A Young Patient with Horseshoe Kidney and Membranous Glomerulonephritis: Case Report

At Nalı Böbrek ve Membranöz Glomerülonefritli Genç Hasta

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ABSTRACT Horseshoe kidney is a fusion anomaly of kidneys. The horseshoe kidney does not produce symptoms, when present, symptoms are usually due to obstruction, stones, or infection. There are a few case reports of nephrotic syndrome in a horseshoe kidney. Here, we describe a case with a membranous glomerulonephritis in a horseshoe kidney. An 18 year-old-man was admitted to nephrology outpatient clinic for evaluation of pretibial edema. On the abdominal ultrasound horseshoe kidney was revealed, there were not stone or obstruction. Because the patient had nephrotic range proteinuria, hyperlipidemia, and hypoalbuminemia renal biopsy was performed. Renal biopsy was consistent with membranous glomerulonephritis. The patient was treated with methylprednisolon, cyclophosphamide, losartan and atorvastatin. There are two cases of idiopathic membranous glomerulonephritis with horseshoe kidney. We thought that the concurrent of two renal pathologies is a coincidence.

Key Words: Glomerulonephritis, membranous; kidney

ÖZET At nalı böbrek, böbreğin füzyon anomalisidir. Genellikle şikâyete neden olmaz, semptomatik olduğunda ise belirtiler genellikle obstrüksiyona, taş veya enfeksiyona bağlıdır. Nefrotik sendrom ile birlikte bulunan birkaç at nalı böbrek vakası vardır. Vakamız, at nalı böbrekte saptanan membranöz glomerulonefrittir. On sekiz yaşında erkek hasta pretibiyal ödem nedeniyle nefroloji polikliniğine başvurdu. Abdominal ultrasonografide at nalı böbrek saptandı, taş veya obstrüksiyon yoktu. Biyokimyasal değerlendirme sonrasında nefrotik sınırda proteinüri, hiperlipidemi, hipoalbüminemi olduğundan böbrek biyopsisi yapıldı. Biyopsisi sonucu membranoz glomerulonefrit ile uyumlu idi. Hasta metilprednizolon, siklofosfamid, losartan ve atorvastatin ile tedavi edildi. Literatürde bildirilen idiopatik membranöz glomerulonefritli iki at nalı böbrek vakası vardır. Bu iki renal patolojinin bir arada olmasının tesadüfi olduğunu düşünüyoruz.

Anahtar Kelimeler: Glomerülonefrit, membranöz; böbrek

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Horseshoe kidney is a fusion anomaly of kidneys, occurred in 1 in 400-1800 births and more common in males (2:1). Patients usually present with complication of reflux, obstruction or stone formation.[1] There are a few case reports of nephrotic syndrome in a horseshoe kidney. We describe a case is a membranous glomerulonephritis (MGN) in a horseshoe kidney.

