

Case Report

Supernumerary Kidney in Conjunction with Horseshoe anomaly-Case report and literature review

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Abstract: A supernumerary kidney is a rare congenital anomaly of the urinary tract. Association of supernumerary kidney with horseshoe kidney is even rarer and true incidence of this anomaly is not known because of its infrequent occurrence. Because of the wide range of associated anomalies and the rarity of such cases, diagnosis and management remains difficult. Ultrasound, Intra venous urography/ Computed tomography urography are usually enough to make the correct diagnosis in majority of the cases. However, if surgical intervention is planned, further radiological imaging studies are recommended.

Keywords: Supernumerary kidney, Horseshoe kidney, Ultrasound, Intra venous urography

INTRODUCTION:

Supernumerary kidney is a rare congenital anomaly, with about 100 cases reported in the literature [1-3]. Very rarely documented is the supernumerary kidney in conjunction with horseshoe kidney, with unknown incidence. Embryologically these kidneys are formed by aberrant division of nephrogenic cord into two metanephric blastemas with bifurcation of one bud [3-5]. We hereby present a rare case of supernumerary kidney in conjunction with horseshoe kidney.

CASE HISTORY:

A 1yr old girl child with coloboma left eye was referred for abdominal ultrasound examination to rule out other congenital anomalies. Her ultrasound examination showed empty right renal fossa. There was a horseshoe kidney. Both renal moieties were caudally located, malrotated with reversed longitudinal axis and anterior renal pelvis, left moiety of horseshoe kidney was smaller than right. Both moieties had separate renal pelvis and ureters. There was no hydronephrosis or calculus and renal echotexture was normal. In addition there was a supernumerary kidney in left renal fossa, small in size with normal echotexture and compact pelvi-calyceal system [Figure 1]. Color Doppler examination revealed two distinct main renal arteries supplying the horseshoe kidney. The right renal moiety

received its arterial supply from a branch of the abdominal aorta just before its bifurcation and left moiety received its arterial supply from a branch left common iliac artery [Figure 2]. The supernumerary kidney in left renal fossa received its arterial supply from aorta distal to origin of superior mesenteric artery. On basis of grey scale and color Doppler ultrasound findings diagnosis of supernumerary kidney in association with horseshoe kidney was made. Intra venous urography (IVU) was advised to confirm the sonography findings. IVU revealed normal excretion and concentration of contrast by horseshoe as well as by supernumerary kidney. On both sides single ureter was visualized with normal opening at vesico-ureteric junction, there was no ectopic ureteral opening bilaterally [Figure 3]. A triple-phasic MDCT scan with intravenous contrast administration revealed normal excretory phase by the kidneys bilaterally. There was bifid ureter on left side. The ureter from the supernumerary kidney joined the ureter from left moiety of horseshoe kidney to form a single ureter opening normally at left vesico-ureteric junction [Figure 4]. The separate arterial supply and venous drainage of horseshoe kidney and supernumerary kidney was confirmed as depicted by color Doppler examination [Figure 5].

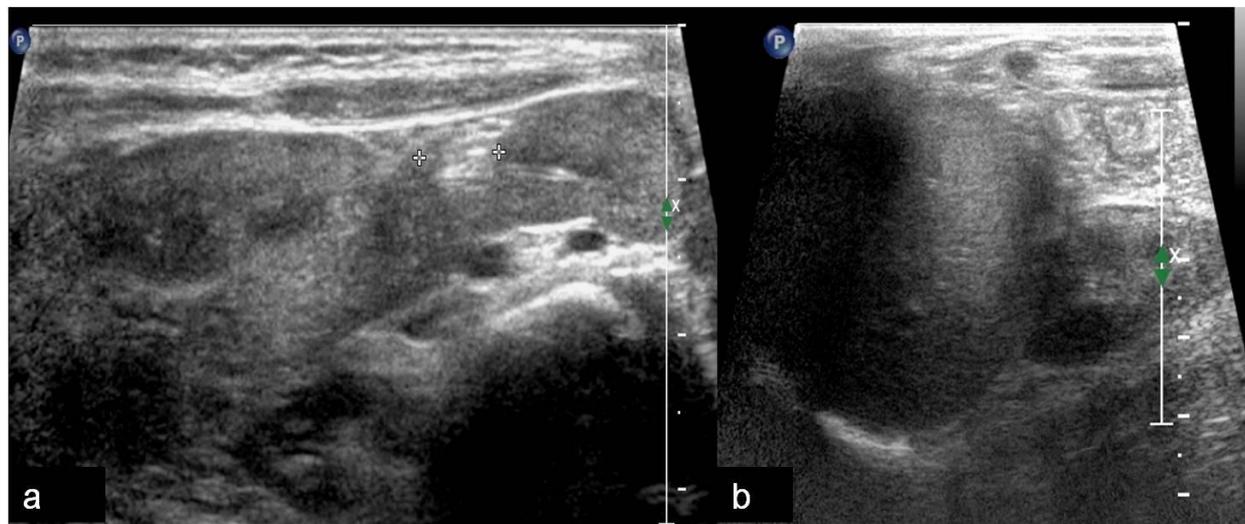


Fig 1(a-b):-Grey scale ultrasonography images showing (a) horseshoe kidney in lower abdomen and (b) small, supernumerary kidney in left renal fossa.

