International Journal of Science and Research (IJSR)
ISSN: 2319-7064
Impact Factor 2024: 7.101

A Rare Case of Malrotated and Fused Supernumerary Kidney

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Abstract: ***Supernumerary kidney is a rare congenital anomaly which may be defined as an additional kidney with separate capsule, blood supply, and collecting system draining into a common or separate ureter. Sometimes small fused supernumerary kidney may be difficult to differentiate from a renal moiety with a separate collecting system including the ureter. There have been fewer than 100 cases reported in literature of fused supernumerary kidney(1). We present a case of S-shaped fused supernumerary kidney.***

Keywords: Malrotation, Supernumerary kidney, Fused, S-shaped fused supernumerary kidney

1. Case Report

A 67-year-old female, known case of type II diabetes mellitus was referred to department of radiodiagnosis of our tertiary care hospital for a routine ultrasound abdomen. She had no pain abdomen, flank pains, recurrent urinary tract infections or hematuria.

At presentation her routine blood parameters were as follows;

1) Blood sugar
   - Fasting – 121 mg/dl
   - Post prandial – 143 mg/dl
   - HbA1c – 7.1
2) TLC – 4300
   - Neutrophil – 61%
   - Lymphocytes – 36%
   - Monocytes – 2%
   - Eosinophils - 1%
   - Basophils - 0%
3) Hb – 11 mg/dl
4) Platelet – 210000/cmm
5) Sodium – 137 mg/dl
6) RFT
   - BUN – 17 mg/dl
   - Creatinine – 1.0 mg/dl
7) Urine RE/ME
   - Sugar (+)
   - Albumin (-)
   - Epithelial cells – 1-2
   - RBC – Nil
   - WBC – 1-2
   - Crystals and casts – nil.
   - 24 hours urinary protein – 87mg.
8) Potassium – 4.1 mg/dl

Ultrasound revealed a mass like lesion in the inferior pole of left kidney with echotexture similar to that of the left kidney. This lesion also had internal structures similar in morphology to the renal collecting system. The right kidney was small in size measuring approximately 7 cm and was normally located in the renal fossa.

A suspicion of supernumerary kidney was made. With consent from the patient, a corroborative NCCT KUB was done from the level of diaphragm to below the symphysis pubis in helical mode. CT revealed a small supernumerary kidney fused in the inferior pole of the left kidney. The upper kidney had a normal, anteromedially oriented pelvis but the lower kidney had an anterolaterally directed pelvis assuming an ‘S’ shape. Both these fused kidneys had separate vascular supplies and separate collecting systems.

The normally located right kidney measured 6.0 x 3.6 cm and among two left kidneys, the cranially located kidney measured 5.8 x 3.4 cm, and the caudally located kidney measured 8.3 x 3.4 cm. Because she was asymptomatic and her kidney functions were well within normal range, she has been placed on a periodic follow up and does not require active intervention as of now.

2. Discussion

Supernumerary kidney is generally asymptomatic and is often discovered incidentally during routine or unrelated imaging studies. Therefore, calculation of true incidence is difficult in cases of supernumerary kidney (2). Palpable lump, abdominal mass, flank pain or fever due to pyelonephritis and rarely with urinary symptoms like incontinence are the common presenting features. The common age at presentation is 40-50 years. Intravenous urography, retrograde pyelography, nuclear scintigraphy, ultrasonography, computed tomography, magnetic resonance imaging, and angiography are the various modalities that can be used to diagnose the condition (3,4).

Most cases of supernumerary kidneys have one additional kidney and are found caudal to the left kidney. Often the additional kidney is smaller than the native kidney(5). It may either be completely separate from the normal kidney or connected through a loose areolar tissue with decreased function (6). In case of cranially located supernumerary kidney in relation to the native kidney, the ureter is usually completely separate and may enter the bladder ectopically (7).

The embryological basis of supernumerary kidney is thought to be due to an abnormal division of the nephrogenic cord into two separate metanephric blastemas giving rise to two kidneys during the fifth to seventh week of gestation eventually leading to the formation of an accessory kidney (8). These can occur either with two separate collecting systems or as a partially duplicated system in which one ureter drains into the other. In even rarer cases, the duplicate ureter might drain into ectopic locations, such as the vagina and present with urinary incontinence (9).

Supernumerary kidney should not be confused with the more commonly occurring duplex kidney, where a kidney has two collecting systems with a single ureter or two separate ureters. The supernumerary kidney is a separate additional kidney, thought to have a separate arterial supply, venous drainage, collecting system, and distinct encapsulated tissue (10).

Supernumerary kidney can be associated with other congenital malformations. These congenital malformations include horseshoe kidney, ventricular septal defects, neural tube defects, and urethral atresia, vaginal atresia, ectopic ureter implantation, imperforate anus, and duplication of urethra (11,12).

