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The Incidental Finding of Horseshoe Kidney in a 31-Year-Old Male Patient with Lupus Nephritis

Khurram Khaliq Bhinder[1], Nishwa Azeem[2], Azza Sarfraz[3], Sameer Saleem Tebha[4], Humaira Bashir[1], Zenab Farooq[1] and Zouina Sarfraz[5*]

[1]Shifa International Hospital, Islamabad, Pakistan.
[2]Lahore General Hospital, Lahore, Pakistan.
[3]Aga Khan University, Karachi, Pakistan.
[4]Jinnah Medical and Dental College, Karachi, Pakistan.
[5]Fatima Jinnah Medical University, Lahore, Pakistan.

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

We describe the case of a 31-year-old-male who presented with an acute flare of SLE, gross hematuria, left lumbar pain, fever, and burning micturition. He had a history of systemic lupus erythematosus, hypertension, myocardial infarction, and weight loss. Radiological examinations with ultrasound and computed tomography imaging revealed fusion of the lower poles of the kidneys rotated at the renal hilum forming a U-shape, in addition to a definite diagnosis of lupus nephritis. The patient subsequently underwent three sessions of plasmapheresis. The final diagnosis was consistent with a co-morbidity of horseshoe kidney and lupus nephritis in the adult male patient who was discharged and advised regular follow-up.

Keywords: Horseshoe kidney; systemic lupus erythematosus; lupus nephritis; Pakistan; LMIC.

**Corresponding author: E-mail: zouinasarfraz@gmail.com;*

1. INTRODUCTION

Horseshoe kidney is a common developmental anomaly due to fusion defect of the kidneys with an incidence of 1:500 with a male predominance [1]. In this defect, both kidneys are fused at the poles, leading to a parenchymal or fibrous isthmus. Before the ascent, both kidneys join in the pelvic cavity, forming a singular and distinct U-shaped kidney. Although the horseshoe kidney is compatible with life and largely remains asymptomatic, it is associated with multiple vascular and genitourinary anomalies [2]. Systematic lupus erythematosus (SLE) is an autoimmune disease that leads to chronic inflammation and damage to more than one organ, notably leading to lupus nephritis (LN) upon kidney involvement. LN is found in 30% of patients diagnosed with SLE [3]. Risk factors include the male gender, racial ethnicities (Asians, Blacks, and Hispanics). To our best understanding, we describe the first co-presentation of horseshoe kidney and lupus nephritis in an adult male patient.

2. CASE REPORT

A 31-year-old male patient with a history of systemic lupus erythematosus (SLE), hypertension (HTN), myocardial infarction (MI), and weight loss presented to the emergency department with an acute flare of SLE, gross hematuria, left lumbar pain, fever, and burning micturition. Urinalysis was performed to rule out infectious etiology. Cystoscopy results were unremarkable.

Left lumbar pain of the patient was acute in onset, persistent, and aching in character. It was temporarily relieved by NSAIDs, was associated with fever which was acute in onset, low grade, intermittent, and relieved by antipyretic medication. The pain was also associated with burning and painful micturition. The patient reported no history of anorexia, sleep disturbances, cough, nausea, vomiting, diarrhea, diabetes, asthma, hepatitis, hematemesis, or melena. The family's health history revealed no congenital or genetic disease. The social and personal history was unremarkable.

On examination, the patient was of average built and height, vitals were stable with pedal edema and mild periorbital puffiness. The cardiovascular, respiratory, and gastrointestinal examination was unremarkable. Complete blood count examination revealed mildly reduced hemoglobin parameters, with an otherwise normal report. Renal function tests showed urea within normal limits, but mildly decreased creatinine was noted. The computed GFR based on the formula GFR (mL/min per 1.73 m2;1.21) = 186.3 x Serum Creatinine (exp[-1.154]) x Age (exp[-0.203]) x revealed a value of 151 mL/min/1.73m. Urine R/E showed WBCs: 15-20, blood ++, leukocyte esterase ++, proteins +++; proteinuria (1.9 g/24h) was quantified on the second day post-hospital admission. C-reactive protein was 21.5 (it decreased gradually after commencement of treatment; it reduced to 6 on the 5th day of treatment). PT, APTT, and INR values were within the normal range. On Urine culture and sensitivity examination, gram staining showed no organism; the initial culture at 24 hours showed no growth.

