

Supernumerary Kidney (Triple Kidney) With Horseshoe Component: A Case Report.

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Citation

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Abstract

Supernumerary kidney is a rare congenital anomaly of the urinary system with less than 100 cases documented in literatures worldwide¹⁻³. Very rarely documented is the fused supernumerary kidney and horseshoe component with unknown incidence, due to the rarity of this complex anomaly. We hereby present a rare case of complex renal anomalies of fused supernumerary kidneys with horseshoe component misdiagnosed as an abdominal tumour on ultrasound, incidentally discovered at Computed Tomography.

Introduction

Supernumerary kidneys are rare congenital anomaly of the urinary system with less than 100 cases documented in literatures worldwide¹⁻³. 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}. Embryologically, supernumerary kidney is believed to result from abnormal division of the nephrogenic cord into two metanephric blastemas that eventually form two kidneys with partial or duplicated ureteral bud^{1,3-5}, this occurs during the period of urogenital system formation about the 5th to 7th week of gestation⁶. No racial predominance is documented in the literature. And most cases of supernumerary kidneys are discovered incidentally⁷. Diagnosis and treatment are commonly challenging due to the rarity of this anomaly, its varied presentation and sparse documentation in literatures^{7,8}. Though horseshoe kidneys are relatively common renal fusion anomaly with an incidence of about one in 400-800 live births⁹. Coexisting supernumerary and horseshoe kidney are however very rarely documented¹⁰.

CASE REPORT

A 25 year old female of Asian origin, who was being managed at a private medical centre for diffuse periumbilical pain of two days duration. No history of vomiting, change in bowel habit or history of melena stool. She had been treated several times at the referral hospital for recurrent urinary tract infection. General examination revealed a mildly tender palpable midline suprapubic mass. Other systemic examinations were essentially normal. Abdominal Ultrasound scan at a private facility reported a well defined, solid, solitary suprapubic intra-abdominal mass measuring 3cm x 4cm x 6cm in its AP x TS x LS, overlying the inferior vena cava (IVC) and abdominal Aorta. The right and left kidneys and other intra-abdominal organs were reported to be within normal limits. The urinary bladder was also normal in outline and content. An impression of an enlarged lymph node probably due to lymphoma or Inflammatory mass was made. Based on the above findings, the patient was referred to our centre for Abdominal CT examination.

An abdominal CT scan using a tri-phasic abdominal protocol

with oral as well as intravenous contrast was performed with 64 slice Toshiba Aquilion CT scanner. The axial, Multiplanar Reconstruction (MPR) and 3-Dimensional images revealed that the right and left kidneys were located in their normal positions. They measure 10.3cm x 4.95 cm and 10.65 cm x 3.9cm in their LS x AP on the right and left respectively.

However, the lower cuts showed a third, midline and obliquely positioned supernumerary kidney anterior to the lower poles of the right and left kidneys, the IVC and abdominal aorta but there was no compression or narrowing of the abdominal aorta. It measures 4.0cm x 7.6cm in its AP x TS dimensions. There is fusion of the Supernumerary kidney with both kidneys anteriorly at their lower poles in keeping with horseshoe kidney configuration.

Figure 1

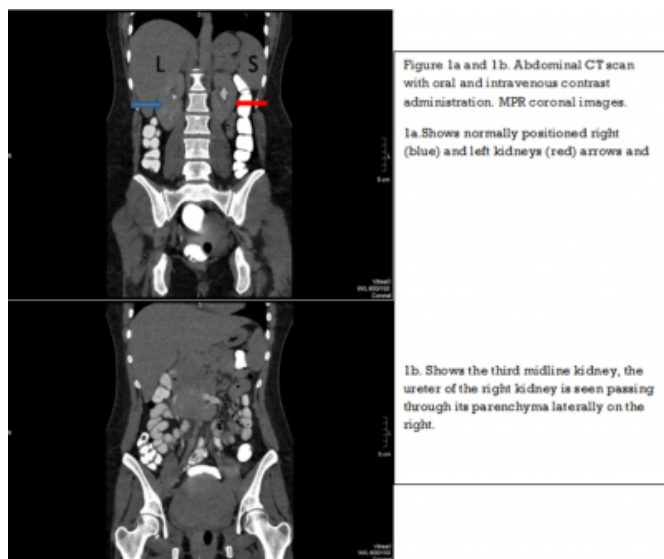
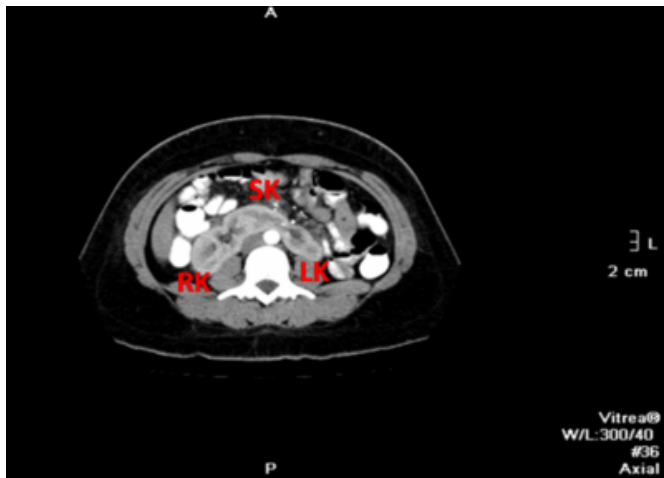


