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Case Report

Congenital kidney malformation: Computed tomography and scintigraphy findings of renal reversed rotation

Francesca Di Giuliano, MD^a, Eliseo Picchi, MD^{a,*}, Adriano Lacchè, MD^a, Alessio Bozzi, MD^a, Yuri Cavaleri, MD^b, Francesco Garaci, MD, PhD^a, Roberto Floris, MD, PhD^a

^aFondazione PTV Policlinico “Tor Vergata”, University of Rome “Tor Vergata”, Department of Biomedicine and Prevention, Radiology UOC, Viale Oxford, 81, 00133 Rome, Italy

^bFondazione PTV Policlinico “Tor Vergata”, University of Rome “Tor Vergata”, Department of Surgery; Urology UOC, 00133 Rome, Italy

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ABSTRACT

Renal reversed rotation is a congenital abnormality due to anomalous kidney rotation around its long axis, resulting in faced laterally renal hilum; this condition is associated with several vascular variants. The aim of this report is to describe the case of a 47-year-old male with renal reversed rotation who came to our attention after ultrasound exam. The association of renal rotation and vascular anomalies increases long-term complications as kidney lithiasis, hydronephrosis, colic pain, hematuria, and renal failure. We decided to study the anatomy and renal function of the reversed rotated kidney by multiphasic computed tomography and renal dimercaptosuccinic acid-scintigraphy. It is critical for urologists and radiologists to recognize this abnormality in order to plan the best treatment and follow-up.

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Introduction

Congenital abnormalities of the kidney and urinary system are the most common abnormalities in newborn and represent approximately 30% of all prenatally diagnosed malfor-

mations [1]. These conditions may be asymptomatic for a lifetime or lead to several complications both in childhood and in adulthood as infections, stone formation, hypertension, and renal failure [2].

Due to various clinical presentations, early diagnosis is critical for the best treatment of these abnormalities;

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* Corresponding author.

E-mail address: eliseo.picchi@hotmail.it (E. Picchi).

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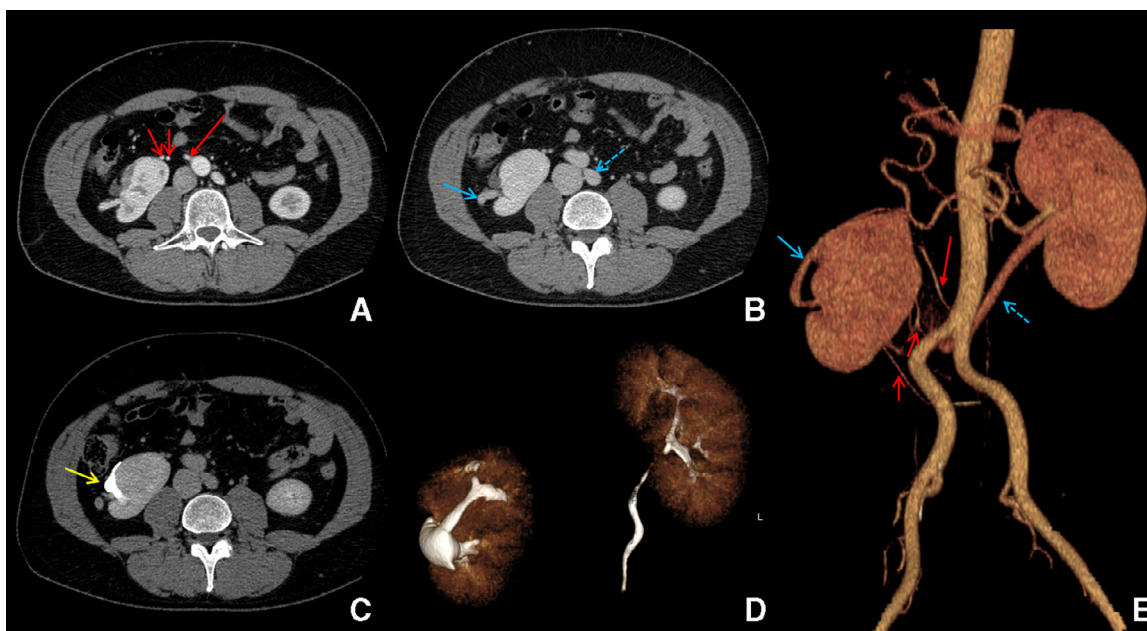