A biopsy of the kidney was performed using H&E stain, which revealed crescent formation, necrotizing lesions, the proliferation of mesangial cells, and segments with endocapillary proliferation. Notably, the kidney biopsy, being the gold standard for the diagnosis of lupus nephritis confirmed SLE in the patient. Post-biopsy ultrasound KUB revealed no hemorrhage or fistula formation, both kidneys appeared normal with the right kidney measuring 9.6 X 5.2cm and the left kidney measuring 9.9 X 4.5cm (Fig. 1). No fluid collection was noted in the left perinephric region. The urinary bladder was empty.

The non-contrast CT abdomen and pelvis was performed due to aggravation of urinary symptoms, which incidentally revealed fusion of lower poles of the kidneys, rotated with renal hilum posteriorly, and the kidney was U-shaped without any hydronephrosis or dilatation (Fig. 2).

A definite diagnosis of Lupus nephritis was made. Plasmapheresis using a double lumen catheter was passed in the right internal jugular vein, under ultrasound guidance. Post incretion, a chest X-ray (CXR) was performed which had no abnormalities and the catheter was in place. A total of three sessions of plasmapheresis were performed with no subsequent complications. Infusion of endoxin (Cyclophosphamide) was started after the first session of plasmapheresis that was well tolerated by the patient. Clopidogrel 75mg started 5 days post-biopsy. The second infusion of endoxin (Cyclophosphamide) was planned 14 days later. The patient was prescribed medications for home use and discharged with the advice of regular follow-up.

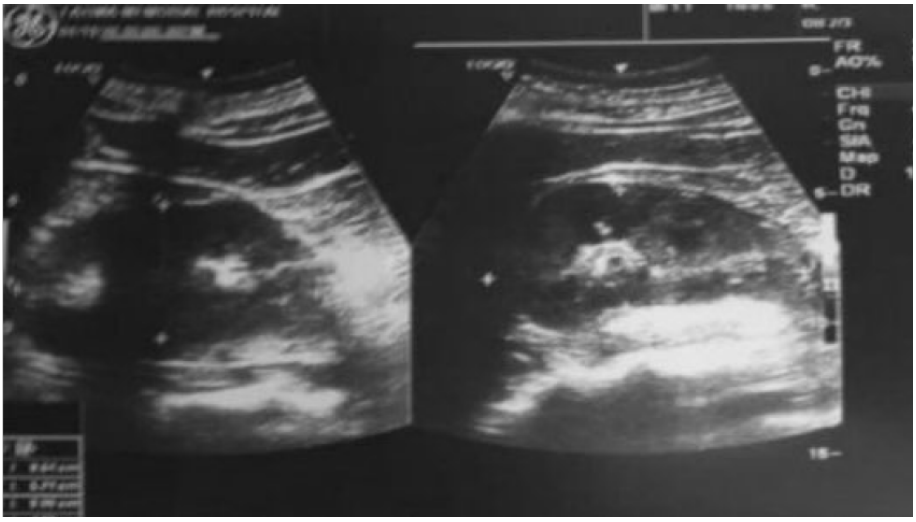
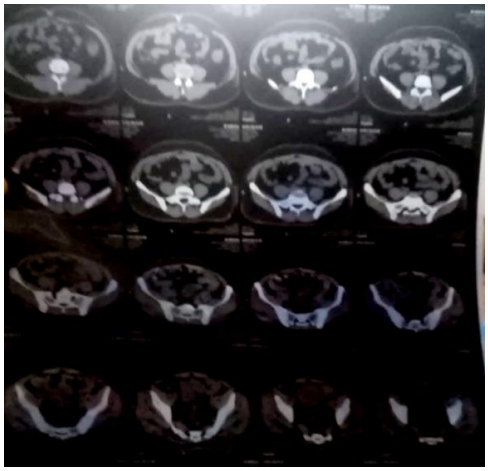


Fig. 1. Post Biopsy Ultrasound. The imaging revealed no fistula or hemorrhage formation with normal-appearing kidneys



