Horseshoe kidney with mesangioproliferative glomerulonephritis and goiter

Salih Kavukçu[1], Barış Şahin[1], Mehmet Türkmen[1], Alper Soylu[1]
Banu Lebe[2], Atilla Büyükgebiz[1]

Departments of [1]Pediatrics and [2]Pathology, Dokuz Eylül University Faculty of Medicine, İzmir, Turkey

SUMMARY: Kavukçu S, Şahin B, Türkmen M, Soylu A, Lebe B, Büyükgebiz A. Horseshoe kidney with mesangioproliferative glomerulonephritis and goiter. Turk J Pediatr 2003; 45: 350-352.

Horseshoe kidney is a relatively common renal anomaly with which many structural and developmental anomalies have been shown to be associated. However, there are only a few case reports regarding the association of membranous glomerulonephritis and focal sclerosing glomerulonephritis in patients with horseshoe kidneys. We report a girl who was evaluated for hematuria and proteinuria, and found to have horseshoe kidney. Renal biopsy demonstrated mesangioproliferative glomerulonephritis. She also had simple diffuse goiter.

Key words: horseshoe kidney, glomerulonephritis, goiter.

Horseshoe kidney occurs in 1:500 live births. Urinary tract infections (UTI), hydronephrosis and calculi are well known complications of horseshoe kidney. Horseshoe kidneys are found in 7% of patients with Turner's syndrome[1]. Although literature includes many reports implying several structural and developmental anomalies associated with horseshoe kidneys, there are a few case reports of patients with membranous glomerulonephritis[2,3] and focal sclerosing glomerulonephritis[4]. There has been no report of a relationship between non-infectious inflammatory nephropathy and horseshoe kidney. We report simple diffuse goiter and mesangioproliferative glomerulonephritis in a girl with horseshoe kidney.

Case Report

An eight-year-old female patient was referred to our clinic for evaluation of proteinuria and hematuria. Her symptoms had begun 45 days earlier with coryza, cough and fever, followed by peripheral edema. Past medical history was marked by recurrent UTI and progressive enlargement of thyroid gland. Family history was marked by first-degree consanguinity of parents and goiter in the mother.

Physical examination was normal except for diffuse enlargement of thyroid gland, and moderate pitting edema of eyelids and extremities.

Dipstick testing of urine revealed a specific gravity of 1025, pH 6, protein (2+), leukocyte (1+) and erythrocyte (1+). Microscopic evaluation of urine sediment demonstrated 10-12 leukocytes, 8-10 erythrocytes and many bacilli per high power field. Complete blood count showed hemoglobin 11.9 g/dl, white blood cell 10,600/μl and platelets 626,000/μl. Biochemical evaluation of serum revealed blood urea nitrogen 15 mg/dl, creatinine 0.8 mg/dl, total protein 4.8 g/dl, albumin 2 g/dl, total cholesterol 171 mg/dl (normal: 122-209 mg/dl), and triglycerides 150 mg/dl (normal: 35-114 mg/dl). The results of other laboratory tests were as follows: erythrocyte sedimentation rate (ESR) 125 mm/h, ASO 44 IU/ml, IgG 544 mg/dl (normal: 608-1572 mg/dl), IgM 187 mg/dl (normal: 52-242 mg/dl), IgA 288 mg/dl (normal: 33-236 mg/dl), C3 175 mg/dl (normal: 72-195 mg/dl), C4 30 mg/dl (normal: 7-40 mg/dl), and anti-nuclear antibody (ANA) negative. HbsAg and anti hepatitis C virus (HCV) antibody were negative. Serum thyroid function tests showed free-T3 4.0 pg/ml (normal: 2.3-4.2), T3 1.7 ng/ml (normal: 0.6-1.8), free-T4 1.3 ng/dl (normal: 0.89-1.8), T4 6.2 μg/dl (normal: 4.5-12.6), and TSH 4.6 μIU/ml (normal: 0.35-5.5) with negative antithyroglobulin and antimicrosomal antibodies. Thyroid ultrasonography disclosed diffuse enlargement of thyroid gland. Urinary protein

excretion was 50 mg/m^2/h, and urine culture yielded Escherichia coli 10^5 colonies/ml. The abdominal ultrasonography and intravenous pyelography demonstrated the presence of horseshoe kidney. Voiding cystourethrography was normal. Urinary tract infection was treated with ceftriaxone, and prophylactic trimethoprim-sulfhamethoxazole treatment was started.

