

Cardiac amyloidosis

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A 39-year-old African-American man with no significant medical history presented with an episode of syncope. For the previous three weeks, he had experienced progressive dyspnea on exertion and lower extremity swelling. On examination, he was slightly tachycardic, at 102 beats/min, and had elevated jugular venous pressure, a grade 2/6 pansystolic murmur, bilateral crackles and moderate pitting edema of both lower extremities. His electrocardiogram showed low voltage in the frontal and anteroseptal leads. Echocardiography showed moderate to severe concentric left ventricular hypertrophy with a ground glass appearance, suggestive of an infiltrative process (Figure 1) (1). The left ventricular ejection fraction was estimated to be 20% to 25% with global hypokinesis, and there was a pseudonormal mitral inflow pattern and a thickened interatrial septum. Coronary angiography showed no significant atherosclerosis. Urine and serum protein electrophoresis demonstrated a monoclonal protein

spike in the beta-1 region, with the diagnosis of amyloidosis confirmed by Congo red staining on bone marrow biopsy (2,3), which showed the characteristic brick-red colour of the affected blood vessel walls and interstitium.

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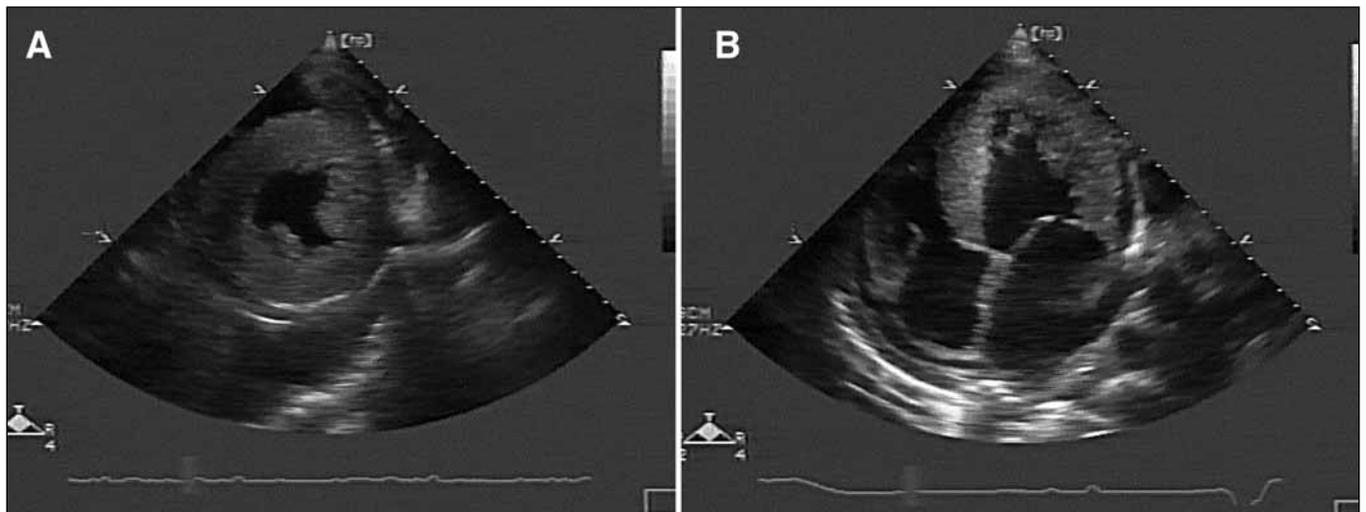


Figure 1 Echocardiography showing moderate to severe concentric left ventricular hypertrophy in the short-axis view (A) and apical four-chamber view (B)

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