

Functional Medicine

Supernumerary Kidney Associated with Horseshoe Malformation: A Case Report and Review of Literature



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ABSTRACT

We report a case of supernumerary kidney associated with horseshoe malformation. A 35-year-old man presented complaining of vague and intermittent left flank pain from few months ago. Ultrasonography of urinary tract showed bilateral hydronephrosis and was suggestive of the horseshoe anomaly. Further evaluation with Intravenous urography showed three renal moieties consisting of a horseshoe kidney and a malrotated right kidney cephalad to and fused with the right moiety of horseshoe kidney.

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Introduction

Supernumerary kidney is an accessory reniform kidney with its own distinct vasculature, collecting system and encapsulated parenchyma. The true incidence is unknown due to the rarity of the abnormality. Supernumerary kidney with a horseshoe component is an extremely rare abnormality. We report a case of supernumerary kidney consisting of 3 renal moieties, including a horseshoe component.

Case presentation

A 35-year-old man presented with vague and intermittent left flank pain from few months ago. His physical examination did not reveal any significant finding. Ultrasonography revealed bilateral hydronephrosis and was suggestive of horseshoe malformation. To better delineate the anatomy of urinary system, intravenous urography was performed and revealed 3 renal moieties; a horseshoe kidney at L2–L3 level and a malrotated reniform kidney cephalad to and fused with the right moiety of the horseshoe kidney. The horseshoe component showed delayed excretion and moderate pyelocaliceal dilation on left side as well as mild hydronephrosis on the right moiety. The pelvis of the supernumerary right kidney was

oriented anteriorly and the ureter was deviated laterally in its course by the horseshoe kidney. Findings of excretory pyelography raised the suspicion for ureteropelvic junction obstruction and the patient underwent 99mTc-DTPA diuretic renography. The renogram did not reveal any pathological process including obstruction; therefore, we decided to observe the patient and follow him with periodic evaluations.

Discussion

Supernumerary Kidney is a rare congenital anomaly of the urinary system with about 100 cases reported in the literature. It may be either totally isolated from the normal kidney or connected through a loose areolar tissue and usually smaller in size with decreased function.¹

Supernumerary kidney may be associated with other kidney anomalies including horseshoe malformation. Although horseshoe kidney is relatively common, affecting 1 in 500 adults,¹ coexistence of both anomalies is extremely rare and to the best of our knowledge, less than 5 reports of supernumerary kidney along with horseshoe anomaly exist in the literature.^{2,3}

Embryologically, supernumerary kidney is believed to result from aberrant division of the nephrogenic cord into two metanephric blastemas at 5th to 7th week gestation that eventually form two kidneys with partial or duplicated ureteral buds. The presence of an additional ureteric bud or a branching one from the initial bud is also a necessary step in embryogenesis of supernumerary kidney.¹

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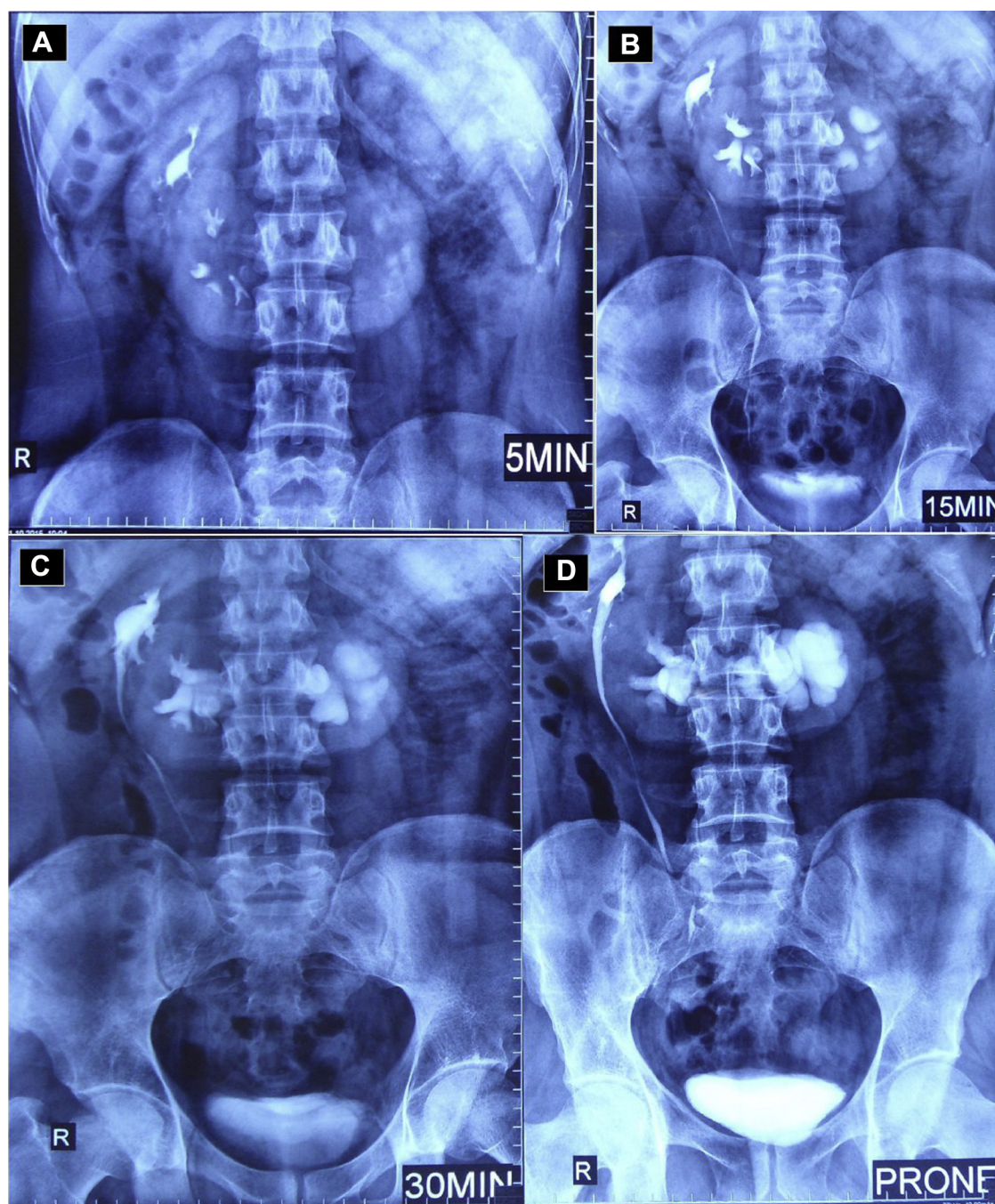


