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Case Series

Radiology Section

Fused Supernumerary Kidneys with S-shaped Configuration: A Case Series

SAI SHANKAR MANKUZHY GOPALAKRISHNAN[1], JENIKAR PAULRAJ[2], R REMYA[3], NILLOHIT SAHA[4], HARSHAVARDHAN BALAGANESAN[5]

ABSTRACT

Supernumerary kidneys are considered as rare congenital renal anomalies (0.04%). Hereby, the authors present a case series comprising of three patients with incidentally discovered supernumerary kidneys on cross-sectional imaging. The first case of 65-year-old male presented with an anterior abdominal mass which turned out to be plexiform neurofibroma and incidentally diagnosed with right supernumerary kidney and type VIII variant of celiac artery. The second 21-year-old male patient came with suspected appendicitis and a supernumerary kidney was found on the left-side. The third case of 45-year-old female presented with abdominal pain and an incidental supernumerary kidney was diagnosed on cross-sectional imaging. Patients were asymptomatic without urinary complaints, were kept under observation, and did not require any treatment regarding supernumerary kidneys. These cases highlight the potential for incidental detection of supernumerary kidneys and the importance of distinguishing them from other renal anomalies/lesions. Regular follow-up is recommended for monitoring potential complications.

Keywords: Computed tomography, Diagnostic imaging, Renal variants

INTRODUCTION

Renal anomalies are fairly common, but the occurrence of a supernumerary kidney is a rare, congenital anomaly with less than 100 cases reported in current literature [1]. The first modern compilation of these cases was made by Kretschmer HL in 1929, during which 30 cases were presented [2]. The supernumerary kidney is typically found on the left-side and is smaller than a normal kidney. Supernumerary kidneys have their own blood supply, collecting system, and distinctly encapsulated tissue [3]. Most additional kidneys are smaller than regular kidneys [4], though they can sometimes be larger [4,5].

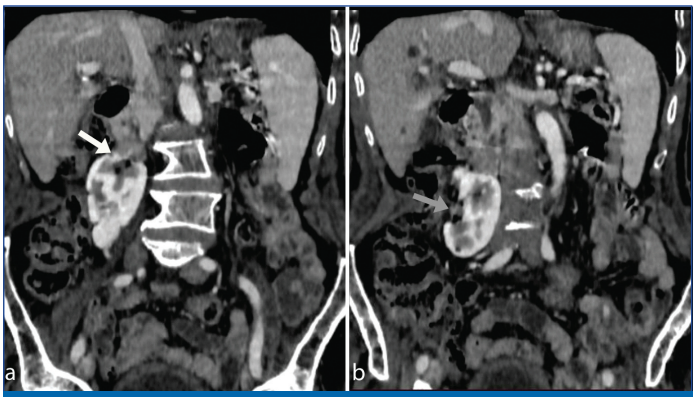
CASE SERIES

Case 1

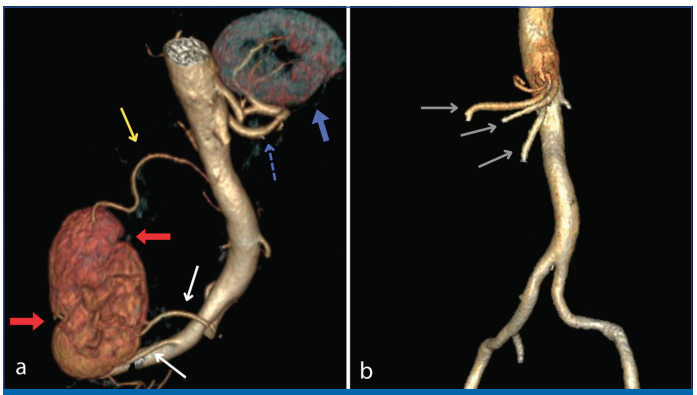
A 65-year-old male, with a known case of type I neurofibromatosis, previously operated on for plexiform neurofibroma involving the anterior abdominal wall along the epigastric region, came to Surgery Outpatient Department (OPD) for recurrence of ulceroproliferative tumour/lesion at the same surgical site and was referred to Department of Radiology to undergo contrast abdomen in suspicion of malignant transformation of plexiform neurofibroma and to evaluate for the deeper extension of lesion.

