Duplex collecting system in a pelvic kidney - an unusual combination

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ABSTRACT

Pelvic kidney is a relatively common renal malformation as well as duplex kidney; however, the unilateral coexistence of these abnormalities is not frequently found. We present a case of a young woman with a history of hypertension in whom a pelvic and dysmorphic left kidney was discovered during ultrasound examination performed for the study of the renal arteries. Magnetic resonance imaging and computed tomography imaging revealed a duplex pelvic kidney. This combination is very rare; we have not found a similar case reported in the literature.

CASE REPORT

CASE REPORT

A 28-year-old woman with a history of hypertension presented at the Radiology Department; her laboratory tests were normal although blood pressure was 190/110 mm Hg.

There was no family history of kidney disease.

US showed an ectopic and dysmorphic left kidney; the renal parenchyma was normal. Color Doppler (CD) study was performed (Fig. 1), but distal tract of renal vessels was not visualized because of gas artifacts. Right kidney had a regular position, morphology and size.

Magnetic resonance imaging (MRI) (Avanto, Siemens Medical Solutions, Forchheim, Germany 1.5 T) of abdomen and pelvis before and after intravenous contrast material was performed. MR Angiography study was not satisfactory for technical reasons. Anyway MRI showed a left pelvic kidney, malrotated, with alterated hilar anatomy and an incomplete duplication of the collecting system (Fig. 2 C). Both suprarenal glands were present at the level of 12th thoracic vertebra.

To get a better visualization of the vascular map, a CT scan (Aquilion 64, Toshiba Medical System, Tokyo, Japan) was performed. The voltage was set at 100 kV, with an automatic mA modulation between 80 and 440 mA and a standard deviation (SD) value of 15. The other parameters were: collimation 64 x 0.5 mm, gantry rotation time 0.5 s, pitch 0.828. Intravenous non ionic contrast material (Iobitridol 350 I/mL; XENETIX 350, Guerbet, Aulnay-sous-Bois, France) was administered through a catheter in the right cephalic vein, followed by administration of 40 mL of saline solution. It was injected at a flow rate of 3.5 mL/second applying an automatic injector.

Longitudinal diameter of the left ectopic kidney was 10 cm, similar to that of the right kidney (9.6 cm). The upper pole was located at the level of L4 and the inferior pole at the S1-S2 intervertebral space. The kidney was malrotated, with altered hilar anatomy and deep lobes; it was vascularised by two distinct arteries: the main artery arose from the aortic bifurcation and descended posteriorly to middle third of kidney; a branch, which penetrated into the parenchyma in the upper pole, originated from this vessel. Another thin artery, that supplied lower pole, originated from the right common iliac artery, just below the aortic bifurcation (Fig. 2, Fig. 3). CD study demonstrated no hemodynamically significant stenoses.

In regard to the venous drainage, there were two renal veins from the upper hilar region, that in turn formed a

common trunk which drained into inferior vena cava. Another independent vein was recognizable at the hilum in lower pole, draining into the right common iliac vein (Fig. 2, Fig. 3).

Contrast excretion was seen normally without any delay. There was no evidence of dilatation of pelvicalyceal system on either side. An incomplete duplication of the collecting system was confirmed: upper collecting system was anterior while lower collecting system was located medially. A short ureter (2.5 cm in length) from the upper system coursed anteriorly and joined the lower pelvis to form a single ureter (12 cm in length) (Fig. 4 A, B), configuring a bifid ureter. The right kidney was normally located; it was supplied by a single renal artery originating from aorta; a single renal vein was seen draining into inferior vena cava. Right pelvicalyceal system and ipsilateral ureter did not show any anatomical variation.

Vescicoureteric junctions of both sides and bladder appeared normal (Fig. 4 C).

Since imaging techniques did not show either significant signs of obstruction with urinary stasis, or signs of infection, as confirmed by laboratory analysis, our patient was chosen to be followed through a regular follow-up with clinical examinations, urine and blood tests, blood pressure monitoring with antihypertensive therapy, with a conservative approach; moreover, since CD study did not demonstrate vascular stenosis neither supine nor in erect station, hypertension was assumed not to be renal-related.

The patient is still followed through a regular follow-up and is alive and in good health.

