

DIAGNOSIS

Echocardiography and cardiovascular magnetic resonance imaging showed signs of amyloid cardiomyopathy. Congo red-stain of abdominal fat pad aspirate samples revealed apple-green birefringence. Since IgM lambda and free light chains (FLC) lambda were elevated, amyloid light-chain (AL) amyloidosis was diagnosed. Additional bone marrow examination showed a lymphoplasmocytic lymphoma. Since he had both lymphoplasmocytic lymphoma and AL amyloidosis, treatment with bortezomib and dexamethasone was started to reduce the production of amyloidogenic FLC. Unfortunately, neither the FLC count nor the nail dystrophy responded to this treatment before it was discontinued because of side effects. He was then treated with dexamethasone, rituximab, and cyclophosphamide. After an initial flare-up, the FLC count decreased somewhat; however, after several months of treatment it increased again. Subsequently, he

was treated with ibrutinib, which was recently approved for lymphoplasmocytic lymphoma. After a short period, he developed atrial fibrillation, a known side effect of ibrutinib, and therefore the treatment was stopped. He suffered from progressive heart failure and died a few months later at the age of 72 years. The appearance of his nails had never improved during treatment.

AL amyloidosis is a rare proliferative clonal plasma cell disorder in which fibrils of monoclonal light chains are deposited in extracellular tissue, such as the heart, liver, kidney or intestines. It is often overlooked at first and diagnosed at a late, irreversible stage. Nail dystrophy is a known but rare clinical finding in AL amyloidosis that is caused by amyloid deposition in the nail bed and nail fold. Early detection of AL amyloidosis, using clinical clues such as nail dystrophy, may enhance treatment options and thereby increase survival in this rare but severe disease.