In present patient, incidentally, two renal kidneys were seen occupying the right renal fossa. Located just below the renal fossa, the hilum of the upper kidney is rotated medially while the hilum of the lower kidney is oriented laterally. Both hila merge, forming an S-shaped configuration [Table/Fig-1a,b]. The kidneys receive blood supply from three arteries: one branch from the abdominal aorta feeding the upper kidney, and two branches from the right common iliac artery supplying the lower kidney. However, the patient did not have any urinary complaint. The contralateral kidney appeared to be normal.

In present patient, interestingly celiac trunk was not visualised. Instead, an arterial variant in which the Common Hepatic Artery (CHA), Superior Mesenteric Artery (SA), and Left Gastric Artery (LGA) were seen as direct branches arising from abdominal aorta. According to Uflacker's classification of celiac trunk anomalies [6], this variant is type VIII where no celiac trunk was detected and instead its branches arise directly from the abdominal aorta [Table/Fig-2a,b].



[Table/Fig-1]: a) Contrast abdomen oblique coronal section in delayed excretory phase showed hilum of cranial kidney directed anteromedially on the right-side (white arrow); b) Hilum of caudal kidney located in anterolateral direction on the right-side (grey arrow).



[Table/Fig-2]: a) A 3D volume rendered image obtained from Contrast-enhanced Computed Tomography (CECT) abdomen study in arterial phase (oblique view) demonstrated S-shaped right kidney with hilum of the kidney directed medially and that of caudal kidney directed laterally (marked by bold red arrows). A branch from the abdominal aorta (yellow arrow) was seen supplying the cranial kidney and two branches from right common iliac artery supplying the caudal kidney (white arrows) on the right-side. The left kidney (bold blue arrow) was seen to be having a single arterial supply (blue dotted arrow) by a branch of abdominal aorta; b) A 3D volume rendered image in the arterial phase showed three different arteries (CHA, SA and LGA) directly arising anteriorly from the abdominal aorta (grey arrows).

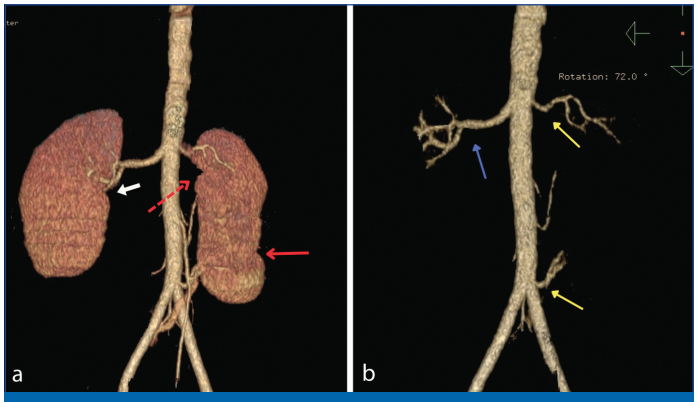
Axial sections of Contrast-enhanced Computed Tomography (CECT) of the abdomen in arterial phase showed a large heterodense enhancing mass measuring approximately 10.2×4.1×3.7 cm (length×width×anteroposterior) showing significant heterogenous enhancement with precontrast hounsefield units ranging from 25 to 30 and post-contrast hounsefield units ranging from 60 to 75. The mass was seen along the epigastric region predominantly involving subcutaneous plane with extension to skin causing skin thickening. Histopathology showed malignant transformation of the plexiform neurofibroma.

The patient was kept under observation and no treatment was required regarding urinary system, as the patient was asymptomatic. After a month, the patient came for a follow-up when the Histopathological Examination (HPE) results showed the malignant transformation of plexiform neurofibroma to fibrosarcoma and the patient was referred to onco surgeon for further management but after which the patient did not turn up for follow-up imaging.

Case 2

A 21-year-old male patient presented with complaints of pain in the right iliac fossa to the Emergency Department with a history of nausea and vomiting for the last three days. The pain was dragging in nature, aggravated by movement. An emergency contrast-enhanced abdomen triple-phase Computed Tomography (CT) study was done to rule out appendicitis, however, the study showed a normal appendix and there were no signs of inflammatory changes except distal ileal loops showing mild inflammation in the form of mild wall thickening and perienteric fat stranding. Few enlarged surrounding mesenteric lymph nodes were seen. Contrast CT abdomen also incidentally discovered a left-sided supernumerary kidney in this patient.

