Case Report of IgA Nephropathy Associated with Ankylosing Spondylitis and HLA-B27

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Dear Sir,

A 32-year-old male patient with concurrent IgA nephropathy and ankylosing spondylitis is described. He had HLA-B27 antigen. Renal involvement in seronegative spondylarthropathies is considered to be very rare. Eighteen such cases reported are reviewed, and a possible common genetic or other pathogenesis discussed.

Case Report

The patient was a 32-year-old Japanese male who was admitted to hospital because of visual disturbance and mild headache. Positive physical findings on admission were a blood pressure of 270/180 mm Hg, a tachycardia of 140 beats/min, grade IV retinopathy, and scotoma of the left eye. Urinalysis revealed 70–80 RBC/HPF, a few hyaline casts and proteinuria of 3.0 g/24 h. Laboratory studies revealed an elevated sedimentation rate (ESR) of 31 mm/h, BUN 22 mg/dl, serum creatinine 2.2 mg/dl, a 24-hour creatinine clearance of 36 ml/min and positive HLA-B27. Plasma renin activity was 5.7 ng/ml/h. Electrolytes and blood gases were within normal limits. The hematocrit was 36%, hemoglobin 12.2 g/dl with a normal WBC count and differential count. The platelet count was 223,000. Serum IgA concentration was 495 mg/dl which was higher than normal (120–450 mg/dl); the level of circulating immune complexes was 1.3 µg/ml (< 5 µg/ml). Chest x-ray showed no pulmonary edema and cardiomegaly. The ECG revealed sinus tachycardia and left atrial enlargement. X-ray of sacroiliac joints and spine showed changes typical of ankylosing spondylitis.

Malignant hypertension was diagnosed and the patient was rapidly treated, initially with nifedipine 30 mg/day, and then with captopril 100 mg/day; the diastolic blood pressure was controlled at approximately 80 mm Hg. The 24-hour creatinine clearance was between 30 and 40 ml/min. Aortography was performed; there was no evidence of aortitis or stenosis of renal arteries.

A renal biopsy demonstrated mild mesangial proliferative glomerulonephritis with focal tubulointerstitial damage. There were three glomeruli of which two showed mild proliferation of mesangial cell and matrix and one moderate mesangial proliferation. The glomerular basement
membranes were thin and smooth without shrinkage as is frequently seen in hypertensive patients. Focal
tubular atrophy with mononuclear infiltrations was observed in the interstitium. Diffuse granular
deposits of IgA(2 +), C3( +) and fibrinogen ( ±) were found in the mesangium by
immunofluorescence microscopy; (fig. 1); deposits of IgG and C1q could not be identified.

Discussion
The clinicopathological features in the previously described patients and in our patient with
nephritis associated with seronegative spondylarthropathies (18 cases) are summarized [1–7].
Almost all patients were males between 20 and 55, and had HLA-B27, proteinuria (~ 14.4
g/day), and complained of back problems 2–25 years prior to the diagnosis of nephritis. Four
patients (22%) progressed to chronic or end-stage renal failure [1,2,4,6]. Kidney biopsy was
performed in all cases. Eleven cases were diagnosed as typical IgA nephropathy, one as mem-
Fig. 1. Diffuse mesangial IgA deposition. × 300.
nephropathy might be expected to be more frequent if such a direct correlation existed. The glomerular IgA deposits in IgA nephropathy have been shown to be IgA originating from the mucosal surfaces [9], and this is unlikely to be associated with ankylosing spondylitis. Thus not only the synthesis or secretion of IgA due to the inflammatory process, but also clearance or immunological regulation of IgA in ankylosing spondylitis might be associated with the development of IgA nephropathy.

A strong association with HLA-B27 in patients with ankylosing spondylitis is well known. IgA nephropathy, on the other hand, has been shown to have an association with HLA-BW35 or HLA-B12, and more recently with HLA-DR4 [10]. Since HLA antigens are known to be associated with several immunopathological diseases, both IgA nephropathy and seronegative spondylarthropathies might be related to certain HLA complexes, but this requires confirmation by further systematic research work.

References