DISCUSSION

Etiology & Demographics:

Congenital abnormalities of the kidney and urinary tract occur in 3-6 per 1000 live births [1] and can be classified on embryological basis into abnormalities of the renal parenchyma development, aberrant embryonic migration and abnormalities of the collecting system [2].

The incidence of ectopic kidney is 1:12,000 clinical and 1:900 post-mortem cases [3]. Ectopic kidney may be abdominal, lumbar or pelvic, based on its position in the retroperito¬neum; it can be placed either ipsilaterally or contralat¬erally [4]. Ectopic kidney is usually smaller with varying degree of malrotation; it can be fused in 85% with the orthotopic kidney [5]. Its blood supply is from iliac artery or infrarenal abdominal aorta with typically multiple arteries and the ureter has a length according to the location of the kidney [1].

Also duplex kidney is a common abnormality, carrying an incidence of approximately 1% [6,7]; it is found to be up to two times more frequent in female patients and bilateral in less than 20% [8].

Clinical & Imaging findings:

Duplex kidney is defined as a renal unit comprised of two pelvicalyceal systems [9,10]. Duplication, often classified as either complete or partial [11], occurs when two separate ureteric buds arise from a single Wolffian duct [12]; it is characterised by an incomplete fusion of upper and lower kidney moieties resulting in a variety of complete or incomplete duplications of the collecting system [8]. Based on the degree of fusion, it can present as bifid renal pelvis, partial ureteric duplication (Y-shaped ureter), incomplete ureteric duplication with ureters joining near or in bladder wall (V-shaped ureter) and complete ureteric duplication with separate ureteric orifices [11,13].

Although these two anomalies are frequent, their association is very rare. We have not found any similar association reported in literature.

Congenital anomalies of the kidney and urinary tract are part of a family of disease with different anatomical origins [14].

Renal ectopia is one of the most common renal abnormalities of kidney development; it is characterized by abnormal location of the kidney outside the flank region and is often associated with other genitourinary malformations. Most ectopic kidneys are clinically asymptomatic and they are not more susceptible to disease than normally positioned kidneys. The abnormal position of the ectopic kidneys may result in a palpable abdominal mass, direct and referred pain, urinary tract infection and stones, vescicoureteral reflux (VUR), vascular malformations and renovascular hypertension secondary to an anomalous blood supply [15].

Various anatomical abnormalities can contribute to stone formation. Renal abnormalities, polycystic kidney, or obstructions at the ureteropelvic junction or at any other level of the excretory system can cause urinary stasis, which increases the risk of stone formation.

Duplex collecting system is also a common congenital abnormality. Duplex kidney is mostly asymptomatic, incidentally detected and of no clinical significance; however, it can be associated with significant pathology, often with long-term morbidity [8], which is specific of the renal moiety (upper moiety: ectopic ureteric insertion, with or without an ureterocele and multicystic dysplastic moiety; lower moiety: VUR, renal scarring and pyelo-ureteral junction obstruction [6,7]).

In our case, unilateral combination of these malformations was observed: we have not found any similar case in literature. Renal function was preserved, the only symptom was arterial hypertension refractory to medical therapy, probably related to vascular tortuosity given the absence of hemodynamically significant stenosis. This could determine a transient increment of vascular resistances in particular positions (e.g. erect station) which could lead to an hypertensive peak.

The left kidney was located in the pelvis and appeared dysmorphic, with deep lobes, simulating a supernumerary kidney fused with a pelvic kidney.

Treatment & Prognosis:

Management of this condition depends on symptoms and the kidney function. No treatment – as well as for ectopic kidney – is required if urinary function is normal and no obstruction to the urine flow is observed [16]; if an obstruction is present, surgery may be required to correct the location of the kidney and to allow better drainage of urine, in order to reduce infections, urinary stasis and consequent stones formation. Reflux can be repaired by surgery to modify the course of the ureter. The position of the kidney, vascular tortuosity and vessels' diameter may determine an increment of blood pressure causing hemodynamically significant stenosis. In case of extensive renal damage or malignancy, nephrectomy is indicated [17].