Two kidneys on the left-side were seen to be fused with two hila directed in opposite directions. The hilum of the cranially placed kidney appeared to be directed anteromedially while that of the caudal kidney was directed anterolaterally, forming S-shaped configuration. The two kidneys on the left-side were seen to get two separate arterial supplies from the abdominal aorta with one branch arising from the abdominal aorta supplying the cranial kidney and another branch arising just before bifurcation from the aorta supplying the lower kidney [Table/Fig-3a,b]. The two kidneys on the left-side showed separate venous drainage and collecting system. However, the right kidney was seen in the right renal fossa with a normal vascular supply and a collecting system. Also, present patient did not have any complaints pertaining to the urinary system and was under observation for a few days before being discharged. The patient did not require any treatment pertaining to supernumerary kidney and did not come back for follow-up.

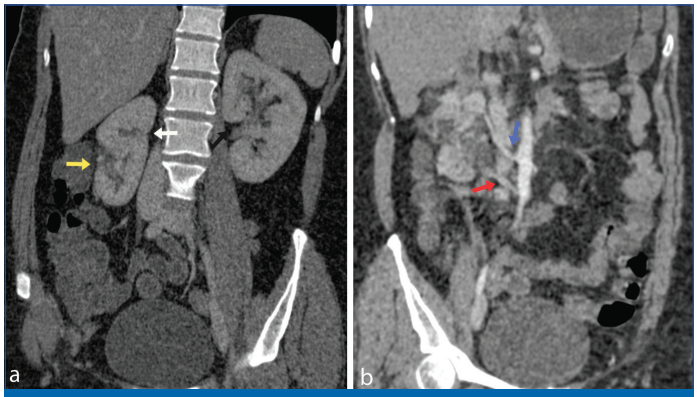


[Table/Fig-3]: a) A 3D volume rendered image of CECT abdomen in arterial phase showed two fused kidneys on the left-side with hilum of the cranial kidney directed medially (red dotted arrow) and hilum of caudal kidney directed laterally (red bold arrow) forming "S" shape. The hilum of right kidney was seen. (white bold arrow); b) A 3D volume rendering obtained from arterial phase of CECT abdomen (oblique view) showing arterial supply of both kidneys. One branch arising from abdominal aorta was seen supplying the upper kidney and one branch arising from the abdominal aorta just before bifurcation was supplying the lower kidney (yellow arrows) on the left-side. The right kidney was seen getting supply by a right renal artery arising from abdominal aorta (blue arrow).

Case 3

A 45-year-old female with no known medical history came to the Emergency Department with diffuse abdominal pain and vomiting for the last five days. The patient underwent contrast CT abdomen study, and it showed some mesenteric fat stranding along the mesenteric vessels with enlarged mesenteric lymph nodes.

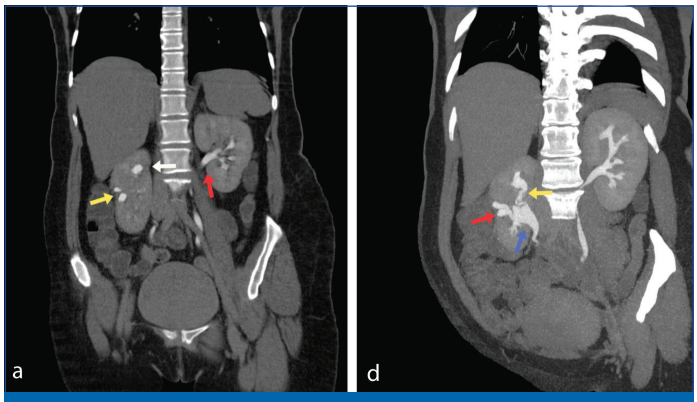
A right-sided supernumerary kidney was an incidental finding in this patient with the hilum of the cranially located kidney facing medially and the caudal kidney was seen fused with the cranial kidney in the right renal fossa and has an incomplete rotation configuration in which the hilum is facing anteriorly and laterally forming S-shaped [Table/Fig-4a,b].



[Table/Fig-4]: a) Contrast abdomen (oblique coronal section) showed hilum of cranial kidney oriented in anteromedial direction (white arrow) and hilum of caudal kidney in anterolateral direction (yellow arrow) with both kidneys fusing to form "S" shape; b) CECT abdomen arterial phase in oblique coronal view showed two separate arteries arising from right lateral aspect of abdominal aorta and the first branch supplying the cranially located kidney (blue arrow) and the other branch supplying the caudally placed kidney (red arrow) on the right-side.

