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Case Report

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Rare Cause of Anuria in a Patient with Horseshoe Kidney

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ABSTRACT

Horseshoe kidney is a common congenital renal fusion anomaly. Acute kidney injury occurs secondary to urinary infection or obstructive uropathy, but glomerular involvement is rarely reported. A 47-year-old female presented with symptoms of urinary infection and was hospitalized in the urology clinic. Investigations revealed renal dysfunction and active urine sediments. The patient did not respond to antibiotics and eventually became anuric after 3 weeks of initiation of symptoms. Abdominal ultrasound was suggestive of horseshoe kidney. Workup for rapidly progressive glomerulonephritis and kidney biopsy confirmed the diagnosis of anti-glomerular basement membrane disease. The patient responded to immunosuppression and plasmapheresis. The above case highlights the importance of timely renal biopsy in diagnosis and management of glomerular disease even in patients with horseshoe kidney.

Keywords: Horseshoe kidney, glomerulonephritis, anti-glomerular basement membrane disease, renal biopsy

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INTRODUCTION

Horseshoe kidney (HSK) is the most common renal fusion anomaly with an incidence ranging from 0.04 to 0.16 per 1000 births.[1] It is associated with complications like urinary tract infections, hydronephrosis, stones, and tumors; however, the occurrence of glomerular diseases has been scarcely reported in the literature.[1-3] This may be due to increased risk associated with the biopsy of HSK due to abnormal structure and complex relationship between HSK blood vessels and adjoining great vessels.

The glomerular diseases described in the literature with HSK are immunoglobulin A (IgA) nephropathy,[3] membranous nephropathy,[4,5] focal and segmental glomerulosclerosis,[6] and membranoproliferative glomerulonephritis.[7] To the best of our knowledge, the occurrence of anti-glomerular basement membrane (anti-GBM) disease in a patient with HSK has not been reported in the literature.

CASE PRESENTATION

A 47-year-old female was admitted in urology in view of symptoms of fever, dysuria, and urgency for 1 week. On admission, she had hemoglobin (Hb) 9.8 gm/dL and leukocytosis (25 000 white blood cells (WBC)/µL) with the following renal parameters: blood urea nitrogen (52 mg/dL) and serum creatinine (1.24 mg/dL). Urinalysis revealed 20-30 WBC/high-power field (HPF), 40-50 red blood cells (RBC)/HPF, and few granular and RBC casts with urine protein 2+. In view of symptoms and pyuria, urine culture which grew *Escherichia coli* was sent (>10 000 colony-forming units/mL) and was initiated on intravenous piperacillin tazobactam as per sensitivity which was later escalated after 5 days to meropenem in view of the deterioration in renal functions. The patient had rapidly progressive renal failure and investigations repeated after 12 days revealed increased blood urea nitrogen (162 mg/dL) and creatinine (7.1 mg/dL) along with low Hb (7.1 g/dL), and total leucocyte count (TLC)

(18100/mL). The patient did not respond to antibiotic treatment and eventually became anuric after 3 weeks of initiation of symptoms.

Ultrasound examination of the abdomen revealed medially directed lower poles of both the kidneys and a band of renal tissue anterior to the aorta, connecting both the kidneys suggestive of HSK. The findings were confirmed by computed tomography (CT) of the abdomen (Figure 1A and B) which did not reveal any other features of obstruction or pyelonephritis. Her chest radiograph was unremarkable.

In view of rapidly progressive renal failure with anuria and active sediments, serological workup was done for the same which revealed normal C3 and C4 and was negative for antinuclear antibodies, anti-neutrophil cytoplasmic antibodies, hepatitis B surface antigen, hepatitis C virus, and human immunodeficiency virus. However, anti-GBM antibody titers were elevated (106.8 U/mL). She underwent renal biopsy following stabilization (2 sessions of hemodialysis and packed cell transfusion) from the left upper pole of kidney under ultrasound guidance. The procedure was uneventful. Light microscopy revealed cellular crescents in 12 out of 16 glomeruli (Figures 2 and 3). Immunofluorescence microscopy showed linear IgG deposits suggestive of anti-GBM disease.

