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Case Report
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Nephrotic Syndrome in Horseshoe Kidneys - An Emerging Association?

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Introduction

Horseshoe kidneys are a frequent yet enigmatic urological malformation. Some frequently associated complications include UPJ obstruction, recurrent urinary tract infections, lithiasis and tumors. The occurrence of nephrotic lesions in horseshoe kidneys is extremely rare with a very few reported and biopsy proven cases. We report a case of an 18 year old boy who was initially diagnosed with hydronephrosis associated with horseshoe kidneys and then 3 years later developed Nephrotic syndrome, with biopsy findings of Focal Segmental Glomerulosclerosis.

Case Presentation

Initial presentation

In this case report, a 14 year old Asian male child presented to us in September, 2017 with the complaints of pain in the left lumbar region for the last 1 month. With the lab workup unremarkable and an ultrasonographic scan was obtained which revealed a grade 3 hydronephrosis. An IVP performed suggested of horseshoe renal anatomy with grade 2 hydronephrosis on the right side and dilated renal pelvis on the left side with bilaterally normal excreting kidneys. A non-contrast CT scan was obtained which suggested presence of horseshoe kidneys with bilateral hydronephrosis (Right more than Left) along with UPJ obstruction. ^{99m}Tc scan was suggestive of slow draining right kidney. The child was managed for his symptoms and advised follow up after 6 months.

First diagnosed with nephrotic syndrome

The child presented 3 years later (August, 2020), at 17 years of age with the complaints of periorbital edema for the past 1 week and abdominal dissension for the last 5 days. Physical examination showed a blood pressure of 130/90 mmHg, with the other vitals being stable.

Laboratory work up was ordered and revealed the following: Hemoglobin 13.5 g/dL, TLC 7.2 m/mm^3, Fasting Blood Sugar 100 mg/dL, Erythrocyte Sedimentation Rate 55, Blood Urea Nitrogen 19 mmol/L, Serum Creatinine 0.8 mg/dL, Serum Na+ 133.6 mEq/L, Serum K+ 4.33 mEq/L, Uric Acid 5.5 mmol/L, SGOT/SBPT 38/26, Total protein 3.3 g/dL, Albumin 1.3 g/dL, Cholesterol 341 mg/dL, Triglycerides 193 mg/dL, LDL 255 mg/dL, HDL 74 mg/dL, Urine Protein+++, Urine Culture Sensitivity-Sterile, ANA-Negative, C3 & C4-Normal Levels, [HIV, Hep B, Hep C] - Non Reactive. Chest X-ray - NAD, Montoux-Non reactive, CBNAAT for *Mycobacterium tuberculosis* - Not Detected. The blood coagulation function of the patient was normal.

A diagnosis of Nephrotic Syndrome was established after infections and secondary causes were ruled out. An ultrasonography was performed, revealing gross ascites and horseshoe kidneys with parenchymatous band and bilateral hydronephrosis. Patient was initiated treatment with prednisolone at 60 mg/kg/m^2 and within 3 days the patient responded adequately. He was discharged on 6 weeks (daily 60 mg/kg/m^2) + 4 weeks (alternate 40 mg/kg/m^2) regimen. The patient defaulted at around 5 weeks into the treatment.

Relapse and biopsy confirmation

The child presented again in July, 2021 with similar complaints of periorbital edema and abdominal dissension. Investigations again revealed hypoalbuminemia, hyperlipidemia and +++ proteinuria, suggestive of nephrotic syndrome. The blood pressure was recorded to be 136/90 mmHg with other vitals being stable. Under ultrasound guidance, a renal biopsy was performed using a Renal Biopsy Gun. H&E, PAS, MT, Silver Methenamine and Congo red sections were studies.

Multiple sections included renal medulla and cortical parenchymal area containing up to 21

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Table 1: Previously published data on glomerular diseases occurring in horseshoe kidneys.

