CASE REPORT

Ectopic Supernumerary Kidney: A Rare Anatomic Variant

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ABSTRACT

Urogenital anomalies are common, correspond to 33% of all congenital anomalies and supernumerary kidney is one of the rarest entity; less than hundred cases have been reported in the past. The Supernumerary kidney is considered a third kidney in addition to the two already present independent kidneys and considered an infrequent developmental anomaly of the urinary tract. The accessory kidney most commonly located on left side caudal to the normal kidney. Because of uncommon occurrence and prevalence, it frequently causes diagnostic challenges in clinical practice. The supernumerary kidney must be differentiated from the more common congenital anomaly, duplex kidney, having two pelvicalceal systems that are connected with single or double ureters. The supernumerary kidney, in distinction, is thought to be an accessory organ with a separate arterio-venous supply, collecting system and a different capsule. The case of a 33 years old male patient is depicted who presented a history of hypertension and was diagnosed as having left sided unilateral supernumerary kidney located cranial to normal renal parenchyma.

KEY WORDS: Urogenital Anomalies, Supernumerary Kidney, Accessory Kidney.


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INTRODUCTION

Supernumerary kidney is the rarest congenital urological anomaly. Since 1656, less than hundred cases have been reported.\(^1\) Supernumerary kidney is considered a third kidney in addition to the two already present independent kidneys and considered an infrequent developmental anomaly of the urinary tract. The true incidence of this anomaly cannot be estimated because it’s occasional incidence\(^2\). The supernumerary kidney must be differentiated from the more common congenital anomaly, duplex kidney, having two pelvicalyceal systems that are connected with single or double ureters. The supernumerary kidney, in distinction, is thought to be an accessory organ with a separate arterio-venous supply, collecting system and a different capsule. It may be either totally separate from the normal kidney or connected to it by loose connective tissue acting as a connection between the two kidneys. Supernumerary kidney is most frequently seen on left side. It may show a discrepancy in size but is usually smaller and caudal in position to the kidney of the same side. When present caudally, a double ureter is the most commonly seen. If the supernumerary kidney is located cranially in relation to the kidney of the same side, the ureter is frequently totally separate and may open into urinary bladder ectopically\(^3\).

CASE

The present case is about a 33 years old male patient, who was referred to Radiology department by nephrologist for Tri-phasic renal CT scan due to suspicion of left renal mass on previous ultrasound conducted at another peripheral setup. Patient has history of hypertension since eight months and taking ACE inhibitor as antihypertensive. General and systemic examinations were essentially normal apart from raised blood pressure. Routine hematological and biochemical investigations were also within normal limits.

A triple-phasic renal CT scan after intravenous contrast administration was performed on 16 slice Toshiba Aquilion\(^\text{TM}\) scanner. The rate of contrast injection was 4 ml/sec, slice thickness was 3 mm and amount of 2 ml/kg contrast was injected. The Axial (A), Multiplanar Reconstruction (MPR) and 3-Dimensional (3D) images were acquired at 1 mm reconstructed slice thickness which revealed fused supernumerary kidney at upper pole of left kidney (Figure 1).

**Figure 1a & 1b:** Coronal and oblique reconstructed images show normal as well as fused supernumerary kidney on left side cranial to normal kidney. Right kidney appears normal.

On arterial phase, there were three renal arteries supplying the left kidney, directly arising from aorta. Two of them are seen supplying to the supernumerary kidney in which proximal one
Ectopic supernumerary kidney shows decreased diameter, which could be the cause of patient's hypertension. Third one supplies the normal left kidney showing normal caliber. Venous drainage of left kidney proceeds through more than one left renal veins, constituting a common trunk, drains into the inferior vena cava (Figure 2). During excretory phase, there was normal and prompt excretion of contrast through pelvicalyeal systems of left supernumerary as well as left kidney.

Figure 2a & 2b: Coronal and oblique reconstructed image showing left supernumerary kidney and their vascular supply.

There was also evidence of duplication of ureters showing unification just proximal to the uretero-vesical junction. One of them draining the supernumerary kidney and another one is seen arising from normal left renal pelvis. No other pathology including cyst, mass, calculi, or hydronephrosis seen. Right kidney was anatomically normal in all post contrast phases showing normal excretion of contrast in to ureter.

DISCUSSION

Urogenital anomalies are common and correspond to 33% of all congenital anomalies. Supernumerary kidney is the rarest congenital urological anomaly with fewer than 100 cases reported in literature since their first description in 1656. Supernumerary kidney may vary in size but is generally smaller and located caudal to ipsilateral normal renal parenchyma. It can be found in another location such as iliac region or anterior to the sacral promontory. The incidence of supernumerary kidney with fusion and distinct ureter is an exceedingly rare anomaly. The supernumerary kidney is distinctly an accessory organ having its own collecting system, blood supply, and encapsulated parenchyma.

Supernumerary kidneys are embryologically formed by anomalous division of the nephrogenic cord into 2 metanephric blastemas with bifurcation of single bud. The supernumerary kidney may be completely separate from the normal kidney or connected to it by loose connective tissue. The true incidence remains unknown, but in most cases males are afflicted. Out of them most are unilateral and on the left side. This anomaly is congenital, is frequently asymptomatic, but may become symptomatic in early adulthood. Congenital anomalies that may be associated with supernumerary kidneys are horseshoe kidney, ectopic ureteral opening, vaginal atresia, double collecting system, coarctation of aorta and nipple anomalies. These patients are usually prone for many complications as in other congenital renal anomalies such as urinary tract infection, stone formation, pyelonephritis, hydronephrosis, pyonephrosis, and malignant degeneration, therefore, long term follow-up is recommended.

Radiological imaging techniques that were used for diagnosis include ultrasound (US), computed tomography (CT) scan, intravenous urogram (IVU), magnetic resonance imaging (MRI),
Computed Tomography angiography (CTA) and dimercapto succinic acid (DMSA) and diethylene triamine pent acetate (DTPA) scans. It is thought that US, IVU and CT scan seem to be enough for the diagnosis in majority of the cases. Further management depends on symptoms and the function of the supernumerary kidney. No treatment is required if the patient is asymptomatic, but regular follow up may be advised. As in my case patient was hypertensive and supernumerary kidneys was functioning so only need hypertensive medical management. Surgical management of diseased or nonfunctional supernumerary kidneys is a big challenge because of the associated anomalies and non-systemized blood supply. Therefore, if any intervention such as calculus extraction or nephrectomy is planned, further radiological modalities should be undertaken to demonstrate the blood supply and the anatomical variation as well as the associated anomalies.

REFERENCES