IgM Nephropathy in a Patient with Kimura's Disease

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Dear Sir,

Kimura's disease is a chronic granulomatous disease of unknown etiology presenting with soft tissue tumor-ous masses. Renal abnormalities, notably proteinuria and the nephrotic syndrome, are its major associated manifestations. We present here a patient with Kimura's disease and IgM nephropathy with relapsing steroid-responsive nephrotic syndrome.

A 25-year-old Chinese male developed a right cervicofacial lump, a biopsy of which showed eosinophil infiltration, lymphoid follicles, fibrosis and vascular proliferation. The diagnosis of Kimura's disease was made, and the mass was resected. Three years later he developed the nephrotic syndrome with proteinuria 19.7 g/24 h, serum creatinine 0.085 mmol/l (0.96 mg/dl), albumin 18 g/l, cholesterol 12.1 mmol/l (467.8 mg/dl), globulin 23 g/l, normal serum sodium, potassium, chloride and bicarbonate levels. There was no eosinophilia. Urinalysis showed no red blood cells, white blood cells or casts. His blood pressure was 120/80 mm Hg. Antinuclear antibody and rheumatoid factor were absent; complement components C3 and C4 were normal. IgG was decreased to 463 mg/dl (normal 700–1,850), IgA 308 mg/dl (normal 90–400), IgM 150 mg/dl (normal 50–300). IgE was raised to 1,918 IU/ml (normal less than 100). Renal biopsy showed minimal increase in mesangial cellularity and mesangial matrix. Immunofluorescence showed weak IgM and Clq mesangial staining. Electron microscopy showed subtotal foot process effacement and a few small parames-angial electron-dense deposits. Proteinuria decreased to 0.08 g/24 h within 2 weeks of starting prednisolone at 1 mg/kg/day. Steroid was tapered off after 12 weeks. In the following 19 months he had 2 more relapses of steroid-responsive nephrotic syndrome treated with 2 courses of oral prednisolone lasting 11 and 8 weeks, respectively. In January 1990, he had a 3rd relapse of the nephrotic syndrome which remitted again with steroid therapy. His renal function remained normal during the 27 months' follow-up.

Proteinuria has been detected in 12% of patients with Kimura's disease with 60% of them manifesting the nephrotic syndrome [1]. Renal pathology, which had been documented in 5 previous reports, consisted of membranous nephropathy, mesangial proliferative glomerulone-phritis, minimal-change disease and diffuse proliferative glomerulonephritis [1–5]. Most authors noted steroid responsiveness of the proteinuria, but no one had reported on the course and long-term renal prognosis. The clinical course in the present case resembles relapsing steroid-responsive minimal-change disease.

The occurrence of proteinuria before the appearance of cutaneous lesions in some cases of Kimura's disease makes it unlikely that the renal pathology is secondary to substances released from the soft tissue lesion. The lymphoid proliferation, eosinophilia and elevated serum IgE often associated with Kimura's disease suggest a disturbance in immune regulation, which may also account for the associated glomerulonephropathy. It had been proposed that in Kimura's disease an unknown antigen was responsible for the stimulation of mast cells to release IgE and initiate a type I hypersensitivity reaction [6]. Relapses of minimal-change nephrotic syndrome have also been linked to hypersensitivity [7]. It is therefore tempting to speculate that a similar hypersensitivity reaction may trigger the nephrotic syndrome as well as initiate the growth of the soft tissue lesion in a genetically predisposed individual thereby manifesting as Kimura's disease and its associated renal abnormality.

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