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Case Report

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Imaging of anatomic variations in a case of supernumerary kidney

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Abstract

Supernumerary kidneys are extremely unusual entities. We present the case of a 14-years-old female with three well-functioning kidneys in whom a reniform abdominal mass was discovered during ultrasound examination performed for right lower quadrant pain. Herein we present the excretory urography, computed tomography, and CT angiography findings in this patient. This paper also describes the aberrant trajectory of some vessels and other associated variants. Knowledge of this condition (and its vascular anatomy) is interesting because it has important implications for any surgical planning and for possible complications.

Keywords: Supernumerary kidneys, Anatomic variation, Renal vein, Pediatric radiology.

INTRODUCTION

Supernumerary kidney is a rare congenital anomaly, with only few cases described in the international literature [1-6]. A fused supernumerary kidney is a still rarer entity [6]. It constitutes a diagnostic and therapeutic dilemma.

The supernumerary kidney is truly an accessory organ with its own collecting system, blood supply, and distinct encapsulated tissue. It may be either totally separate from the normal kidney or connected to it [2].

Because of the associated anomalies, different anatomical environment and non-systemized blood supply, surgical and endovascular management of these patients is very difficult. Therefore, correct diagnosis is crucial before any surgical intervention although many cases have not been discovered until the operation. The aim of this report is to contribute with a case of supernumerary kidney and describe its anatomic variants.

CASE REPORT

A 14-year-old female patient presented to the emergency room with complaints of right lower quadrant pain of variable intensity. A gynecologic transabdominal ultrasound showed a right complex ovarian mass and some days later a follow-up one showed non relevant gynecologic findings but a reniform mass was view in the right-middle abdomen arouses the suspicion of right supernumerary kidneys.

Urine analysis and culture as well as serum blood values were within normal limits.

Plain-Film: Convex leftward scoliosis. Transitional vertebra at the level of the thoracolumbar junction with partial fusion of the two vertebral bodies. Posterior defect of the first sacral arch. Rachischisis of the last sacral segments.

Contrast-enhanced CT revealed three kidneys (Fig. 1); two right malrotated renal units joined by a well functionating section of parenchyma (Figs. 1 c and 2 b) and one left kidney (Figs. 1 b and 2). The supernumerary (lower) kidney lay to the right in mesogastrium at the level of the ipsilateral common iliac artery, next to the anterior abdominal wall and with its upper pole fused with the lower pole of the superior right kidney (Figs. 1 c and 2), its hilum was turned outwards (Figs. 1 a and 2 a) and was bigger (93 x 42.24 x 32.30 mm) than superiorly located kidney (79 x 33.79 x 41.66 mm). Collecting systems of the two right kidneys are united by a bifid ureter connecting at the level of the ureteropelvic junction of the lower

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kidney. The three kidneys had good corticomedullary differentiation with appropriate parenchyma and normal nephrographic and excretory phases.

Multidetector CT Angiography (MDCTA) was performed to demonstrate the renal vascular anatomy, and revealed three right renal arteries; two of them were originating from the abdominal aorta (Fig. 2) (the first branch toward the upper unit and the second branch toward the lower unit). The third right renal artery (supplying too the lower kidney) was arising from the posterolateral wall of the right common iliac artery and passed with an ascending course outwards the posterior aspect of the organ (Fig. 2). On the left side, there were two renal arteries (Fig. 2 a): one arising from the lateral aspect of abdominal aorta (as usually) at the level of superior mesenteric artery (higher than habitually) and the second one arising below the inferior mesenteric artery more ventral than the upper polar artery.

The two rights units had separate arterial supply but unified venous drainage (Figs. 2 a, and 3). A bridging vein between right renal units could be seen (Fig. 2 a). Inferior Vena Cava (IVC) received as tributary only one thick venous trunk from the two right renal units (Fig. 3) while two left renal veins were well opacified (Figs. 2 a and 4). The right single venous trunk empty into IVC at an acute angle to its course (instead of the habitual approximately right angle) and it converged in the IVC at a similar level of the superior left renal vein. At this point, the right renal vein had a caliber of 12 mm while its left counterpart has a diameter of 6.02 mm. The inferior left renal vein had a diameter of 5.89 mm. The left superior renal vein had an anterior course to the aorta (classic anatomy) (Fig. 4 a) and had two branches. The left inferior renal vein has initially a downward path, and then, the vein curves medially passing behind the aorta (retroaortic vein) (Figs. 2 b and 4 b). It drains separately from the kidney and independently into the inferior vena cava. A small section of a posterior branch (not opacified appropriately on late arterial phase images) connected to the left inferior renal vein, that was difficult to differentiate from a lumbar or ascending lumbar vein, was encountered (Fig. 2 b).

