopsy specimen obtained from the right axillary lymph node revealed diffuse deposition of amorphous and acellular matter without abnormal cells (Fig. 2a). Apple-green birefringence was detected under polarized light, which was considered to be amyloid deposition (Fig. 2b). On immunohistochemistry, the biopsy was positive for λ-chain and negative for κ-chain (Fig. 2c,d). Biopsy of the rectal mucosa also revealed amyloid deposition. Based on the above findings, a diagnosis of primary AL amyloidosis was established.

Primary AL amyloidosis shows a variety of symptoms, which may vary depending on the site of amyloid deposition, such as congestive heart failure, arrhythmia, hepatic dysfunction, or renal failure. Amyloid fibrils have affinity for calcium ions [1], and calcification associated with amyloidosis has been sporadically reported in cases presenting with mesenteric lymphadenopathy [2], pericardial thickening [3], or gastrointestinal amyloidosis [4]. However, systemic lymphadenopathy with calcification, as observed in this case, is extremely rare.

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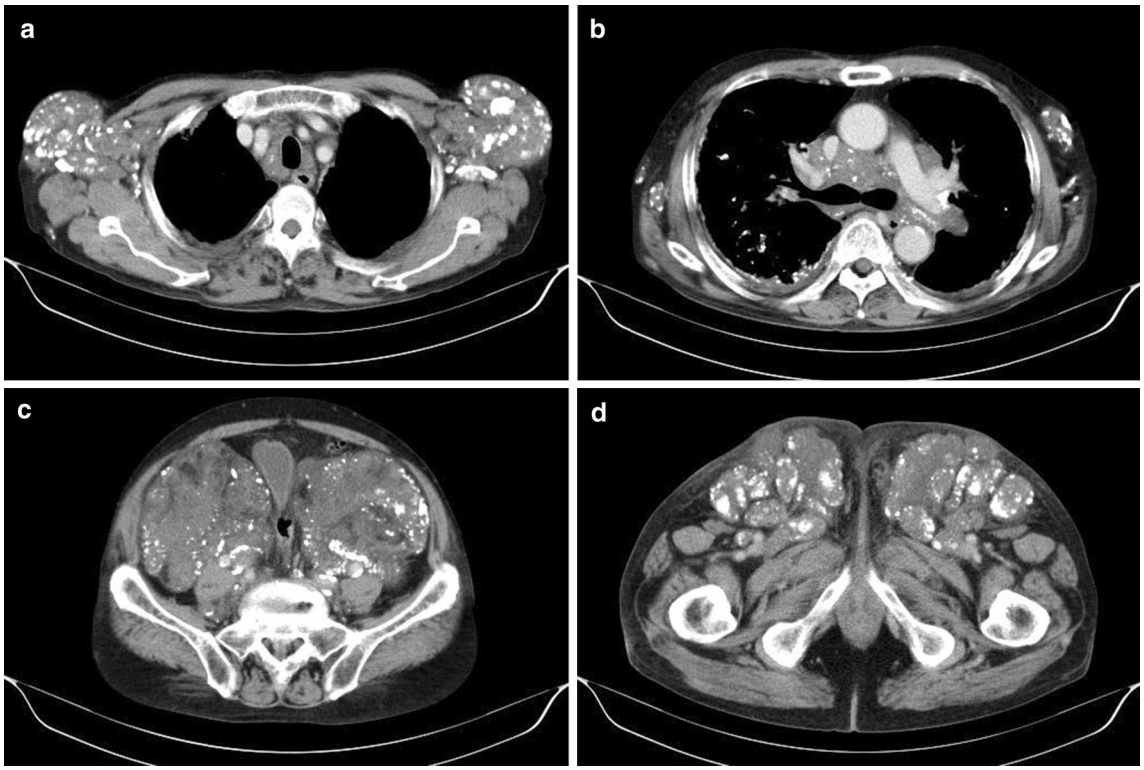


**Fig. 1** CT findings

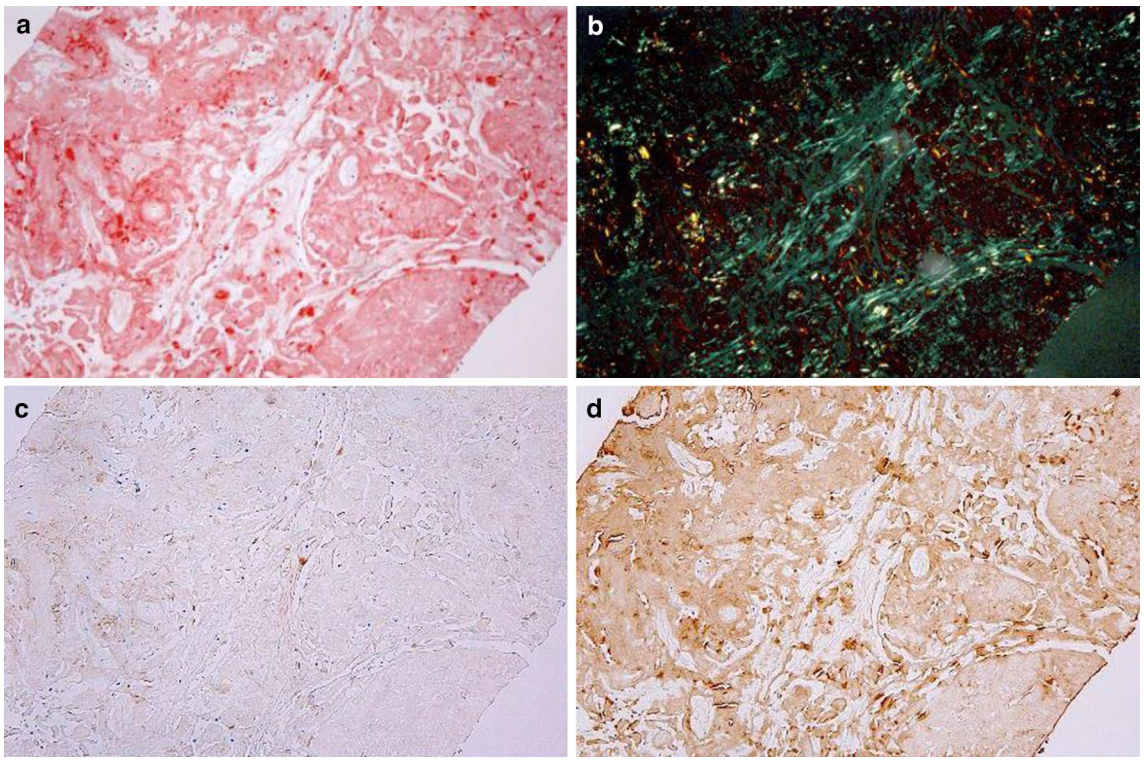


**Fig. 2** Histological findings of the lymph node. **a** Congo-red staining. **b** Polarized microscope. **c** Immunostaining for Aκ light chain. **d** Immunostaining for Aλ light chain

**Compliance with ethical standards**

**Conflict of interest** None declared.

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