An open renal biopsy was performed revealing diffuse mesangioproliferative glomerulonephritis with the following features: On light microscopy, narrowing of Bowman's capsular space by mesangial proliferation, diffuse thickening of capillary basement membranes, and mild segmentation of some glomerular capillaries were observed (Fig. 1). Proximal and distal tubular epithelial cell cytoplasms demonstrated a granular appearance. Immunofluorescence study showed diffuse (2+) mesangial and membranous deposition of IgM and a small amount of IgG without IgA, C3, or C1q deposition. Prednisolone treatment was started and the clinical gins and symptoms of the patient improved, but she had two occasions of heavy proteinuria on follow-up. First relapse was treated with prednisolone. She is still under control with prednisolone and cyclophosphamide combined treatment after a second relapse.
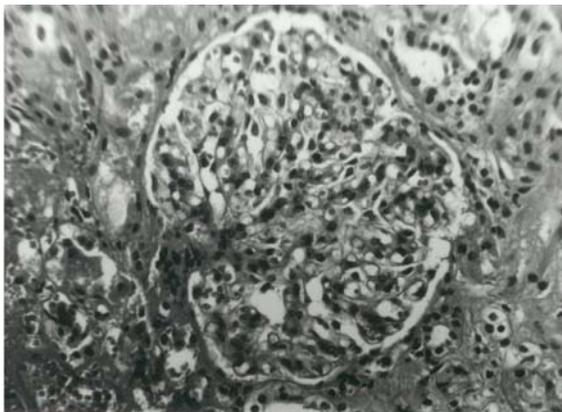

Fig. 1. Light microscopy showing diffuse thickening of the glomerular capillary walls and mesangial cell proliferation (hematoxylin and eosin stain x 400).

Discussion

The association of different glomerulopathies, including focal and sclerosing glomerulonephritis and hypothyroidism[4] and membranous glomerulopathy in horseshoe kidney were considered to be coincidence in adults[2,3]. Horseshoe kidney and simple goiter are common in the general population. Our case is unique in that the patient is in childhood and presented with the association of mesangioproliferative glomerulonephritis in horseshoe kidney and simple diffuse goiter.

Urinary tract infection in the absence of vesicoureteral reflux is common among girls. Our patient had a history of recurrent UTIs and a renal anomaly that could predispose her to UTIs. On the other hand, development of UTI during nephritic syndrome in this case suggests that this infection could also be due to the predisposition created by the nephritic state. Although vesicoureteral reflux was not demonstrated, concerning the nephritic syndrome and immunosuppressive treatment, a six-month course of antibiotic prophylaxis was planned for this patient.

Coexistence of simple goiter with other pathologies could be a coincidence in this patient. The rate of simple goiter in our area is a high as 6.25%[5]. Thus, given the high rate of horseshoe kidney in the population, its association with simple goiter could be regarded as a coincidence.

The mechanism of the association between the horseshoe kidney and inflammatory renal pathologies could not be explained in the previous reports. Horseshoe kidney is a structural and developmental anomaly. Wilms tumor has been reported to be seen four times more in this group of patients[1]. In addition, chromosomal abnormalities like Turner syndrome and trisomy 18 are also seen more commonly in patients with horseshoe kidney[6]. Non-infectious glomerular inflammatory pathologies have not been reported frequently in these chromosomal diseases or Wilms tumor. Furthermore, glomerulonephritis incidence has not been reported to increase in structural and developmental anomalies of the kidney like dysplasia or hypoplasia. Horseshoe kidney differs from the other anomalies and from the normal kidneys with respect to its size and blood supply. These characteristics could be speculated as being predisposing factors for immune complex accumulation. On the other hand, given the high rate of horseshoe kidney in the papulation (1/500), glomerulonephritis

should be more frequent in horseshoe kidneys in the presence of such a predisposition. However, there is no literature data indicating a high rate of glomerulonephritis in horseshoe kidneys. There are only a few case reports of horseshoe kidney with glomerulonephritis in the literature as mentioned before.

In conclusion, this case indicates that the response to the treatment of glomerulonephritis in horseshoe kidney does not differ from the glomerulonephritis in normal kidneys. In addition, the mechanism of the association of horseshoe kidney with glomerulonephritis should be evaluated and screened further, keeping in mind that it could just be coincidental.

REFERENCES

1. Elder JS. Urologic disorders in infants and children. In: Behrman RE, Keligman RM, Jenson HB (eds). Nelson Textbook of Pediatrics (16th ed). Philadelphia:WB Saunders Company; 2000: 1620.

2. Chen A, Ko W-S. Horseshoe kidney and membranous glomerulonephropathy. Nephron 1990; 54: 283.

3. Fujimoto S, Hirayama N, Uchida T, et al. Horseshoe kidney and membranous glomerulonephritis with cold activation of complement. Intern Med 1992; 31: 625-628.

4. Abson C, Jones M, Palmer A, Persey M, Gabriel R. Horseshoe kidney, focal and sclerosing glomerulonephritis and primary hypothyroidsm. Nephron 1991; 58: 124.

5. Ünak P, Darcan Ş, Yurt F, Biber Z, Çoker M. Determination of iodide amounts in urine and water by isotope dilution analysis. Biol Trace Element Res 1999; 71-72: 463-470.

6. Kass EJ, Bloom D. Anomalies of the upper urinary tract. In: Edelmann CM (Ed). Pediatric Kidney Disease (2nd ed) Vol. 2. Boston Little, Brown and Company; 1992: 2031.