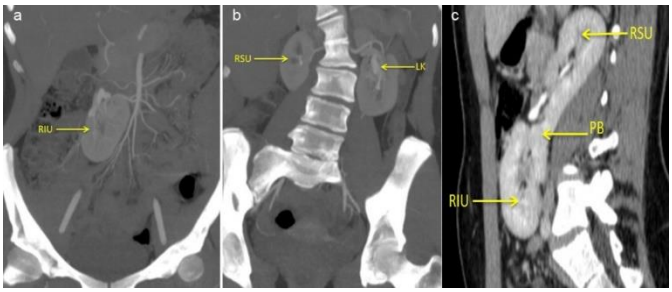

Figure 1: A and B. Coronal reconstruction of contrast enhanced CT images showing right supernumerary kidney (RIU), markedly malrotated, lower and more ventral than the right superior kidney (RSU). LK: left kidney. C. Sagittal reconstruction of contrast enhanced CT image showing the parenchymatous isthmus (PB) that joins the two right renal units. RSU: right superior unit. RIU: right inferior unit.

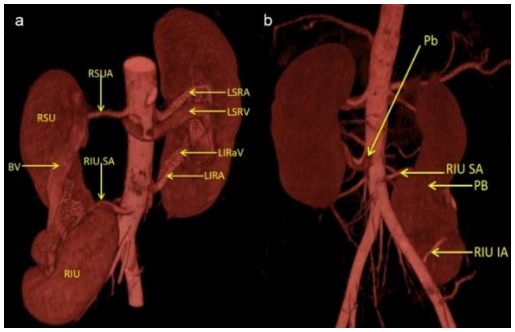

Figure 2: Multidetector CT angiograms. A. Anterior volume-rendered 3D images. A. RSUA: Right superior unit artery. RSU: Right superior unit. BV: Bridging vein between right renal units. RIU SA: Superior artery for the inferior unit arising from aorta. B. Posterior volume-rendered 3D image. Pb: Posterior branch of the left inferior and retroaortic renal vein that was difficult to differentiate from the lumbar vein or ascending lumbar vein. RIU SA: Superior artery of the right inferior unit. PB: Parenchymatous bridge between the two rigt units. RIU IA: Inferior artery of the right inferior unit originating from the right common iliac artery.

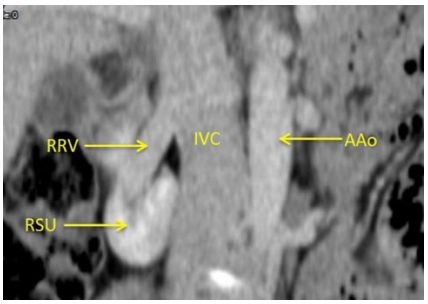

Figure 3: Coronal reconstruction of contrast enhanced CT image. Inferior Vena Cava (IVC) receives as tributary only one vein (RRV) from the two right renal units. RSU: right superior unit. AAo: Aorta.

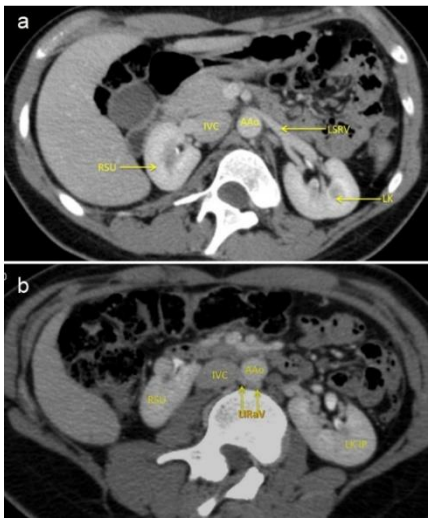

Figure 4: Axial contrast enhanced CT images. A: At the level of the left superior vein. LSRV: Preaortic (normal) left superior vein with two branches converging to 18.5 mms from the left lateral wall of the aorta. LK: Left kidney (superior pole). B: At the level of the left inferior vein. LIRaV: Left inferior (retroaortic) vein. LK IP: Left kidney inferior pole. A and B. IVC: Inferior Vena Cava. AAo: Aorta. RSU: right superior unit.