The patient was managed with supportive care including hemodialysis, packed cell transfusion, and antibiotics. She was treated with 9 sessions of single-volume plasmapheresis, 5 pulses of injection methylprednisolone (500 mg) followed by oral prednisolone (1 mg/kg/day) and oral cyclophosphamide (2 mg/kg/day). Her anti-GBM titers became undetectable after plasmapheresis.

The patient responded to immunosuppression and her renal functions settled at creatinine of 1.8 mg/dL after 3 months of therapy. Currently, she is on low-dose prednisolone (5 mg/day) and her anti-GBM titers are negative.

MAIN POINTS

- The most important is incidental detection of horseshoe kidney (HSK) disease and the first-ever case report of anti-GBM condition in such a patient.
- A high index of suspicion was kept for RGPN given urinary sediments with persistent anuria, dialysis dependency, and kidney biopsy, which eventually led to the diagnosis.
- Kidney biopsy is usually avoided in patients with HSK with the difficulty and the possibility of complications. This case highlighted the importance of timely kidney biopsy and was started on immunosuppression based on cellular crescents findings despite being dialysis-dependent. The treatment prevented permanent kidney damage, and she became dialysis independent.

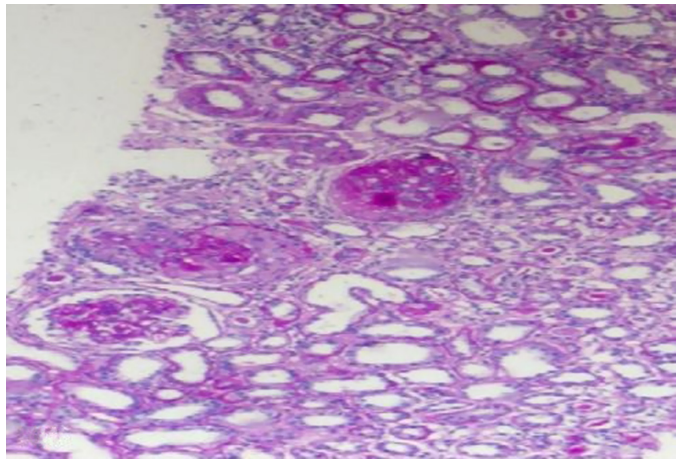

Figure 2. Pathological findings of the renal biopsy. Light microscopy showing 3 glomeruli with a cellular crescent. The tubulointerstitial compartment shows interstitial inflammation and acute tubular injury. (PAS stain, magnification 10×). PAS, periodic acid–Schiff.

Informed consent from the patient has been taken for the case report.

DISCUSSION

Horseshoe kidney is the most common renal fusion anomaly, with a prevalence of 0.25% among the general population.[8] It consists of kidney fusion across the midline. Horseshoe kidney can present with a wide variety of associated anomalies. The most frequent include ureteropelvic junction anomalies, stones, infections, and a variety of benign and malignant tumors.[1,8] Glomerular diseases in HSK have scarcely been reported in the literature due to the increased risk associated with renal biopsy. There have been case reports in the literature where the association between glomerular diseases and HSK has been linked to either coincidence or possibility that HSK facilitates immune complex deposition. Although left renal pole is the preferred site for kidney biopsy, in HSK, 90% of the fusion occurs in inferior poles and since the bilateral lower renal poles are closer to abdominal aorta, there is increased risk of renipuncture and bleeding during the kidney biopsy. Since the anatomical and vascular abnormalities like accessory renal arteries make renal biopsy difficult in HSK, careful evaluation should be done including abdominal CT angiography if necessary to rule out abnormal vasculature. Thus, upper poles of the kidney should be preferred for biopsy in HSK and the biopsy should be done under ultrasound guidance by experienced doctors.[3,5]

Our patient also had HSK. She presented with rapidly progressive glomerulonephritis (RPGN) as evident by the renal deterioration and progression to anuria along with active urinary sediments within a span of 2 weeks. Serological workup for the same revealed elevated anti-GBM antibody. Doppler studies of renal vessels and CT scan were done to rule out abnormal great