Figure 1. (A–D): Intravenous urography shows a horseshoe kidney and a malrotated supernumerary kidney cephalad to and fused with the right renal moiety. Horseshoe kidney shows delayed excretion and moderate hydronephrosis of left moiety as well as mild pyelocaliceal dilation on the right side.

Supernumerary kidney usually lies caudal to the ipsilateral kidney and sometimes in the iliac fossa or anterior to the sacral promontory¹; however, in our case it was located cranially.

The ureteral interrelationship in patients with supernumerary kidney may be variable. The ureter of supernumerary kidney may traverse independently or more frequently as a bifid ureter.^{1–4} Cranially located supernumerary kidney usually drains through a separate ureter and the Weigert–Meyer principle is typically followed. The ureter may also open into the vagina resulting in urinary incontinence.⁵

Supernumerary kidney usually has a separate arterial supply; however, sophisticated anatomical variations may exist. Therefore,

a thorough evaluation of vascular anatomy is of utmost importance in patients who require surgery to prevent inadvertent damage to the supernumerary renal moiety.

Associated congenital anomalies include urethral atresia, vaginal atresia, complete duplication of urethra and penis, ectopic ureteral opening, horseshoe kidney, imperforate anus, ventricular septal defects, meningomyeloceles, and coarctation of aorta.^{2–5} Due to high prevalence of associated anomalies thorough physical exam and precise work-up is important in such patients (Fig. 1).

This anomaly is usually asymptomatic; however, fever, hypertension, abdominal discomfort or palpable mass may be presenting symptoms. Several pathologic conditions including

hydronephrosis, pyelonephritis, pyonephrosis, renal and ureteral calculi, as well as malignant and benign neoplasms might affect renal moieties in patients with supernumerary kidney. The aforesaid pathologies may occur more frequently when supernumerary kidney is associated with other renal anomalies, i.e. horseshoe kidney. Horseshoe anomaly is considered the most common renal fusion anomaly and a significant proportion of patients with horseshoe kidney suffer from additional urologic and non-urologic defects¹ leading to more symptomatic presentation. Early fusion of the developing kidneys prevents ascent and rotation of the kidneys and results in impaired drainage and urinary stasis. We also noted urinary stasis in our patients without clear evidence of obstruction. Urinary stasis predisposes these patients to stone formation and urinary tract infections and warrants close follow up.

Various imaging modalities, i.e. ultrasonography, intravenous urography and computed tomography scanning are of value for diagnosis. However, evaluation with nuclear scintigraphy and/or magnetic resonance imaging/urography may also be necessary in patients with more complicated anomalies.

Management of these conditions depends on the function, symptoms and associate complications. Asymptomatic or

minimally symptomatic cases are considered for regular follow up for early detection of potential pathologies and complications. However surgical intervention might be necessary in patients with certain pathologies associated with supernumerary kidney.

Conflict of interest

There is no conflict of interest.

References

1. Ramanathan S, Kumar D, Khanna M, et al. Multi-modality imaging review of congenital abnormalities of kidney and upper urinary tract. *World J Radiol.* 2016;8.
2. Mustafa M. Bilateral supernumerary kidneys in conjunction with horseshoe anomaly. *Saudi J Kidney Dis Transpl.* 2012;23:1243.
3. Fathollahi A. Supernumerary kidney with a horseshoe component. *Urol Case Rep.* 2014;2:162.
4. Macpherson RI. Supernumerary kidney: typical and atypical features. *Can Assoc Radiol J.* 1987;38:116.
5. Gonzalvo Perez V, Ramada Benlloch F, Blasco Alfonso JE, et al. Supernumerary kidney with ectopic ureteral opening to the vagina associated with horseshoe kidney. *Actas Urol Esp.* 1992;16:796.