Supernumerary kidney usually does not require any treatment and only a periodic follow up with imaging and renal function test suffices. Nephrectomy is done for diseased or non-functional kidney (13,14).

This case contributes to the existing literature by providing detailed radiological findings of a rare S-shaped fused supernumerary kidney, which can aid in differential diagnosis and clinical management of similar cases.

Teaching point:
Supernumerary kidneys are extremely rare and fused supernumerary kidneys are even rarer. Supernumerary kidney should be differentiated from duplex kidney.

Authors' contributions
Nipun Rajgarhia - Radiologist primarily involved in diagnosing and reporting the case.
Amit Bajpai - Radiologist involved in diagnosing and reporting the case.

Disclosures
None

Consent
Yes

Human and animal rights
Not applicable

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Figures:

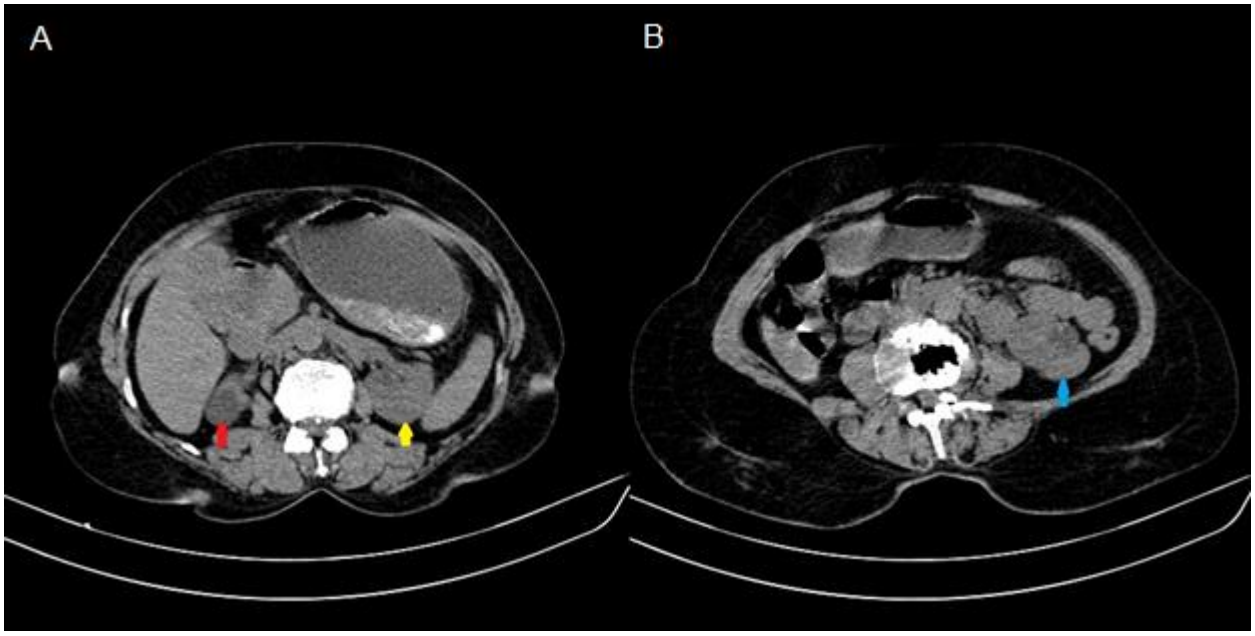

Figure 1: Axial non-contrast CT images. **A,** right kidney (red arrow) and left kidney (yellow arrow) normally located in respective renal fossae. A simple cortical cyst is seen in the right kidney. **B,** supernumerary kidney in the left (blue arrow) with antero-laterally directed renal pelvis.