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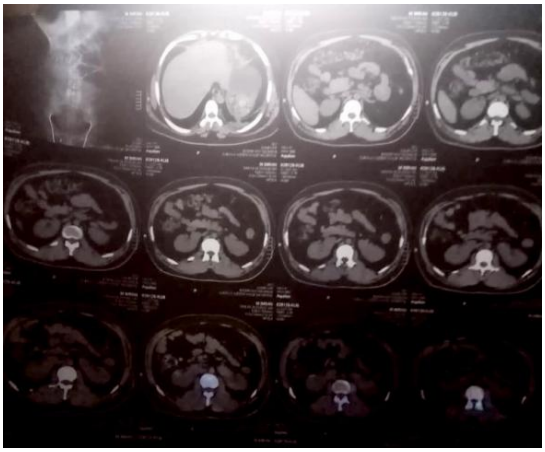

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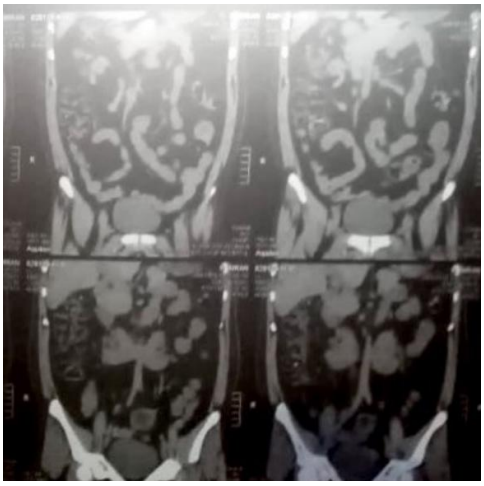

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Fig. 2. Computed Tomography (CT) scan findings. The CT abdomen and pelvis revealed a fusion of the lower poles of the kidneys that are rotated with the renal hilum posteriorly. The U-shape is visible, without the presence of any dilation or hydronephrosis

3. DISCUSSION

The presentation of lupus nephritis with horseshoe kidney is extremely rare with ours being an exceedingly rare one in scientific literature so far. A case was documented in a 13-year old boy who was evaluated for hematuria and proteinuria [4]. SLE was diagnosed based on laboratory findings upon admission and a complete physical examination, which was solidified by abdominal ultrasonography and by computed tomography. Patients with horseshoe kidneys are at an increased risk of infection, obstruction, urolithiasis, and malignant transformation. Hydronephrosis occurs due to obstruction at the level of the ureteropelvic junction that could be because of a higher level of the ureter at the renal pelvis [5]. There is an association of this anomaly with other chromosomal and genetic abnormalities such as Trisomy 18, Trisomy 9, and Turner syndrome. As many as 33% of patients with horseshoe kidneys have anomalies other genito-urinary, gastrointestinal, respiratory, and skeletal system defects. There is also an association with malignancies such as Wilm's tumor, transitional cell carcinoma, and/or carcinoid tumor [6-9].

Horseshoe kidney occurs due to a developmental defect during the 7^{th} and 9^{th} weeks of gestation [10]. The metanephric blastemal arising from the intermediate mesoderm remains unseparated. The ureteric buds come in contact with the fused nephrogenic cords with cranial growth. As the isthmus makes contact with the inferior mesenteric artery it stops its ascent. The fusion thus prevents normal posterior rotation of the kidney that causes the anterior orientation of the renal pelvis. The fusion typically happens at the lower ends while the upper and middle are rarely involved [11]. A majority of cases of horseshoe kidney are asymptomatic with a male predominance. However, Wilson and Azmy reported 15 children among a population of 20 who presented symptomatically [12]. In another study of 30 cases among them, a majority were male; 22 cases presented with abdominal pain, 12 with hematuria, and 2 with pyuria.

It must be acknowledged that such cases may be underrepresented in developing countries like Pakistan [13-14]. As the fifth most populous country in the world, it is essential that Pakistan continues to represent the multitude of clinical findings in scientific literature [15].

4. CONCLUSION

We present a rare case of horseshoe kidney and lupus nephritis in an adult male patient. The diagnosis is often established upon uncovering classical radiological findings on ultrasound, CT, and MRI scans in addition to examination findings and classical symptomatology. Our case emphasizes the need to carefully evaluate

presentations of acute SLE in emergency units. We also highlight that patients may have longstanding symptoms that are oftentimes missed on routine testing at secondary or tertiary care hospitals in low and middle-income countries like Pakistan. It is essential to manage patients upon correct radiological diagnosis and conduct regular medical, extracorporeal therapy, and regular symptomatology follow-ups.

CONSENT AND ETHICAL APPROVAL

As per international standard or university standard guideline Patient's consent and ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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