Authors	Sex	Age	Serum Creatinine	Urine Protein	Pathological Findings
Chen et al. [7]	M	20	0.9 mg/dL	300 mg	Membranous glomerulonephropathy
Abson et al. [8]	M	52	1.5 mg/dL	7.7-14.4 g/24h	Focal segmental glomerulosclerosis
Fujimoto et al. [9]	M	48	0.7 mg/dL	0.6 g/24h	Membranous glomerulonephropathy
Matyus et al. [10]	M	38	210 µmol/L	9 g/24h	Membranoproliferative glomerulonephritis
Alagözlü et al. [11]	F	18	0.8 mg/dL	8-14 g/24h	Membranous glomerulonephropathy
Kavukcu et al. [12]	M	8	0.8 mg/dL	50 mg/m2/h	Mesangioproliferative glomerulonephritis
Kayatas et al. [13]	M	23	0.9 mg/dL	9.2 g/24h	Focal segmental glomerulosclerosis
Kayatas et al. [13]	M	28	0.8 mg/dL	5.8 g/24h	Renal amyloidosis
Rivera et al. [14]	M	38	1.6 mg/dL	18 g/24h	Focal segmental glomerulosclerosis
Hu et al. [15]	F	15	66.9 µmol/L	1.7 g/24h	Henoch-Schonlein purpura nephritis
Hu et al. [15]	M	26	108.2 µmol/L	1.4 g/24h	Immunoglobulin A nephropathy
Chaabouni et al. [16]	M	22	95 µmol/L	22 g/24h	Minimal Change Disease
Shi SS et al. [17]	M	48	75 µmol/L	7.54 g/24h	PLA2R-Positive Membranous Nephropathy

glomeruli. None globally sclerosed. The glomeruli revealed focal dilation and congestion of capillary lumina. 6 glomeruli revealed segmental tuft sclerosis with intraglomerular foam cell change (Table 1). On immunostaining, IgA, IgG, IgM, C3, C1q, Kappa Light Chain or Lambda Light Chain depositions were not found. There was no evidence of crescent formation, capillary wall thickening, tuft necrosis, congophillic deposits in the visualized glomeruli. The findings were suggestive of Focal Segmental Glomerulosclerosis involving 6/21 of sampled glomeruli.

The patient was re-initiated on daily prednisolone therapy at 60 mg/kg/m^2 and went into remission after 8 days. DVT prophylaxis, Rosuvastatin and Enalapril were also prescribed and the patient was discharged.

Discussion

One of the most frequent fusion malformations is horseshoe kidneys (1:400 people; twice as common in male as in females). Although it has been observed among monozygotic twins and siblings within the same family, no genetic factor has yet been found. Horseshoe kidneys are indolent in and of themselves, but because to their embryogenesis and anatomy, they are prone to a greater rate of illness than regular kidneys. It is characterized by fusion at the lower poles by parenchymatous or fibrous isthmus, resulting in malrotation. Midline fusion occurs in up to 42% of instances, with lateral fusion occurring in 58 percent of cases, of which 70% are left dominant. This further leads to incomplete cephalic migration of the kidneys as during ascent, the fused lower poles become trapped under the inferior mesenteric artery. Hence, the isthmus lies at L3-L4 vertebral level, at the origin of IMA from the abdominal aorta. The pelvis and ureters are usually anteriorly/anteriomedially placed with the calyces pointing posteriorly. Associated ureteral compression is common (>35%) occurring due to anteriorly displaced ureter or from aberrant vessels leading to hydronephrosis, calculi formation and increased propensity for infections.

A horseshoe kidney with nephrotic syndromes is extremely rare. Primary causes of nephrotic syndrome include minimal change disease, focal segmental glomerulosclerosis, membranous glomerulonephritis; it can also be due to secondary causes such as drugs, vasculitis, amyloidosis, diabetes, allergens and toxins.

In our patient, a renal biopsy was performed and studied by experienced doctors revealing multiple non-global segmental sclerotic changes in the glomeruli. A diagnosis of Focal Segmental Glomerulosclerosis was made and the patient was started on prednisolone, to which he responded adequately. Further Rosuvastatin and Enalapril were also prescribed to manage hyperlipidemia and proteinuria respectively [1-6].

Conclusion

Previous studies were unable to elucidate the specific mechanism between the two pathologies and there is no evidence in the literature that horseshoe kidneys have a high rate of glomerulonephritis. The co-occurrence of two renal disorders in this patient could be just coincidental, but a study by Rivera F. et al, concluded that horseshoe kidneys could be associated with renal adenocarcinoma and nephrotic syndrome with FSGS, secondary to hyper-filtration that progresses to chronic renal failure. Further studies are needed to be certain; meanwhile one should keep in mind the possibility of renal disorders other than reflux nephropathy in patients with horseshoe kidney.

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