The patient was asymptomatic. A conservative approach was decided with periodic follow-up including urine analysis and culture, blood analysis and abdominal ultrasound.

DISCUSSION

Supernumerary kidneys are formed by aberrant division of a single nephrogenic blastema or initiate as two separate nephrogenic blastemas [2-4, 6, 7]. Two separate encapsulated kidneys are formed when partially or completely duplicated ureteral stalks enter these blastemas [2, 6].

Few reports describe the associated anomalies like ureteral and vaginal atresia, complete duplication of urethra and penis, ectopic ureteral opening, horseshoe kidney, coarctation of the aorta and megaureter [2, 3, 4]. However in this case, none of these anomalies could be detected.

Most of the patients are symptomatic. Usual presenting symptoms are fever, pain, and palpable abdominal mass [2, 6]. Nevertheless, other patients are asymptomatic. Our patient complained about pain in the right inferior quadrant related with the ipsilateral ovary but not with the supernumerary kidney although the abdominal mass could be easily palpable.

Because of the high rate of complications such as hydronephrosis, stones, pyonephrosis, pyelonephritis and malignant changes (Wilms' tumor, clear cell carcinoma) reported [2, 3, 6], long term follow-up is mandatory. Management depends on the function and associated symptomatology.

Ultrasound, nuclear scintigraphy (also determine the level of function of each kidney), CT scan, and MRI are enough to make the correct diagnosis of supernumerary kidneys and associated anomalies in majority of the cases [1-3, 5, 6, 8]. However, if surgical intervention is planned, further radiological imaging studies are recommended [3]. MDCTA or MR angiography could be performed to demonstrate the blood supply and the anatomical environment as well as the associated anomalies [2, 3]. MR angiography is the alternative to MDCTA to visualize the vessels in cases of iodine allergy.

In our case CT was diagnostic and clearly demonstrated the fusion of two right kidneys. MDCTA allowed us to understand the complex vascular anatomy of kidneys non-invasively. We did not need to perform scintigraphic examination because a satisfactory drainage from the collecting systems had been showed by CT. We believe that ultrasound and CT (MDCTA) seem to be enough for the diagnosis of the condition and associated anomalies.

The highly complex embryological development of the left renal vein compared to its right counterpart results in greater variations which are clinically significant [8]. There are two left renal veins in our patient separated to a distance of 24 mm so we consider them two veins (a main superior and an additional inferior) for separate (Figs. 2 and 4) more than a renal collar. Additional renal vein is any additional vessel that drains separately from the kidney and independently into the inferior vena cava and should be considered as a normal variation. Retro-aortic vein is a single ectopic trunk in a relatively low position, with a trajectory that is oblique inferiorly and retro-aortic while renal collar (circum-aortic venous ring, circum-aortic vein, circum-aortic renal venous collar) is defined as the occurrence of a renal venous channel coursing both anteriorly and posteriorly to the abdominal aorta [8]. The circum-aortic renal venous collar is composed from a ventral inter-subcardinal anastomosis and by a small part of the right and left sub-posterior cardinal anastomosis. On either side, the right and left sub-supracardinal anastomosis forms the ring, while dorsally it is complete by the inter-supracardinal anastomosis. Usually the ventral portion of the circum-aortic plexus persists as the normal left renal vein. If the dorsal portion of the plexus persists, then the left renal vein is posterior to the aorta (retro-aortic renal vein). If both the dorsal and the ventral portions persist, there will be a circum-aortic venous collar [8, 9].

In this case a vein with two tributaries that converge to 18.5 mm of the left lateral wall of the aorta, near of the zone of late confluence [10], drains the blood of the superior half of the left kidney (Fig. 4 a).

After discussing this case with our pediatric urologist and due to her age and the absence of associated complications, we suggest sonographic examination once a year if the patient is not symptomatic.

Considering the complex congenital abnormality, it is advisable to visualize the vascular anatomy before any surgery in all cases of supernumerary kidneys. Preoperative knowledge of vascular variants help the surgeon anticipate these anomalies, facilitates the safe performance of many abdominal surgeries and avoid inadvertent ligation or transection of these vessels.

Conflict of interests. The authors have none to declare

Informed consent. Was obtained from the mother.

Author contributions

Case management – V.F., C.M. Patient follow-up –V.F. Literature Search – V.F. C.M. Writing – V.F.; Critical Reviews – V.F., C.M. All authors approved the final version of the manuscript.

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