CASE REPORT

An 18 year-old-man was admitted to nephrology outpatient clinic for evaluation of pretibial edema. He had no medical history of chronic systemic disease or drug exposure. On the physical examination, arterial blood pressure was 130/70 mmHg, pulse was 86/min, temperature was 36.5 °C and there were 3(+) pitting edema and ascites. Serum testing showed blood urea nitrogen 10 mg/dL, creatinine 0.69 mg/dL, sodium 140 mEq/L, potassium 4.6 mEq/L, calcium 8.1 mg/dL, phospfor 4.9 mg/dl, total cholesterol 326 mg/dL, LDL cholesterol 258 mg/dl, total protein 3.8 g/dL, albumin 1.3 g/dL, aspartate aminotransferase 14 IU/L, alanine aminotransferase 11 IU/L, alkaline phosphatase 80 IU/L, gamma-glutamyl transferase 7 IU/L, erythrocyte sedimentation rate 78 mm/hour, C reactive proteine 6 mg/L. His whole blood counts were; hemoglobine 12 g/dL, white blood cell count 10.9 K/mm^3 and platelets 234 K/mm^3. Examination of urine sediment showed 6-8 red blood cells, 1-2 white blood cells. Repeated urine and blood cultures were sterile. The patient's creatinine clearance rate was estimated at 109 mL/min and his 24-hour urine protein level was 8 g/day. Serological tests for anti-nuclear antibodies and for antibodies to cytomegalovirus, Epstein-Barr virus, hepatitis A, B and C viruses, and *Toxoplasma* spp. were negative. Complement levels were normal. On the abdominal ultrasound horseshoe kidney was revealed, there were not stone or obstruction (Figure 1). Renal biopsy was performed for diagnosis of nephrotic syndrome. No complications were observed after biopsy. Histopathological examination revealed thickening of glomerular basal membrane, expansion of the glomerulus and mesangial matrix, focal interstitial fibrosis, tubular epithelial edema and infiltration of mononuclear inflammatory cells on light microscopy (Figure 2, 3). Immunoflourescence study showed granular deposits of IgG and C3 but no deposits of IgA, IgM, and C1q along the glomerular capillary walls. Those finding were similar to MGN seen in patients without horseshoe kidney. Amyloid stain was negative. Electron microscopic evaluation could not be done because of the technical problems. According to that pathologic evaluations the diagnosis was consistent with MGN. Based on the clinical, laboratory and pathological findings, the patient was diagnosed with idiopathic MGN and treated with methylprednisolon (0.5 mg/kg/day), cyclophosphamide 500 mg/month, losartan 50 mg/day, atorvastatin 40 mg/day for six months. Methylprednisolon was gradually decreased and stopped within that time. Proteinuria decreased to 250 mg/day, serum albumin levels increased to 3.6 gr/dl and edema resolved. Losartan was maintained at dosage of 50 mg/day.

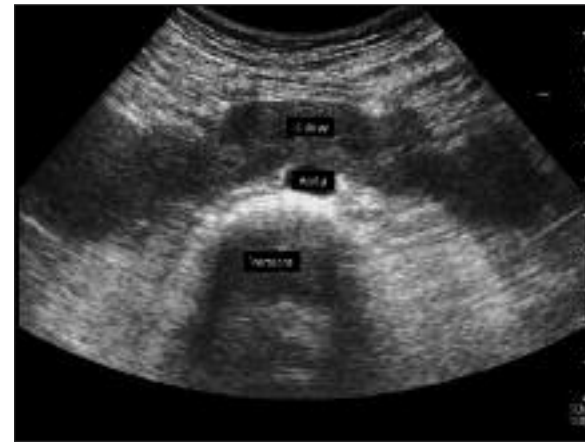

FIGURE 1: Ultrasonographic image of horseshoe kidney.

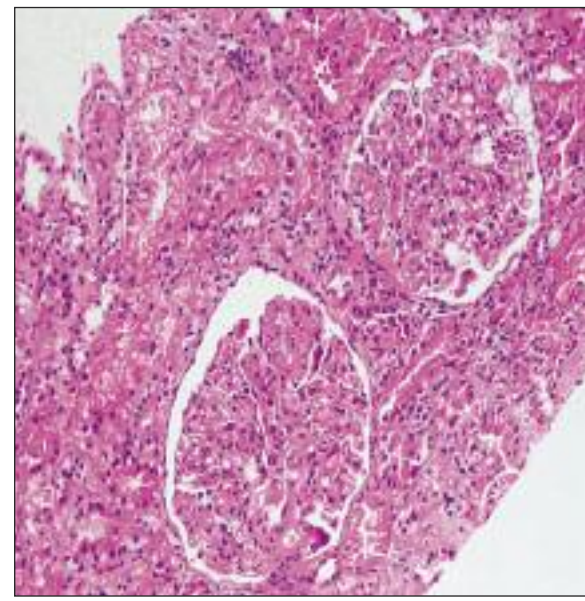

FIGURE 2: Thickening of glomerular basal membrane and expansion of the glomerulus. Hematoxylin eosin stain, magnification x200.