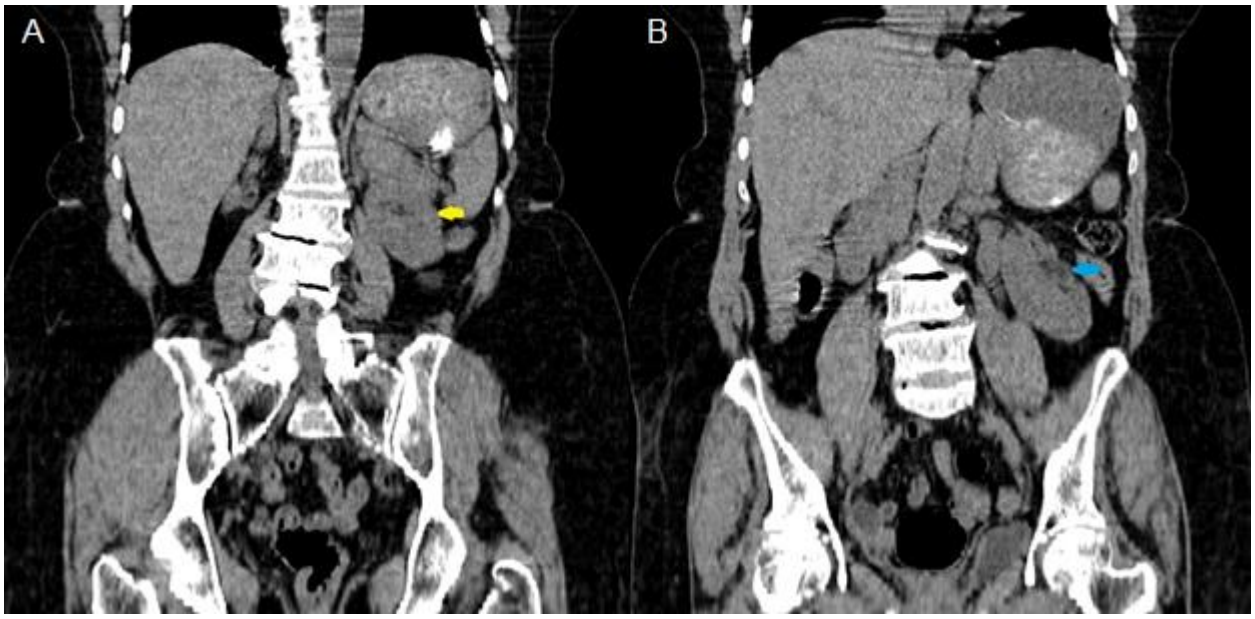

Figure 2: Coronal reformat images. **A,** Cranially located normal left kidney (yellow arrow) with normal collecting system. **B,** Caudally located supernumerary kidney (Blue arrow) with laterally directed collecting system.

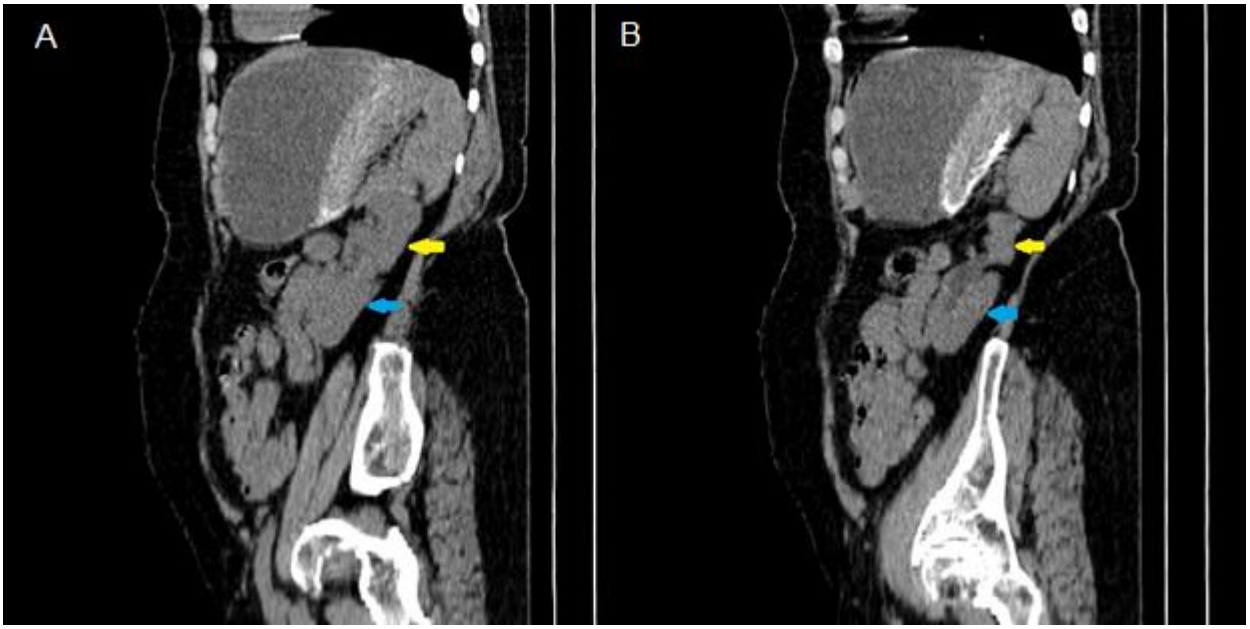

Figure 3: Sagittal reformat images. **A** and **B,** Cranially located normal left kidney (yellow arrow) and caudally located supernumerary kidney (Blue arrow).