Both the kidneys on the right-side had two separate small renal arterial supply originating from the abdominal aorta. The venous drainage was through separate venous channels which were seen fusing and forming a common right renal vein just before its drainage to the Inferior Vena Cava (IVC).

Both the kidneys on the right-side had separate collecting systems and were seen fusing at the hilar region adjacent to the lower kidney. The kidney on the opposite side was in a normal anatomical position with the left renal artery supplying the left kidney and getting drained by the left renal vein. The left kidney had a single collecting system draining into the bladder [Table/Fig-5a,b].



[Table/Fig-5]: a) In the same patient, contrast abdomen oblique coronal section of excretory delayed phase showed two collecting systems on the right-side; one arising from the medial aspect of upper kidney (white arrow) and other arising from lower kidney in lateral aspect (yellow arrow). The left kidney was seen in left renal fossa with a single hilum (red arrow); b) Oblique coronal Maximum Intensity Projection (MIP) image showed hilum of the upper kidney arising from medial aspect (yellow arrow) and hilum of lower kidney arising from lateral aspect of lower kidney (red arrow) and coursing postero-laterally to fuse with the collecting system of upper kidney (blue arrow) on the right-side and continuing as a single right ureter to drain into urinary bladder.

The patient was asymptomatic and kept under observation and did not undergo any intervention regarding this incidental finding of supernumerary kidney. The patient came back for follow-up to Surgery Department in study Institute and was not sent for further radiological investigations as the patient had no urinary complaint.

DISCUSSION

Supernumerary kidneys are rare and very few cases have been mentioned in literature till now. The supernumerary kidney is an accessory kidney with its own collective system, arterial supply and venous drainage and well-defined encapsulated renal tissue [3]. The prevalence of supernumerary kidneys cannot be estimated as it is one of the rarest abnormalities [6].

The embryological basis of this is the abnormal premature division of the nephrogenic cord into two metanephric blastemas that form two kidneys with partial or complete duplication of the ureteric bud [7].

A survey of collected cases indicates that most supernumerary kidneys are located below the normal kidney. However, in few instances, supernumerary kidneys have been found above the normal kidney [4]. In all present cases, the supernumerary kidney was found located caudal to the normal kidney. If, the extra kidney is situated above the normal kidney, its ureter typically remains entirely separate and might enter the bladder in an abnormal position. In such instances, the Weigert-Meyer rule may apply, with the ureter inserted into the bladder in a medial and inferior location [8]. It affects males and females equally and usually affects the left-side [9].

The supernumerary kidney is usually located on the left-side of the abdomen [8]. In present cases, however, in two patients the extra kidney was seen on the right-side and it was located on the left-side in the other patient.

The size and functionality of the supernumerary kidney also appears to be reduced when compared to normal kidneys [10]. In all present cases, the supernumerary kidney appeared to be smaller.

It is important to differentiate the supernumerary kidney from the more commonly occurring duplex kidney. A duplex kidney is characterised by having two pelvicalyceal systems associated with either a single ureter or double ureter and a single vascular supply within a single renal capsule [7]. A supernumerary kidney can be identified as an accessory organ with its separate vascular supply and collecting system [11].

Previously the supernumerary kidneys were usually found only during surgery or autopsy [12]. Several methods such as urography, retrograde pyelography were not useful [13,14]. The diagnosis of supernumerary kidneys has become more common with the use of ultrasound and Computed Tomography (CT) [15]. Scintigraphy techniques with Diethylene-triamine-pentaacetate (Tc-99m DTPA) or Technetium-99m -mercaptotriacetyltriglycine (Tc-99m MAG3) are more accurate in diagnosing supernumerary kidneys [16,17]. CT imaging had distinctive findings, clearly showing the combination of kidneys. CT angiography is valuable for noninvasively identifying the complex anatomy and origin of renal arteries. Magnetic Resonance (MR) angiography could serve as an alternative for detecting renal artery origins [18,19]. Supernumerary kidneys are often smaller and may have reduced function, evaluating excretory urograms less reliable. Conrad GR and Loes DJ reported that Dimercaptosuccinic acid (DMSA) scintigraphy provides superior imaging and functional assessment of supernumerary kidneys compared to excretory urograms [20].