Figure 2

Figure 2. Abdominal CT scan with oral and intravenous contrast administration. Axial images of the arterial phase show fusion of the supernumerary kidney (SK) with the right (RK) and the left kidney (LK), anteriorly at their lower poles.



The arterial supply of the normally located right and left kidneys is via single normal sized arterial supply from the abdominal aorta on either side following the normal anatomical origin. The caudally placed obliquely oriented midline supernumerary kidney, however receives an average size single artery directly from the abdominal aorta, distal to the origin of the inferior mesenteric artery. This artery almost immediately bifurcate into two branches, each supplying the right and left side of the supernumerary kidney,

Figure 3

Figure 3: Volume rendering of the arterial Phase CT Abdomen. Image show the right (RRA) and left (LRA) renal arteries originating from the Abdominal aorta. Supernumerary kidney (SK) fusion with the .Right kidney (Blue arrow) and Left kidney (Green arrow) anteriorly at their lower poles.

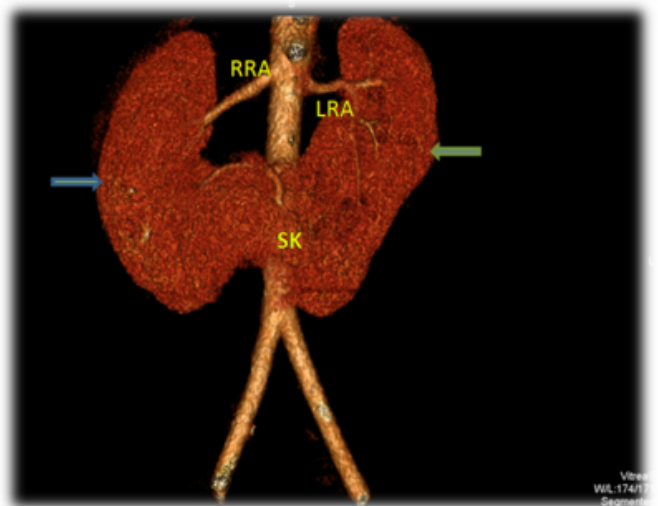
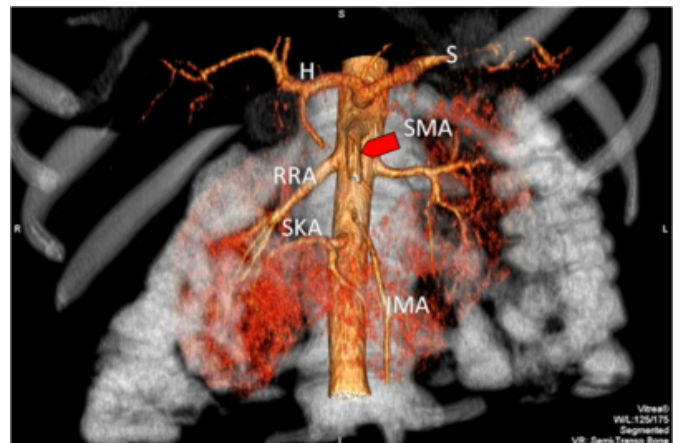


Figure 4

Figure 4 CT. 3-D volume rendering of the arterial phase (oblique view) showing the vascular anatomy. Splenic artery(S), Hepatic artery (H), Superior(SMA, red arrow) and inferior mesenteric artery(IMA) as well as the arterial supply of the right(RRA), left and the supernumerary kidney(SKA).