Differential Diagnoses:

Supernumerary kidney can be distinguished from duplex kidney [18], which is more often encountered. It is a rare anatomic anomaly with less than 100 cases reported in the literature; it is generally diagnosed in young people, affects males and females equally and involves mostly the left side [14]. A supernumerary kidney may be of the same size or more commonly smaller than usual kidney and located caudally rather than cranially to the ipsilateral kidney [19]. It can be totally apart from the original kidney or connected to it by a sheath of connective tissue [20] or by a portion of parenchyma [21]. As concern excretory system, the supernumerary kidney can be drained by a bifid ureter or by a separate ureter [18], the former condition being a little more frequent than the latter (53%) [22]. The supernumerary kidney is considered to be an accessory organ with a separate arterial supply, venous drainage and usually separate encapsulated tissue [23].

Duplex kidney, however, has a single continuous capsule and does not have a separate arterial supply. The number of calyces is greater in supernumerary kidney, while it is equal to that of the opposite kidney in the duplex system [18].

In our case report CT scan showed a pelvic duplex kidney, because the two moieties of the kidney did not have a separate arterial supply (the main artery supplied both upper and lower portion of the kidney, the accessory one supplied the lower portion); furthermore, only a single capsule was identified. The vein anatomy (two veins) as well as excretory system anatomy (double system with a Y-shaped ureter) did not give an useful contribution to differential diagnosis.

TEACHING POINT

Pelvic kidney is a relatively common renal malformation as well as duplex kidney but the unilateral coexistence of these abnormalities is not frequently found. Ultrasonographic imaging, as first line imaging is not always diriment; it may highlight an ectopic kidney and/or a malrotation, To get a better visualization of the vascular map, a CT scan must be performed, also to obtain useful information in anticipation of an eventual surgical approach. Management of this condition depends on symptoms and on the kidney function and no treatment is required if urinary function is normal and no obstruction to the urine flow is observed with imaging techniques.

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FIGURES

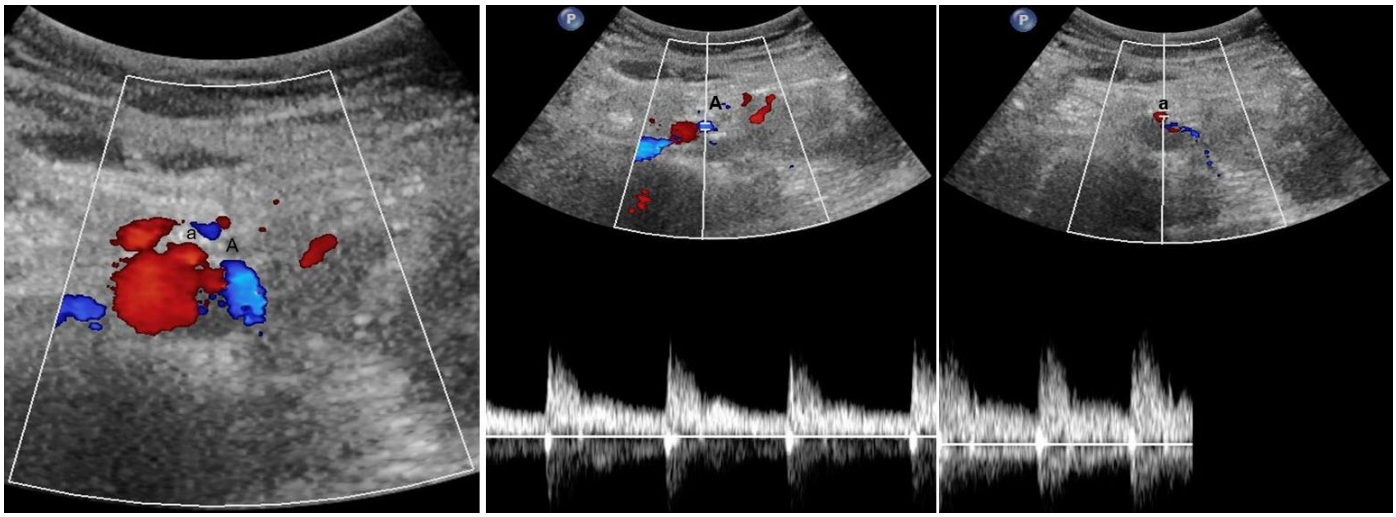

Figure 1: 28-year-old woman with duplex collecting system in a pelvic kidney.
Findings: CD-US images demonstrate two distinct arteries: the main artery (A) and another thin artery (a), that supplied lower pole, originated from the right common iliac artery, just below the aortic bifurcation. The exam demonstrated no hemodynamically significant stenoses.

