Fused supernumerary kidney

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A partially fused supernumerary kidney with bifid ureters was diagnosed in a 35-year-old female patient using ultrasound and triple-phased computed tomography (CT). The CT also revealed two separate renal arteries supplying each of the left kidney and multiple left renal veins, forming a common trunk to drain into the inferior vena cava. Supernumerary kidney is a rare congenital anomaly; fewer than 100 cases have been reported over the years. A fused supernumerary kidney is rarer still.

Case report

A 35-year-old female presented with pain in the right iliac fossa over a period of two days. General examination revealed tenderness at McBurney's point. Routine hematological and biochemical investigations were within normal limits except for a raised total leucocyte count.

Ultrasonography of the abdomen revealed two kidneys on the left, with the pelvicalyceal system of one directed medially (measuring 8.1 x 4.0 cm) and the other directed laterally (measuring 8 x 3.8 cm). The two kidneys were seen to be attached loosely by a parenchymal bridge. The right kidney measured 9.1 x 4.8 cm and appeared normal in site. The right iliac fossa was obscured by gas.

A triple-phasic CT scan with intravenous contrast administration revealed a normal right kidney measuring 10.1 x 5.1 cm and two left kidneys. The cranially located kidney measured 8.3 x 3.7 cm, and the caudally located kidney measured 8.0 x 3.5 cm. These kidneys were partially fused and demonstrated a small parenchymal bridge between them (Fig. 1).

Two distinct main renal arteries supplied the fused left kidneys. The cranially placed kidney received its arterial supply from a branch of the abdominal aorta. The caudally placed kidney received its arterial supply from a branch of the left common iliac artery (Figs. 2, 3). The caudally placed kidney drained through two renal veins, and the cranially placed kidney drained through one renal vein. These veins in turn formed a common trunk and drained into the inferior vena cava (Fig. 4). The vascular supply to

Figure 1. 35-year-old female with right iliac fossa pain. Triphasic CT with 60ml of intravenous contrast administration. 3-D volume rendering of the arterial phase (oblique view) showing a normal right kidney (white arrow) and two left kidneys (yellow arrows) partially fused by a parenchymal bridge between them (blue arrow).
The right kidney was through a single main renal artery and renal vein and did not show any anatomical variation. A normal excretory phase was demonstrated by the kidneys bilaterally, with no delay in excretion of contrast. The supernumerary kidneys on the left showed a bifid ureter. The ureter from the cranially placed kidney passed anteriorly and joined the pelvis of the caudally placed kidney at the pelviureteric junction to form a single ureter (Fig. 5). The pelvicalyceal systems of both left kidneys were not dilated. The pelvicalyceal system of the kidney lying cranially was normally sited (Fig. 6). The caudally placed kidney was malrotated, with the pelvicalyceal system facing laterally (Fig. 7). The right pelvicalyceal system and right ureter did not reveal any anatomical variation or pathology. Ureterovesical junctions were normal, and there was no ectopic ureteral opening bilaterally. CT further revealed features consistent with acute appendicitis, and the patient was surgically managed for this.

Discussion

A supernumerary kidney is a third kidney (in addition to the two independent kidneys). This is a rare congenital anomaly of the urinary tract. The true incidence of this anomaly cannot be calculated because of its infrequent occurrence (1). The embryological basis for this anomaly is thought to be the abnormal division of the nephrogenic cord into two metanephric blastemas that then form two kidneys, in association with either a partially or completely duplicated ureteral bud (2). The supernumerary kidney needs to be differentiated from the more commonly occurring duplex kidney, which is defined as having two pelvicalyceal systems that are associated with a single ureter or with double ureters. The supernumerary kidney, in contrast, is thought to be an accessory organ with a separate arterial supply, venous drainage, collecting system, and distinct encapsulated tissue. It may be

Figure 2. 35-year-old female with right iliac fossa pain. Triphasic CT with 60mL of intravenous contrast administration. 3-D volume rendered image in the arterial phase shows a branch from the abdominal aorta supplying the cranially placed kidney (yellow arrow) and a branch from the left common iliac artery supplying the caudally placed kidney (white arrow). Single right renal artery (red arrow) is also visible.

Figure 3. 35-year-old female with right iliac fossa pain. Coronal reconstruction of contrast-enhanced CT images in the venous phase shows the three renal veins forming a common trunk and joining the inferior vena cava. The two renal veins drain the caudally placed kidney (white arrow), the renal vein drains the cranially placed kidney (red arrow) and in turn drains into the common trunk (yellow arrow) before joining the inferior vena cava. (Kvp: 120, mas: 100, 5 mm thickness, 60 mL of omnipaque intravenous contrast)
either totally separate from the normal kidney or connected to it by loose areolar tissue acting as a bridge between the two kidneys (2, 3).

The supernumerary kidney is most often seen on the left side of the abdomen (2). It may vary in size but is usually smaller and caudal in position to the ipsilateral normal kidney. When present caudally, a bifid ureter is the most commonly seen presentation (2).

If the supernumerary kidney is located cranially in relation to the normal kidney, the ureter is usually completely separate and may enter the bladder ectopically (2), in which case the Weigert-Meyer rule may be followed and the ureter may insert medially and inferiorly into the bladder (2). Sometimes the ureter of the supernumerary kidney may be associated with an ectopic opening, such as into the vagina (2). In this case, it may present with symptoms of urinary incontinence.

Although the present case had no symptoms, symptoms have been noted in about two-thirds of the cases of supernumerary kidney (4). The most commonly associated pathologies include hydronephrosis, pyelonephritis, pyonephrosis, renal and ureteral calculi, carcinoma, papillary cystadenoma, and Wilm’s tumors (5, 6).

A few anomalies have also been associated with supernumerary kidneys such as ureteral atresia, vaginal atresia, horseshoe kidney, complete duplication of urethra and penis with ectopic ureteral opening into the vagina or introitus, imperforate anus, ventricular septal defects, meningo-myelocoeles, and coarctation of the aorta (1, 7, 8, 9).
For the diagnosis of supernumerary kidney, intravenous pyelography, ultrasonography, nuclear scintigraphy (for function), CT, and MRI can be used.

Management of this condition depends on symptoms and the function of the supernumerary kidney. If the patient is asymptomatic, as in our case, no treatment is required, but regular followup maybe advised. If the kidney is diseased or nonfunctional, nephrectomy is usually the preferred procedure (6, 8).

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