Case Report

Renal amyloidosis and angioimmunoblastic lymphadenopathy

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Case report

In 1992 a 33-year-old Afro-Carribean male presented with pyrexia, generalized muscle weakness, and tenderness and significant proteinuria (> 2 g/24 h). He was known to carry sickle cell trait and took no medications. Clinical examination was unremarkable and investigations revealed a serum creatinine of 180 μmol/l and a creatinine kinase of 508 (normal range 24–195 U/l). A screen for autoantibodies, viral serology, and antibodies for HIV were all negative. Renal biopsy revealed acute tubular necrosis with IgA deposition in glomeruli and a muscle biopsy showed myopathic change. His symptoms of myalgia, fever, and heavy proteinuria settled spontaneously and no cause was found.

He then re-presented in July 1993, again with generalized myalgia and weakness, but had also developed alopecia, bilateral retinal vasculitis, anterior uveitis and painful subcutaneous nodules in both arms. Investigations revealed elevated plasma levels of immunoglobulin G and immunoglobulin M but ANCA, ANA and cryoglobulins were not detected. Serum creatinine was 63 μmol/l with a normal creatinine kinase and AST. Muscle biopsy revealed non-specific atrophy and a skin biopsy showed necrotising vasculitis. Serum angiotensin converting enzyme level was normal and a Kveim test negative. Renal angiography and coeliac axis studies were also normal. A diagnosis of systemic vasculitis was assumed and the symptoms responded to high dose steroid treatment (prednisolone 60 mg per day) which was tapered out over several weeks.

A year later he presented again with generalized myalgia, proteinuria 5 g/24 hour and right axillary lymphadenopathy. On this occasion, a screen for autoantibodies revealed a weakly positive ANA (1: 40), whilst C3 was elevated along with C-reactive protein (102 mg/l, normal range < 10). A chest X-ray showed interstitial shadowing and a thoracic CT identified pulmonary infiltrates and confirmed the presence of right hilar and axillary lymphadenopathy. Transbronchial biopsy demonstrated lymphoid infiltrates and a renal biopsy showed deposition of amyloid within glomeruli (Figure 1). Amyloid was confirmed to be of the secondary type by immunoperoxidase staining for amyloid A and by abolition of Congo red staining by potassium permanganate. Excision biopsy of an axillary lymph node revealed expansion of paracortical areas with immunoblasts and increased vascularization consistent with angioimmunoblastic lymphadenopathy (AIL) (Figure 2). A bone marrow biopsy showed no abnormality of lymphocytes.

He was prescribed a course of chemotherapy consisting of cyclophosphamide 750 mg/m², doxorubicin 50 mg, vincristine 1.4 mg and prednisolone 40 mg to prevent further impairment of lung function and abate the progression of his renal amyloidosis. Eight months

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Patients with AIL, as in many people with HIV, suffer from chronic idiopathic thrombocytopenic purpura, autoimmune haemolytic anaemia and perhaps autoimmune myeloid suppression. These manifestations are occasionally of such severity as to require chemotherapy [4].

AIL mainly affects people aged between 40 and 90 years. The development of lymphadenopathy may be preceded by rashes or a previous history of autoimmune disease, such as Sjogren’s syndrome, hypothyroidism, or pernicious anaemia, and patients can develop a lupus like syndrome with anti-DNA antibodies [3].

There have been 10 published cases of AIL with renal involvement, three of which were described before 1978. Prior to this date, diagnosis of AIL was unreliable. All the cases describe a proliferative glomerulonephritis and in some, granular deposits of IgG, C3, and IgM within glomeruli [5–10]. Renal amyloid has previously not been described. It could be postulated that the development of secondary amyloid is consistent with either that of an inflammatory response as part of the autoimmune state or a low grade malignancy, both of which have been hypothesized as the aetiology of AIL.

**References**


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**Fig. 2.** Axillary lymph node showing preservation of a few follicles, with expansion of the paracortical zone consistent with angioimmunoblastic lymphadenopathy. H&E × 20.