Technique: CD performed on PHILIPS IU22 with 5-2MHz Curved Transducer at a frequency of 5.0 MHz.

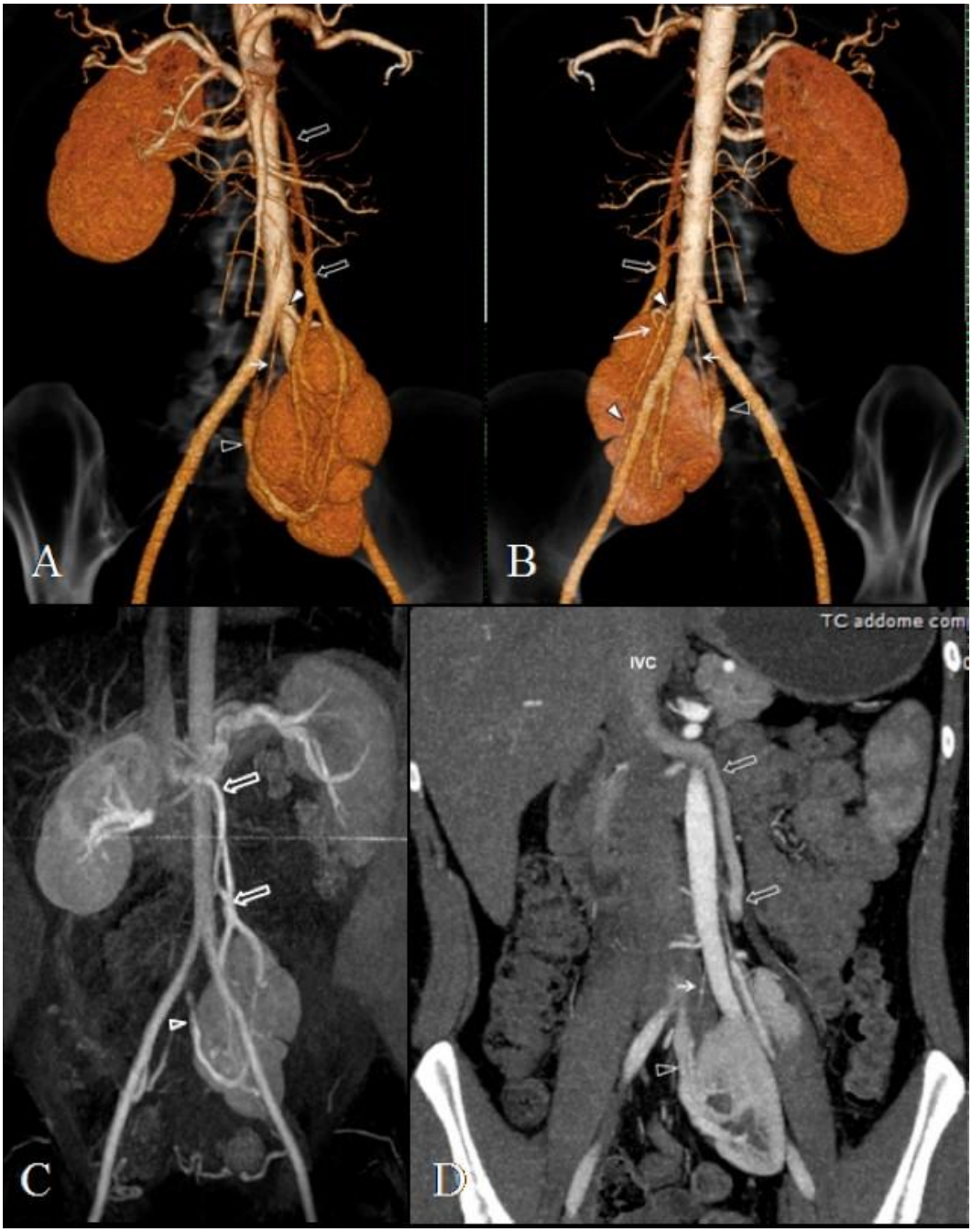

Figure 2: 28-year-old woman with duplex collecting system in a pelvic kidney.
Findings: CT-Angiography VR anterior view (A), CT-Angiography VR posterior view (B), MR-Angiography coronal MIP (C) and CT-Angiography coronal MIP (D) images demonstrate a left pelvic kidney, dysmorphic and malrotated, with deep lobes, altered hilar anatomy and complex vascularization. The main renal artery - that arises from the aorta, at the bifurcation, in the middle seat - is seen for a short tract (full arrowhead), as it moves to the posterior surface of the kidney and descends to the middle third. A branch, originating from the main artery, penetrates into the parenchyma at the upper pole (long arrow). An accessory renal artery (short arrow) arises from the right common iliac artery, just below the bifurcation, on the medial side and supplies the lower pole. Two renal veins, that originate from upper renal hilum, form a common trunk (empty arrows) which drains into the inferior vena cava (IVC). Another independent vein is recognizable at the lower hilum (empty arrowhead), draining in the right common iliac vein.

Technique: Scan acquired in the angiographic phase on Aquilion 64, Toshiba Medical System CT scanner (Tokyo, Japan). The voltage was set at 100 kV, with an automatic mA modulation between 80 and 440 mA and a standard deviation (SD) value of 15.; beam collimation 64 x 0.5 mm, gantry rotation time 0.5 s, pitch 0.828.
110 cc Iobitridol 350 mgI/mL intravenous contrast administered at 3.5 mL per second. MR-Angiography sequence acquired on Avanto, Siemens Medical Solutions, 1.5 T (Forchheim, Germany) Spoiled Gradient Echo (TR 2.94ms TE 1.06ms) slice thickness 1.10mm GAP 0. 7.0 mL Gadobutrol 1.0 mmol/mL contrast injected at 3.0 mL per second.