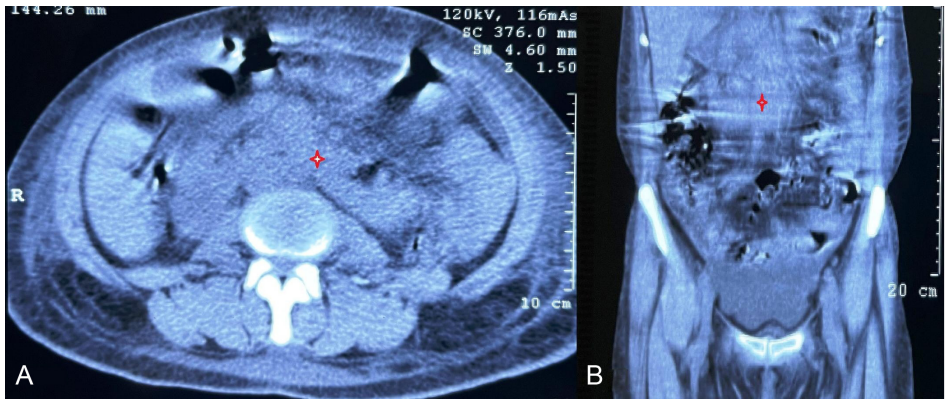

Figure 1. (A) Non-enhanced axial CT scan of the abdomen of the patient showing an HSK with isthmus (highlighted) lying anterior to the aorta and (B) depicting a coronal section with isthmus of HSK highlighted. CT, computed tomography; HSK, horseshoe kidney.

vessel around HSK, following which renal biopsy from the left upper pole was performed.

Our patient had many poor predictors of renal survival including high serum creatinine and the need for dialysis during the hospital stay. Current Kidney Disease: Improving Global Outcomes guidelines do not recommend immunosuppression in patients with the anti-GBM disease who are dialysis-dependent, have 100% crescent in a biopsy, and do not have a pulmonary hemorrhage.[9] In spite of the complexities associated, kidney biopsy was performed as histopathology is the gold standard in the diagnosis of RPGN. Apart from diagnosis, the biopsy also helps in deciding the level of immunosuppression to be given along with assessing the long-term prognosis of the patient. The kidney biopsy showed a predominantly cellular crescent which raised the hope of salvaging renal function. Fortunately, the patient responded to treatment with immunosuppressive therapy and plasmapheresis and became dialysis-independent.

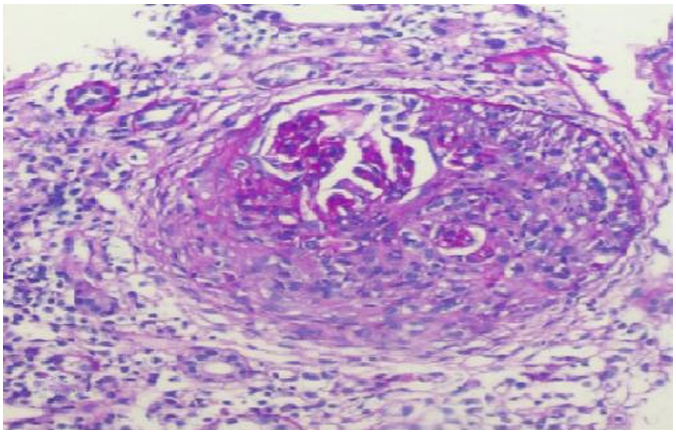

Figure 3. High magnification shows 1 glomeruli with complete cellular crescent, compression of glomerular tuft, and breakage of Bowman capsule with sunburst appearance (PAS stain, magnification 40×). PAS, periodic acid–Schiff.

Whether the association of HSK with RPGN and anti-GBM presentation is a mere coincidence or HSK can predispose to glomerulonephritis is yet to be established. Since only a few patients with HSK receive a renal biopsy, there is a lack of evidence elucidating the causal relationship between glomerulopathy and HSK.

To conclude, this is the first case of RPGN with anti-GBM disease occurring in a patient with HSK. This case highlights the importance of doing a timely renal biopsy and early initiation of therapy to achieve a better outcome.

Informed Consent: Informed consent was obtained from the patient.

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