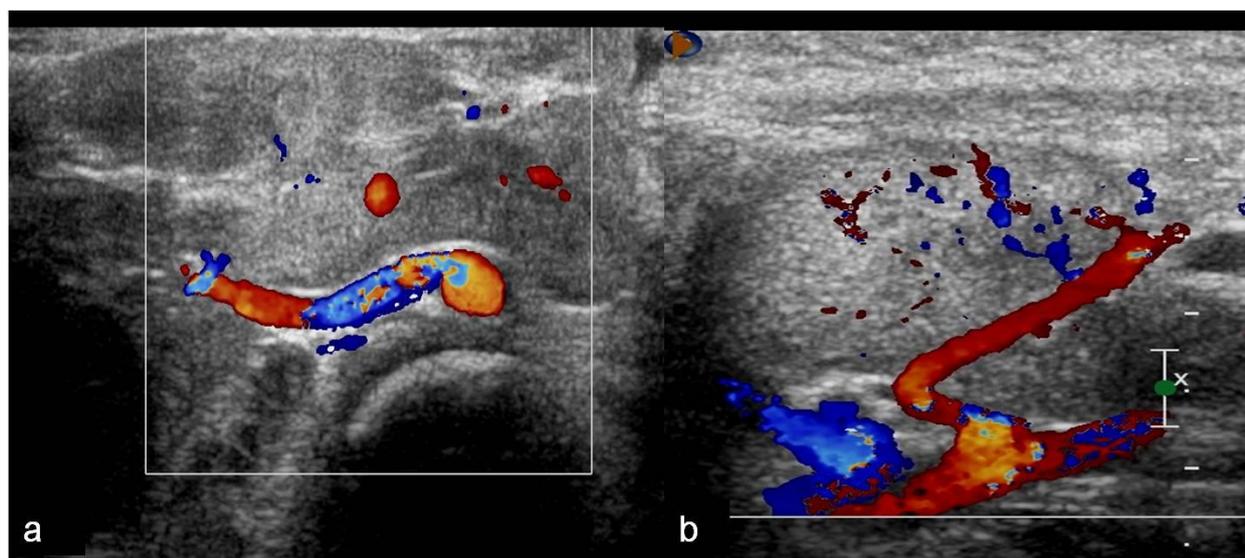


Fig 2(a-b):- Color Doppler images showing (a) right renal moiety of horseshoe kidney receiving arterial supply from branch of distal aorta and (b) left renal moiety receiving arterial supply from branch of left common iliac artery



Fig 3(a-b):- Intravenous urography showing horse shoe kidney with characteristic flower vase appearance of pelvi-calyceal system with ureters opening at vesico-ureteric junction. Note excretion of contrast from supernumerary left kidney (arrow)

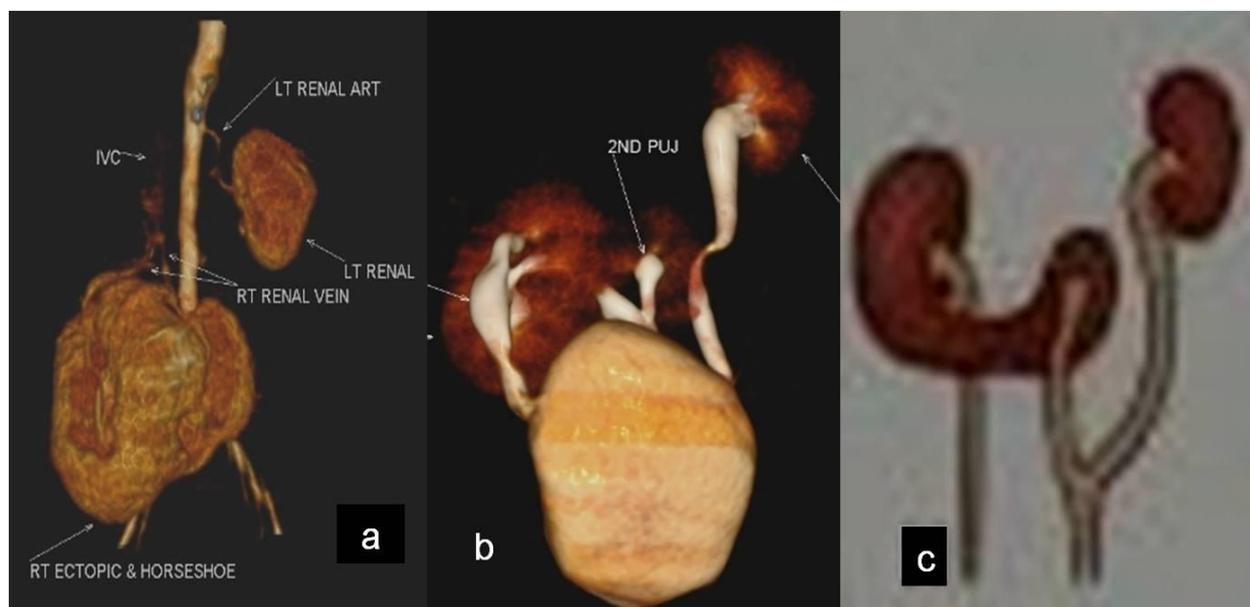


Fig 4(a-c):- (a,b) Volume rendering technique(VRT) MDCT images showing supernumerary kidney in association with horse shoe kidney with separate pelvi-calyceal system.(c) schematic representation of the anomaly