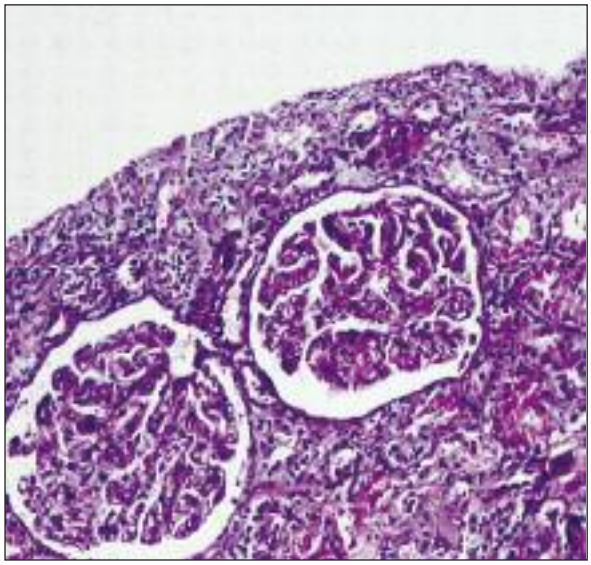

FIGURE 3: Thickening of glomerular basal membrane. PAS stain, magnification x200.

DISCUSSION

The horseshoe kidney is the most common type of renal fusion anomaly. No genetic determinant is known. The horseshoe kidney does not produce symptoms, when present, symptoms are usually due to obstruction, stones, or infection. The rates of hydronephrosis, stone formation, infection, and certain cancers (transitional cell cancer, Wilms and carcinoid tumors) are higher. Horseshoe kidney can be a feature of genetic disorders such as Turner syndrome, Trisomy 13, 18 and 21. Ureteropelvic junction obstruction occurs in up to 35% of patients. Stone formation range from 20-60%, which is associated with hydronephrosis or ureteropelvic junction obstruction that causes urinary stasis. The variable blood supply, presence of isthmus, high insertion point, abnormal course of the ureters contribute to these problems.[1,2] There is no information about whether the frequency of these complications increased in patient with nephrotic syndrome.

Membranous glomerulonephritis is one of the more common forms of idiopathic nephrotic syndrome in the adult population (30%). Immunologic disease (such as diabetes, lupus, rheumatoid arthritis), infectious and parasitic diseases, drugs, toxins can cause MGN. The pathologic features of MGN evolve from initial formation of subepithelial immuneo complexes of IgG and complement along the outer surface of the capillary wall. Changes first occur in the glomerular epithelial cells, then in the glomerular barrier function leading to proteinuria, then in the renal interstitium and finally in the glomerular basal membrane itself. Glomerular basal membrane becomes thickened through the accumulation of additional matrix material along the outer surface, often irregular, or spike-like pattern.[3]

There are several reports of horseshoe kidney with nephrotic syndrome. Those are amyloidosis, cryoglobulinemic membranoproliferative glomerulonephritis associated with hepatitis C virus infection, mesangial proliferative glomerulonephritis, focal and sclerosing glomerulonephritis and MGN.[4-12] Four MGN cases are reported, two of them were idiopathic. The first case was a twenty-year-old man and open renal biopsy was performed and treated with prednisolon.[9] The second one did not have proteinuria and biopsy was performed during hepatic hydatid cyst operation.[10] The other one was 48 year-old-woman with positive hepatitis C antibody and cold activation complement. In that case, viral antigenemia was suggested to be the cause of MGN.[11] The last one was 18-year-old woman with idiopathic MGN and was treated with methylprednisolon.[12] Our patient was treated with methylprednisolon (0.5 mg/kg/day) and cyclophosphamide 500 mg/month for 6 months. Proteinuria decreased to 250 mg/day, edema resolved.

Even though the frequency of stone, ureteropelvic junction obstruction, certain tumors are higher and horseshoe kidney can be a feature of some genetic disorders, it is early to say the increased frequency of MGN or whether MGN is one of the complications of horseshoe kidney. It is unclear if that anomaly can cause immune dysfunction or predispose to immune complex deposition in the glomerulus. There are only two cases of idiopathic MGN in the horseshoe kidney. More such cases will be necessary to conclude the causal relationship between these two conditions. Renal biopsy should be performed if there is not any contraindication in those patients with proteinuria. The concurrent of these two renal pathologies may still be a coincidence.

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