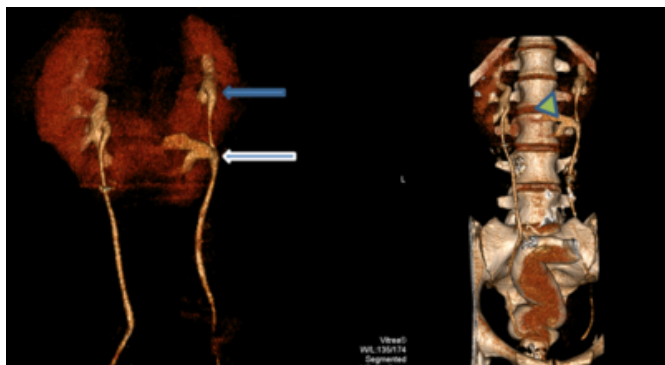


A normal and prompt excretion was demonstrated by the three kidneys. There is malrotation of the right and left kidneys as shown by the left pelvicalyceal system being rotated anteriorly while that of that of the right kidney is laterally rotated. The supernumerary kidney showed a superolaterally directed pelvis to the left. The right ureter arises laterally and it is minimally compressed as it passes through the parenchyma of the supernumerary kidney but its

proximal and distal parts were normal. The ureter from the left kidney show normal caliber proximally and reduction in caliber as it descends caudally to fuse with the pelviureteric junction of the supernumerary kidney to form a single ureter distally. The common ureter continues as a single ureter down to its insertion into the urinary bladder. No pelvicalyceal, ureteral dilatation or any other urinary system pathology is seen. The common left ureter and the right ureter showed normal insertion into the urinary bladder. The urinary bladder and ureterovesical junctions were normal, and there was no ectopic ureteral opening bilaterally. No calculi, cyst or mass lesion was seen in the kidneys, no other intra-abdominal mass or pathology was also found.

Figure 5

Figure 5 A & B: CT scan 3-D volume rendering of the excretory phase (oblique view) of the pelvicalyceal systems and ureters shows anterolaterally rotated right pelvicalyceal system and ureter. On the left, the pelvis from the normally placed kidney passes antero while its ureter joins the pelvis of the caudally placed supernumerary kidney at its pelviureteric junction (white arrow) to form a single ureter. Both ureters shown down to their distal part (Fig B).



DISCUSSION

Supernumerary kidney is a rare congenital anomaly of the urogenital system¹⁻³. A Supernumerary kidney is an accessory/extra or third kidney present along with two normally located kidneys. They are normal in shape and function and have distinct normal encapsulated tissue and separate arterial supply and venous drainage. Supernumerary kidneys may be a separate entity from the other kidneys or connected by loose areolar tissue to the normal kidney. They are usually smaller in size but vary in size, compared to the other kidneys and in most cases its location is caudal to the ipsilateral kidney and more common on the left^{3,5,7,11}. Other documented location is in the iliac or anterior to the sacral promontory^{3,12}. Supernumerary kidneys either have a bifid ureter in most cases, or a separate one. Bifid ureters are commonly associated with caudal position while cranially

located supernumerary kidneys have separate ureter that inserts ectopically, medial and inferior to the normal kidney's ureter, according to Weigert-Meyer rule, into the urinary bladder^{1,4}. The ureter may also open into the vagina and patient presents with urinary incontinence^{1,4}.

In this case, the supernumerary kidney although caudally located in the upper pelvis, is in the midline, and connected to the inferior poles of both kidneys anteriorly giving the horseshoe configuration (Fig-2).

The left ureter inserts into the pelviureteric junction of the Supernumerary kidney. Literature review shows that supernumerary kidneys usually have a separate arterial supply. This is seen in this case report with the supernumerary kidney been supplied by a short segment single artery originating from the abdominal aorta, distal to the inferior mesenteric artery. And immediately dividing into two arteries, each supplying the right and the left components of the midline kidney (Fig. 4).

A common differential diagnosis is a duplex kidney which is associated with two pelvicalyceal systems with a single or double ureter^{4,5}, duplex kidneys however does not have a separate arterial supply and distinct encapsulated tissue^{3,4,5}. Associated congenital anomalies includes; ureteral atresia, vaginal atresia, complete duplication of urethra and penis, ectopic ureteral opening, horseshoe kidney, imperforate anus, ventricular sepal defects, meningomyeloceles, and coarctation of aorta^{1,2,13-16}. Apart from the coexisting horseshoe anomaly, none of the other associated anomalies were present in this case.

Clinical symptoms such as fever, abdominal pain, and abdominal mass^{1,7,12} are common in most cases and associated complications include hydronephrosis, pyelonephritis, pyonephrosis, renal and ureteral calculi, carcinoma, papillary cystadenoma, and Wilm's tumors^{1,3,13,15,17}. This case report was only associated with history of recurrent abdominal pain and symptoms of recurrent urinary tract infection.

Diagnostic modalities employed in the evaluation of Supernumerary kidney are intravenous pyelography, ultrasonography, nuclear scintigraphy, CT, and MRI^{1,3}.

Management option is dependent on the presence or absence of function in the supernumerary kidney, symptoms and complications^{1,3}. While surgical interventions are reserved for symptomatic cases with complications or non-functional

supernumerary kidney^{1, 3, 15, 16}. Asymptomatic cases are followed up regularly^{3, 15} for early detection of any pathology or complication within the kidney.

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