Fig 5(a-c):- Coronal multiplanar reformatted MDCT images show separate arterial supply to both moieties of horse shoe kidney and supernumerary kidney in orthotopic position

DISCUSSION:

A supernumerary kidney is a third kidney (in addition to the two independent kidneys). This is a rare congenital anomaly of the urinary tract. Association of supernumerary kidney with horse shoe kidney is even rarer and true incidence of this anomaly is not known because of its infrequent occurrence [2]. The supernumerary kidney needs to be differentiated from the more commonly occurring duplex kidney, which is defined as having two pelvicalyceal systems that are associated with a single ureter or with double ureters. The supernumerary kidney, in contrast, is thought to be an accessory organ with a separate arterial supply, venous drainage, collecting system, and distinct encapsulated tissue [4, 5]. Two types of supernumerary kidneys exist [6] (i) drained by a bifid ureter and (ii) drained by a separate ureter. When a bifid system is present, supernumerary kidney lies caudally, and when a separate ureter is seen then the supernumerary kidney is located cranially in relation to normal kidney. In such a case, the ureters enter the bladder ectopically according to the Weigert-R meyer rule, the ureter may insert medially and inferiorly into the bladder. Sometimes the ureter of the supernumerary kidney may be associated with an ectopic opening, such as into the vagina [4]. If both horseshoe kidney and a supernumerary kidney coexist then the half of the horse shoe kidney on the side of the body containing the supernumerary kidney is always small [7]. In our case also left moiety of horseshoe kidney was smaller however the location of the supernumerary kidney was orthotopic which is very rare. There was a bifid left ureter draining supernumerary kidney and left moiety of

horseshoe kidney and draining through single opening at left vesico-ureteric junction.

The embryological basis for this anomaly is thought to be the abnormal division of the nephrogenic cord into two metanephric blastemas that then form two kidneys, in association with either a partially or completely duplicated ureteral bud [3-5]. The true incidence is unknown due to the rarity of the anomaly and it is believed to be found in both sexes with equal frequency [1, 2]. Though horseshoe kidneys are relatively common renal fusion anomaly with an incidence of about one in 400-800 live births. Coexisting supernumerary and horseshoe kidney are however very rarely documented [8].

Although the present case had no urinary symptoms, symptoms have been noted in about two-thirds of the cases of supernumerary kidney [4]. The most commonly associated pathologies include hydronephrosis, pyelonephritis, pyonephrosis, renal and ureteral calculi, carcinoma, papillary cystadenoma, and Wilms tumors [9-11]. A few anomalies have been described in literature found in associated with supernumerary kidneys such as ureteral atresia, vaginal atresia, complete duplication of urethra and penis with ectopic ureteral opening into the vagina or introitus, imperforate anus, ventricular septal defects, meningo myeloceles, and coarctation of the aorta [2,11-13].

Because of the wide range of combined congenital anomalies and the relative rarity of such cases, it is difficult to standardize a protocol for diagnosis and, thus, over diagnosis with many

unnecessary imaging tests is done. Therefore, diagnosis of patients with supernumerary kidneys represents a challenge. In the literature, majority of these cases were diagnosed using various imaging techniques, including ultrasound, CT scan, IVU, magnetic resonance imaging, CT angiography and dimercapto succinic acid (DMSA) and diethylene triamine pentacetate (DTPA) scans. While some authors have reported that IVU, CT and ultrasound are adequate for the diagnosis of supernumerary kidneys, [14] other studies have included MRI, DTPA, DMSA and CT angiography in addition [15, 16]. We believe that ultrasound, IVU/CT urography seem to be enough for the diagnosis in majority of the cases. CT urography has advantage over IVU in depicting anatomy of pelvi-calyceal system and ureter better as well as demonstrating the arterial supply and venous drainage in this rare anomaly.

Management of this condition depends on symptoms and the function of the supernumerary kidney. If the patient is asymptomatic, as in our case, no treatment is required, but regular follow-up maybe advised. If the kidney is diseased or nonfunctional, nephrectomy is usually the preferred procedure [10, 12].

CONCLUSION:

Supernumerary kidneys are very rare congenital anomalies of urinary tract. Very rarely documented is the supernumerary kidney in conjunction with horseshoe kidney, with unknown incidence. Because of the wide range of associated anomalies and the rarity of such cases, diagnosis and management remains difficult. Ultrasound, IVU/ CT urography are usually enough to make the correct diagnosis in majority of the cases. However, if surgical intervention is planned, further radiological imaging studies are recommended.

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