Association of IgA Nephropathy and Myasthenia gravis

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Dear Sir,

We were interested to read the recent article in Nephron by Miyazaki et al. [1] which described 3 patients in whom IgA nephropathy was associated with myasthenia gravis. In all 3 cases, myasthenia gravis appeared after the discovery of glomerulonephritis. We have also noted a patient with both of these conditions in whom myasthenia gravis and a subsequent total thymectomy preceded the IgA nephropathy.

A 25-year-old male presented with ocular myasthenia gravis in 1981 and was initially treated with steroids and azathioprine. There was a poor response to this regime and shortly thereafter a total thymectomy was performed with a good initial result. No thymoma was demonstrated. In 1986, he was first noted to have microscopic haematuria and hypertension and at that time was on oral steroids for an exacerbation of this myasthenic symptoms. He presented to a medical ward 1 year later with a vasculitic skin rash affecting his feet, calves, hands and forearms. He continued to have microscopic haematuria and remained hypertensive (blood pressure 170/130). He never noted macroscopic haematuria.

Investigation results were as follows: plasma creatinine 84 µmol/l, urinary protein 0.26 g/24 h, intravenous urogram normal; C3, C4 and C3d were all within the normal range. Autoantibodies were negative, serum IgG and IgM levels were normal but serum IgA level was mildly elevated at 4.54 g/l (normal range 1.25–4.25 g/l). Renal biopsy yielded twenty-six glomeruli of which two were globally sclerosed. The remainder were normal on light microscopy but electron microscopy and immunoperoxidase studies demonstrated large mesangial IgA deposits with smaller amounts of C3 in a similar distribution and in small blood vessels. A punch biopsy of skin included a purpuric macule 0.4 cm in diameter. This biopsy showed severe acute neutrophilic vasculitis largely confined to upper dermal vessels. In the mid-dermis a larger muscular artery demonstrated neutrophilic vasculitis and fibrinoid necrosis of the walls. The skin rash slowly subsided spontaneously but the patient continues to require antihypertensive medication. The most recent plasma creatinine was 93 µmol/l.

This individual was thought to have both IgA nephropathy and a vasculitis of the skin. It is also possible that the skin rash and renal lesion are both manifestations of Henoch-Schönlein purpura. Although the skin changes are in keeping with this diagnosis, the renal lesions show no evidence of vascular disease and are characteristic of IgA nephropathy. IgG and IgM are absent and there are no basement membrane deposits such as would be expected in Henoch-Schönlein purpura. As the microscopic haematuria was identified 1 year before the clinical manifestations of skin vasculitis this would seem to be a separate unrelated...
entity. To our knowledge, however, there have been no reports of Henoch-Schönlein purpura in association with myasthenia gravis. In contrast to the cases reported by Miyazaki et al. [1], this case demonstrates myasthenia gravis and total thymectomy preceding IgA nephropathy. It is possible to speculate on a common T cell abnormality responsible for myasthenia gravis, IgA nephropathy and vasculitis. The previous total thymectomy makes a common link difficult to sustain although this may itself be involved in the aetiology of the skin and renal lesions as total thymectomy results in a decrease in the function of some subpopulations of T lymphocytes [2].

References