Letters to the Editors

Amyloid myopathy masquerading as polymyositis

Sirs,

Polymyositis is an idiopathic inflammatory myopathy manifesting as proximal weakness, raised muscle enzymes and histopathological abnormalities on muscle biopsy. Here, we present a case of systemic amyloidosis presenting with polymyositis-like symptoms and a true amyloid myopathy,

A 58 year old woman with no past medical history presented to the Rheumatology clinic with a 2 month history of gradually progressive proximal muscle weakness. She had specifically noticed difficulty climbing stairs and washing her hair. She remained systemically well and continued to work as a cleaner. There were no associated symptoms suggestive of a connective tissue disorder, notably, no rash, Raynaud's, shortness of breath or dysphagia, and no weight loss. She was an ex-smoker of 8 years and abstained from alcohol. On examination, there was no evidence of muscle pseudohypertrophy, and the muscles did not feel firm or "woody" on palpation. Using manual muscle testing, power was 3/5 bilaterally in the proximal arms and leg musculature with evidence of truncal weakness. No other focal neurological features were present. Initial investigations included ALT 52 IU/l, GGT 124 IU/l, ESR 37 mm/hr, CRP 1 mg/l and CK 1,567 IU/l, IgG 18.6 (6-16), IgA 0.5 (0.8-4), IgM 0.28 (0.5-2) and negative antibody screen using immunoprecipation. Protein electrophoresis showed a compact band with immune paresis (suppression of other normal immunoglobulins), paraprotein load 15 g/dl and negative Bence Jones protein on urine electrophoresis. A bone marrow aspirate subsequently revealed 25% plasma cell infiltrates. Chest x-ray and abdominal ultrasound imaging were normal. An MR of the thigh musculature showed bilateral patchy active myositis on Short Tau Inversion Recovery sequences with fatty streaking on T1 sequences reflecting low grade fatty replacement of muscle. Electromyography testing showed a moderate excess of brief duration motor units of simple and complex morphology consistent with moderate myopathic changes, but no abnormal spontaneous activity was seen indicative of a more active myositic process. Urine dipstick showed proteinuria and urine protein:creatinine ratio was 279 mg/mmol. A quadriceps muscle biopsy was performed, and reported as showing the presence of extensive perimysial amyloid deposits with perivascular accentuation (Fig. 1-2). No inclusion bodies or rimmed vacuolated fibres were present. Electron microscopy confirmed focal aggregate/deposits of amyloid fibrils around the vessels and muscle fibres, in relation to the latter showing focal areas of scalloping of the fibres. Immunostaining revealed positive labelling of potential amyloid deposits which were lambda light chain restricted. The muscle biopsy was also sent to the National Amyloidosis Centre although insufficient sample was available for Congo Red staining. Low and subsequently high dose prednisolone failed to halt the patient's progressive weakness and CK rise.

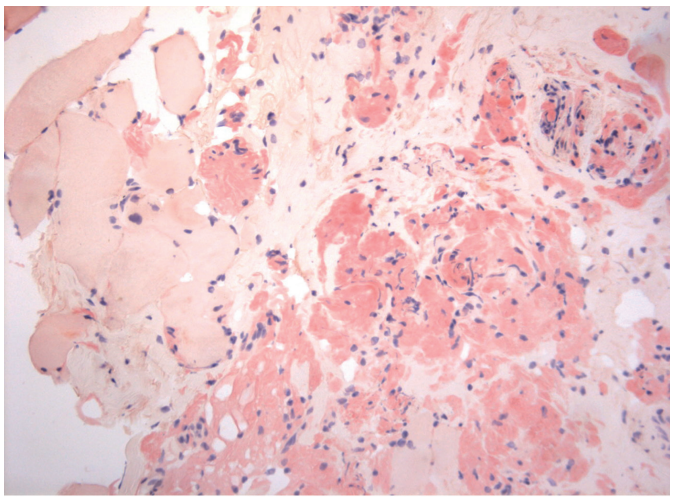

Fig. 1.
CRx200 = Congo Red stain (non-polarised) show extensive perimysial amyloid deposits with perivascular accentuation (original magnification x200).

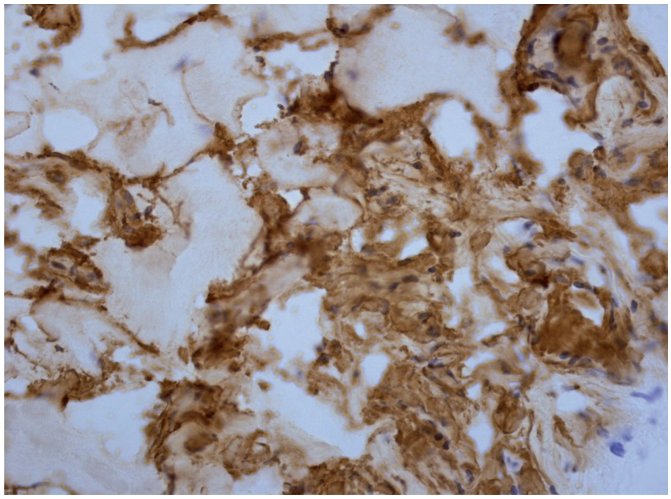

Fig. 2.
LLCx400 = Lambda light chain immunopositive amyloid deposits confirmed in peri- and endomysial stroma (original magnification x400).

Three months post-presentation, the patient became breathless and developed bilateral pedal oedema. An echocardiogram revealed a small left ventricular cavity with moderate concentric hypertrophy, and the echogenic appearance of the myocardium was consistent with cardiac amyloid. Further assessment and treatment at the National Amyloidosis Centre was sought. Bone marrow trephine confirmed amyloid of AL type (lambda subtype) and SAP scan showed muscle, liver, cardiac and renal involvement. Treatment with cyclophosphamide (5 days per week), dexamethasone (4 days every fortnight) and daily thalidomide was initiated. Unfortunately, despite serological response to this regimen, little clinical improvement has been observed.

The phenomenon of amyloid myopathy is rare and there are only few previous descriptions in the literature (1-3). Skeletal muscle amyloid deposition may be seen in patients with plasma cell dyscrasia with the patients classically exhibiting abnormal muscle firmness, pseudohypertrophy and macroglossia (4). However, an alternative phenotype has also been described which occurs more commonly in patients with AL amyloidosis, and as in our case is characterised by proximal muscle weakness and progressive atrophy (4, 5). The diagnosis can be confirmed using Congo Red muscle biopsy staining as other diagnostic tools such as muscle MRI can be non-specific (6). Unfortunately, even once the diagnosis is made, prognosis is generally poor with median survival from diagnosis being less than 15 months (7).

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