IgA Nephropathy in a Patient with Unilateral Renal Agenesis

S.	Shinobu	Waga
Y.	Yoshiki	Kakizaki
K.	Kazuhiko	Sugimoto
H.	Hiroshi	Tanaka
M.	Masaru	Yokoyama

Department of Pediatrics, Hirosaki University School of Medicine, Hirosaki City, Japan

Shinobu Waga, Department of Pediatrics, Hirosaki University School of Medicine, 5-Zaifucho, Hirosaki, 036 (Japan)

Dear Sir,

The clinical relevance of the observation that reduction of renal mass in rats promotes the development of proteinuria and progressive renal failure due to focal glomerular sclerosis (FGS) is controversial. Similar mechanisms were postulated in the patients with unilateral renal agenesis, oligomega-nephronia, or unilateral small kidney, but Novick et al. [1] suggest that patients with a solitary kidney had an increased risk for progressive nephropathy after partial nephrectomy. These observations suggest that patients with a solitary kidney need further loss of nephrons in the remaining kidney due to resection or other structural abnormalities to be comparable to the rat model. Therefore, exposure of glomerulonephritis to a solitary kidney may also be relevant, but there have been few reports. Since IgA nephropathy is a disease with mesangial proliferation, which may cause progressive renal insufficiency, it is interesting to know how IgA nephropathy in a solitary kidney influences the outcome of renal function, and how the disease itself is influenced by a solitary kidney. We describe a boy with agenesis of the right kidney who had IgA nephropathy associated with nephrotic syndrome.

A 9-year-old boy was found to have heavy proteinuria and hematuria on 27th July 1992. Proteinuria or hematuria had not been discovered by annual screening for urinary abnormality at school in the previous 4 years. On admission, he weighed 28 kg and, within a short time, gained 5 kg. Blood pressure was 114/40 mm Hg. No skin or mucosal lesions were detected. His testes

Fig. 1. Diameter of glomerulus in the patient and in age-matched controls with IgA nephropathy. Only glomeruli passing through the hilus on the section were measured. Mean diameter $\pm$ SD of each control is shown on the abscissa.

IgM, 126 mg/dl, and C3, 120 mg/dl. Other routine blood examinations and antibody titers related to glomerulonephritis were normal. Ultrasonogram, intravenous pyelogram, and enhanced CT scan revealed no evidence of the right kidney, but an enlargement of the left kidney (length 13.5 cm)

were not defective. Urine analysis showed proteinuria of 470 mg/dl and many sediments of red blood cells per high power field. Laboratory studies revealed the following: serum total protein, 4.9 g/dl; total cholesterol, 334 mg/dl; BUN, 17 mg/dl; creatinine, 0.8 mg/dl; IgG, 212 mg/dl; IgA, 146 mg/dl;

KARGER

E-Mail karger@karger.ch Fax + 41 61 306 12 34 <http://www.karger.ch>

©1996 S. Karger AG, Basel 0028-2766/96/0742-0479\$ 10.00/0

without distortion of calyces. A voiding cystourethrogram revealed no reflux. Prednisolone therapy at a dose of 40 mg/day (40 mg/m²) for 4 weeks did not affect his nephrotic syndrome. Light microscopic findings for open biopsy of the right kidney revealed that most of the 17 glomeruli obtained showed moderate mesangial proliferation with focal accentuation. Partial cellular crescent was found associated with segmental sclerosis in 2 glomeruli. Immunofluorescence studies revealed staining for IgA with intensity of (+++), IgG (+), IgM(+), Clq(±), C3(-), C4(-), C5(-), and fibrinogen (-). Thus, he was diagnosed as having IgA nephropathy. Prednisolone was tapered. By the beginning of January 1993, urinary protein gradually decreased to 120-250 mg/dl, restoring the serum level of total protein and total cholesterol. Prednisolone was ceased on 30 July 1995, when urinary protein level and hematuria improved to trace.

In our patient, histology of renal biopsy was indistinguishable from FGS, but nephrotic range of proteinuria and hematuria subsided during a 3-year observation, while trace amount of proteinuria remained without deterioration of renal function. One explanation for the residual proteinuria is that his IgA nephropathy may be in the way of complete remission, since the disease has the property of spontaneous improvement in some patients [2]. The other is that FGS may have been promoted by exposure of IgA nephropathy to his solitary kidney containing hypertrophied glomeruli. Bhathena et al. [3] demonstrated that patients with solitary kidneys associated with proteinuria and FGS had at least 1.75 times increases of mean glomerular diameters compared to controls, and that the functionally fully compensated kidney is characterized by the absence of FGS and by a 1.24 times smaller increase in glomerular mean diameter. In our patient, glomerular diameter was increased 1.27-1.53 times compared to controls, who have IgA nephropathy without kidney deficit and supposedly normal glomerular diameter (fig. 1). This indicates that the patient's glomerular enlargement had not reached its limits. Therefore, the residual proteinuria is likely to reflect the course of IgA nephropathy itself, although a longer period of follow-up including serial biopsy should be required to elucidate which possibility is more likely.

References

Novick AC, Gephardt G, Guz B, Steinmuller D, Tubbs RR: Long-term follow-up after partial removal of a solitary kidney. *N Engl J Med* 1991;325:1058-1062.

Kusumoto Y, Takebayashi S, Taguchi T, Ha-rada T, Naito S: Long-term prognosis and prognostic indices of IgA nephropathy in juvenile and in adult Japanese. *Clin Nephrol* 1987;28: 118-124.

Bhathena DB, Julian BA, McMorro BW, Baehler RW: Focal sclerosis of hypertrophied glomeruli in solitary functioning kidneys of humans. *Am J Kidney Dis* 1985;5:226-232.

Waga/Kakizaki/Sugimoto/Tanaka/ Yokoyama