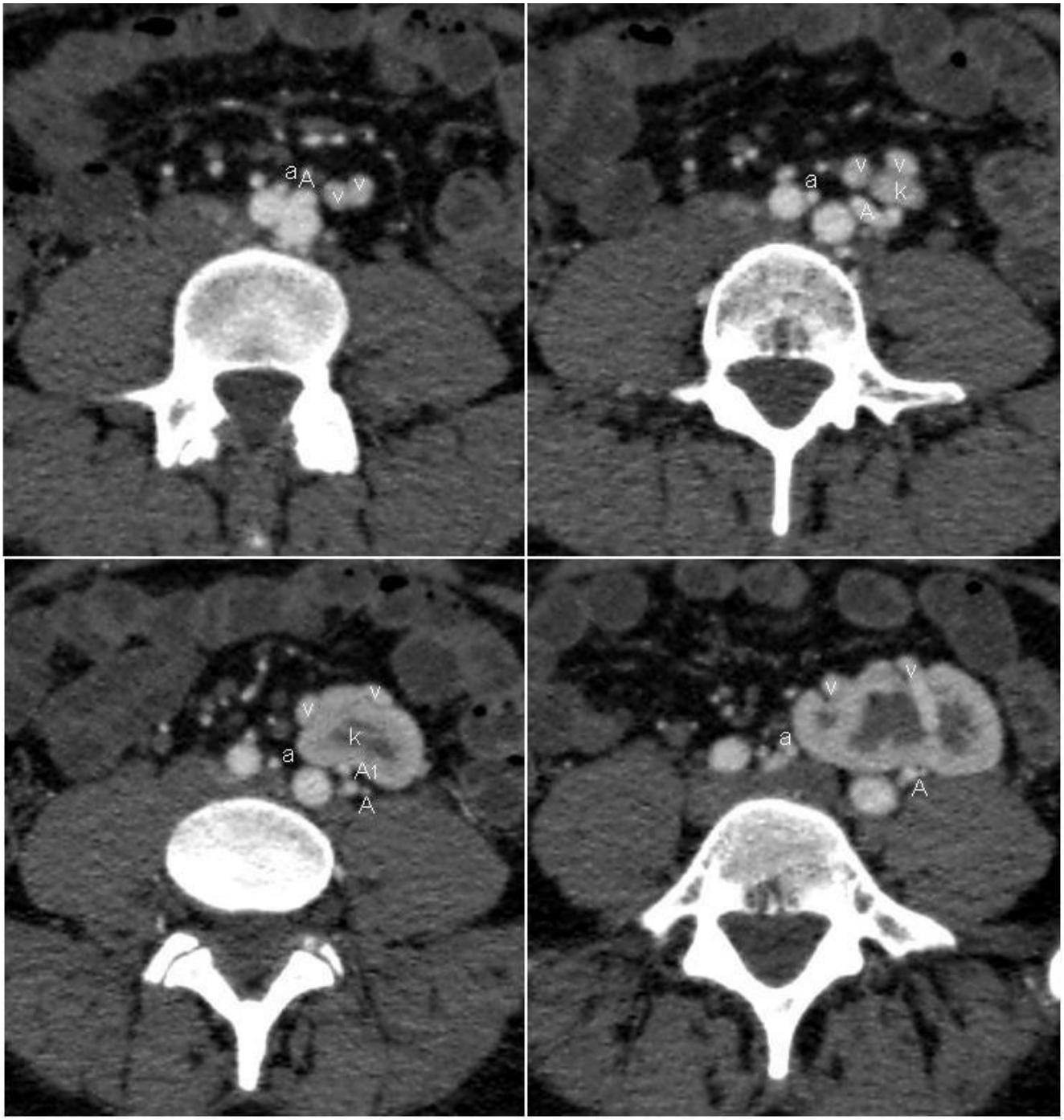

Figure 3: 28-year-old woman with duplex collecting system in a pelvic kidney.
Findings: Serial axial contrast-enhanced CT images of the abdomen from superior to inferior (A, B, C, D).
The main renal artery (A) arises from the aorta, at the carrefour, in the middle seat; it moves to the posterior surface of the kidney (k) and descends to the middle third . A branch, originating from the main artery, penetrates into the parenchyma at the upper pole (A1). An accessory renal artery (a) arises from the right common iliac artery, just below the bifurcation, on the medial side; it supplies the lower pole. Two renal veins (V) originate from upper renal hilum which is rotated anteriorly.

Technique: Scan acquired in the angiographic phase on Aquilion 64, Toshiba Medical System CT scanner (Tokyo, Japan). The voltage was set at 100 kV, with an automatic mA modulation between 80 and 440 mA and a standard deviation (SD) value of 15.; beam collimation 64 x 0.5 mm, gantry rotation time 0.5 s, pitch 0.828. 110 cc Iobitridol 350 mgI/mL intravenous contrast administered at 3.5 mL per second.