Supernumerary kidneys have no intrinsic clinical significance [3]. However, they are often linked to infections, pyonephrosis, urolithiasis or hydronephrosis [18,19]. Supernumerary kidneys can be susceptible to traumatic injuries [20]. Some cases have been reported with Wilms tumour or adenocarcinoma [21,22]. Supernumerary kidneys have been linked to some abnormalities, such as ectopic ureteral opening into the vagina, ureteral atresia, vaginal atresia [23,24].

Supernumerary kidneys are treated individually and there is no standard protocol. Asymptomatic patients with incidental diagnosis of supernumerary kidneys as seen in present cases can be kept under observation and no treatment is required. However, regular follow-up can be advised. In cases of malignancy, severe or repeated infection, and multiple stones, open supernumerary nephrectomy or uretero-nephrectomy is a possible treatment option [25]. A laparoscopic approach has been mentioned as another option for surgical removal [1,26].

The findings of recent similar case reports of supernumerary kidney are tabulated in a [Table/Fig-6] with clinical and imaging findings, diagnosis, and management details [8,27-30].

Name of author	Age (year)	Gender	Symptoms	Imaging findings	Side	Treatment	Outcome
Tefera AT et al., [27]	35	Male	Right flank pain, tenderness	CT: Bilaterally fused, malrotated supernumerary kidneys each with its own vascular supply and obstructive stone	Bilateral	Open pyelolithotomy	Symptoms improved after surgery
Yener S and Lice Z [28]	8	Male	Urinary incontinence (day and night), right inguinal swelling	USG/CT: Right supernumerary kidney with rotational anomaly; DMSA: normal uptake; MRI: Malrotated supernumerary small kidney on the right-side	Right	Right orchiopexy, circumcision, anticholinergics	Complete resolution of incontinence
Krakhotkin DV et al., [29]	51	Male	Left flank pain	USG, CT: Two left kidneys, one with solitary cyst, bifid ureter	Left	Observation	Symptomatic relief
Kumar M et al., [30]	30	Female	Abdominal pain	USG, CT: Fused kidneys, separate pelvicalyceal systems, ureters join below supernumerary kidney	Right	Observation	Follow-up
Mebis W et al., [8]	50	Male	Abdominal pain	CT: S-shaped kidney with separate ureters and multiple renal arteries	Left	None specified	No clear explanation for abdominal pain found

[Table/Fig-6]: Recent cases of supernumerary kidney presented in literature [8,27-30].
USG: Ultrasonagraphy; DMSA: Dimercaptosuccinic acid; MRI: Magnetic resonance imaging

CONCLUSION(S)

Most of the cases of supernumerary kidneys are asymptomatic. In present cases, none of the patients had any urinary complaint and all of them were diagnosed incidentally on CT where an encapsulated retroperitoneal organ with its own blood supply and collecting system was detected. It is of utmost importance to diagnose supernumerary kidney correctly to avoid unnecessary surgical interventions. Regular follow-up is required after diagnosis since, there is risk of developing complications such as hydronephrosis, pyelonephritis, calculi or malignancy.

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PARTICULARS OF CONTRIBUTORS:
1. Associate Professor, Department of Radiodiagnosis, Shri Sathya Sai Medical College and Research Institute, SBV, Chengalpattu, Tamil Nadu, India.
2. Associate Professor, Department of Radiodiagnosis, Shri Sathya Sai Medical College and Research Institute, SBV, Chengalpattu, Tamil Nadu, India.
3. Assistant Professor, Department of Radiodiagnosis, Shri Sathya Sai Medical College and Research Institute, SBV, Chengalpattu, Tamil Nadu, India.
4. Junior Resident, Department of Radiodiagnosis, Shri Sathya Sai Medical College and Research Institute, SBV, Chengalpattu, Tamil Nadu, India.
5. Assistant Professor, Department of Radiodiagnosis, Shri Sathya Sai Medical College and Research Institute, SBV, Chengalpattu, Tamil Nadu, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Nillohit Saha,
Junior Resident, Department of Radiodiagnosis, Shri Sathya Sai Medical College and Research Institute, Sri Balaji Vidyapeeth, Chengalpattu-603108, Tamil Nadu, India.
E-mail: drnillohit@gmail.com

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