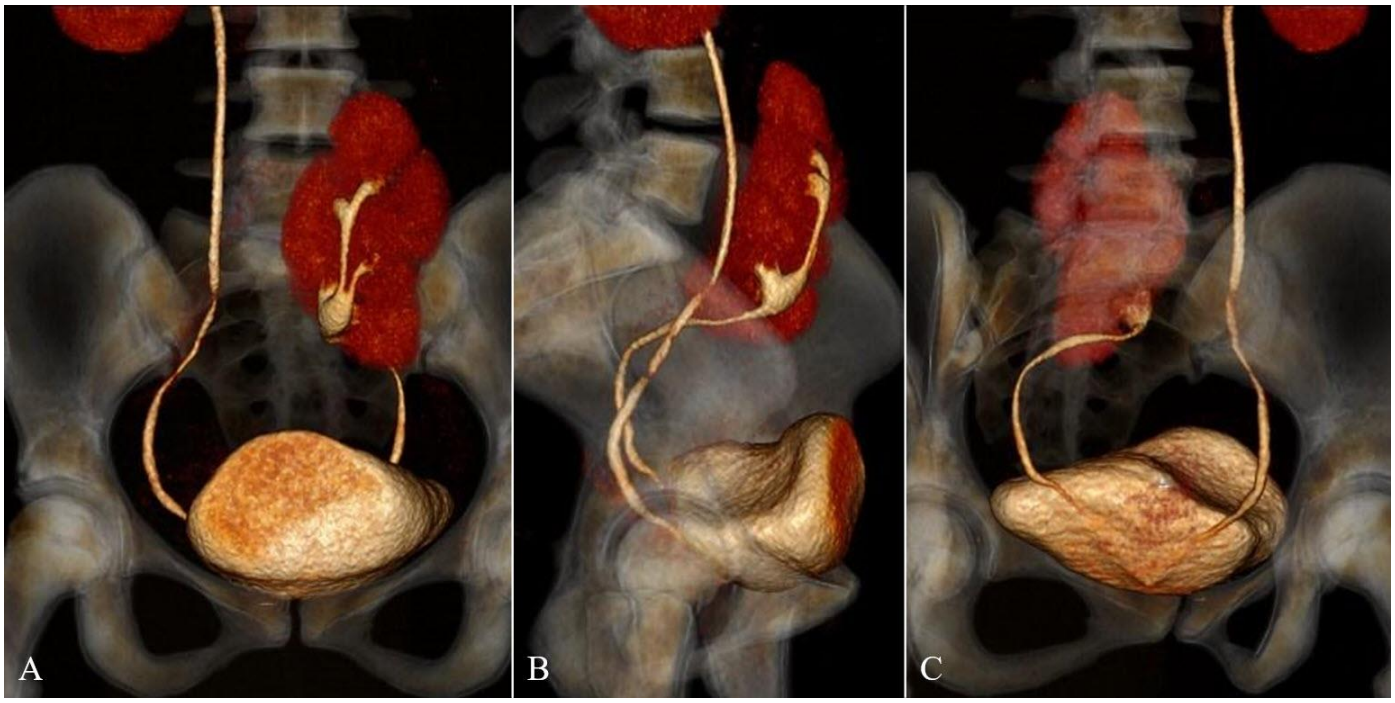

Figure 4: 28-year-old woman with duplex collecting system in a pelvic kidney.
Findings: 3D VR images (anterior view: A; lateral view: B; posterior view: C) demonstrate a left pelvic kidney with a duplex excretory system. A, B) Upper collecting system is anterior, while lower collecting system is located medially; a short ureter (2.5 cm in length) from the upper system courses anteriorly and joins the lower pelvis to form a single ureter (12 cm in length). C) Vescicoureteric junctions of both sides are normal.

Technique: Scan acquired in the excretory phase (12 minutes after contrast administration) on Aquilion 64, Toshiba Medical System CT scanner (Tokyo, Japan). The voltage was set at 100 kV, with an automatic mA modulation between 80 and 440 mA and a standard deviation (SD) value of 15.; beam collimation 64 x 0.5 mm, gantry rotation time 0.5 s, pitch 0.828. 110 cc Iobitridol 350 mgI/mL intravenous contrast administered at 3.5 mL per second.

	Supernumerary	**Duplex**
Etiology	Congenital	Congenital
Incidence	The true incidence of this anomaly cannot be calculated because of its infrequent occurrence (less than 100 cases in literature)	1:100
Gender ratio	-	F:M=2:1
Age predilection	-	-
Risk factors	-	-
Treatment	None if asymptomatic	None if asymptomatic
Prognosis	Influenced by urinary function and eventual complications	Influenced by urinary function and eventual complications

Table 1: Summary table for supernumerary kidney and duplex kidney.

	Supernumerary	**Duplex**
Parenchyma	Usually completely separate/ partial fusion	Both poles attached to each other
Capsule	Separate or continuous	Continuous
Number of calyces	Greater	Does not exceed from the opposite side
Vascular supplies	Separate arterial supply and venous drainage	Same vascular supply
Pattern of contrast enhancement	Generally homogeneous (influenced by urinary function and eventual complications)	Generally homogeneous (influenced by urinary function and eventual complications)
Radiopharmaceutical uptake	Generally homogeneous (influenced by urinary function and eventual complications)	Generally homogeneous (influenced by urinary function and eventual complications)

Table 2: Differential diagnosis table for supernumerary kidney and duplex kidney based on imaging.

ABBREVIATIONS

CD = Color Doppler
CT = Computed Tomography
MRI = Magnetic Resonance Imaging
SD = Standard Deviation
US = Ultrasound
VUR = Vescicoureteral Reflux

KEYWORDS

Kidney; Ectopic; Duplex; Abnormalities; CT