Fig. 1 – Dynamic contrast-enhanced CT study on axial plane at level of L3 and Volume Rendering. (A) arterial vessel (long-close red arrow: main renal artery; short-open red arrow: accessory renal artery); (B) renal veins (blue arrow: right renal vein; blue-dotted arrow: retro-aortic course of left renal vein); (C) urography phase (yellow arrow: left renal pelvis); (D) Volume Rendering reconstruction of the urography phase showing anomalous left renal pelvis; (E) Volume Rendering reconstruction of the main right renal artery arising at aortic bifurcation (long-close red arrow) and of the accessory arteries (short-open red arrows) arising from the common right iliac artery; left (dotted blue arrow) and right renal vein (blue arrow). Note the anomalous course of the left renal vein which drains into the inferior venous cava near confluence of the 2 common iliac veins. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

therefore, radiologists and urologists need to promptly recognize kidney malformations and their potential complications during imaging studies in order to begin timely treatments.

Case report

A 47-year-old male was scheduled for computed tomography (CT) exam further to a previous abdomen ultrasound, performed for screening that showed alteration of his right kidney. The patient had no history of fever, pain or surgical intervention; blood and urine tests were normal.

Contrast-enhanced CT examination was performed by a 64-slice CT scanner (Lightspeed; General Electric Healthcare, Waukesha, WI). The protocol included a noncontrast CT scan, with 5 mm of thickness, and dynamic acquisition at 35, 90, and 300 seconds after administration of iodine contrast medium (Iobitridolo; volume: 120 mL; flow rate: 2.5 mL/s), slice thickness: 2.5 mm, that showed ectopic, smaller, dysmorphic right kidney with reverse rotation. While right kidney pelvis was turned sideways, the renal arterials were located anteriorly. The lateralized calyces were distorted without visible obstruction. The right kidney showed 4 renal pelvis and 4 blood renal arteries, one of which originating from the aortic carrefour 2 from the proximal third of the right common iliac artery and the last one from its medium third. It also revealed 4 renal veins, the biggest of which with a retro-renal

course. Its right ureter, due to hyper-rotation, featured a slower excretory phase (Figs. 1 and 2). Renal dimercaptosuccinic acid-scintigraphy showed inhomogeneous parenchymal radiotracer distribution with stasis of radioactive urine in the lower right calyceal group: this finding was partially resolved in orthostatism (Fig. 3).

Glomerular filtration rate, calculated with Gates method, was normal (87.35 mL/min; lower limit for age 80 mL/min) with asymmetrical distribution between the kidneys (64% left, 36% right). Serum creatinine level was normal: 0.85 mg/dL (0.70-1.30 mg/dL). Also, blood urea nitrogen was in the normal range: 33 mg/dL (15-50mg/dL) and so were blood pressure values after 24-hour Holter monitoring.

The patient showed no symptoms, so he was advised on regular follow-up via blood tests and ultrasound scans.

Discussion/conclusion

Congenital abnormalities of kidney and urinary tract occur in 3.3%-11.1% of the population. Renal malrotation is a rare congenital variation of kidneys and hilum position, more common in males, with a prevalence of 1 in 2000 autopsies [3]. Usually renal malrotation occurs unilaterally and many patients might show no symptoms at all during life.

Referring to the renal hilum and position, malrotation may be:

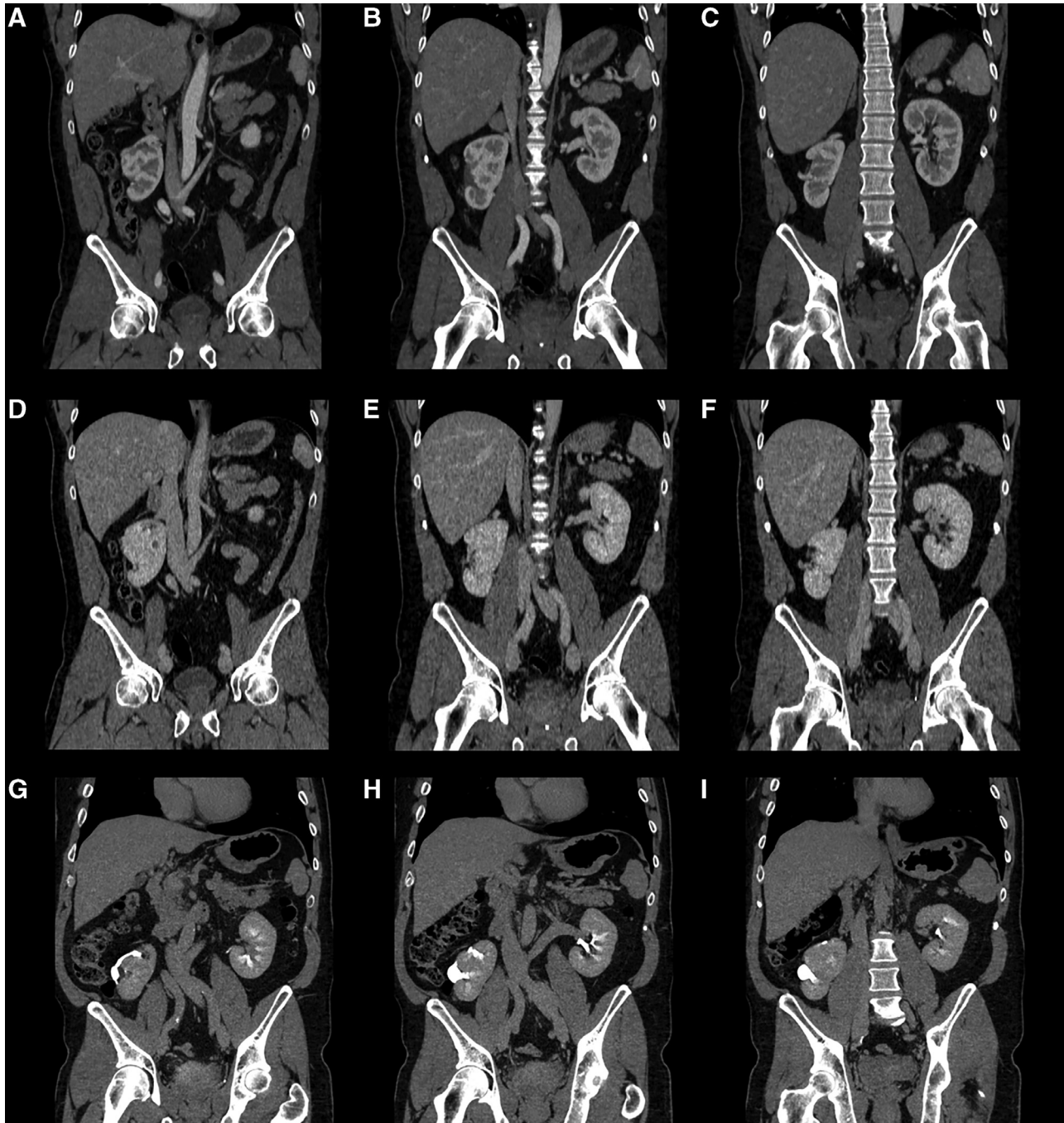


Fig. 2 – Dynamic contrast-enhanced CT study on coronal plane, acquisition at 35 (A-C), 90 (D-F), and 300 (G-I) seconds, showing ectopic, smaller, dysmorphic right kidney with reverse rotation and pelvis facing laterally. The lateralized calyces are distorted without visible obstruction.

- incomplete rotation or nonrotation (most common type): anteriorly faced hilum;
- excessive rotation or hyper-rotation: posteriorly faced hilum;
- reversed rotation: laterally faced hilum.

During the embryogenic period, kidney develops from metanephros located in the sacral region and the hilum is directed anteriorly. Both kidneys then gradually ascend to the

lumbar region while the hila rotate medially [4]. During their ascent, kidneys are initially vascularized by branches of common iliac arteries and later by the abdominal aorta.

At the ninth week of intrauterine life, renal ascent stops and kidneys receive arterial blood from renal arteries only. Other mesonephric arteries, besides renal arteries, include the middle supra-renal, gonadal, and inferior phrenic arteries. The particular development and continued changes of blood supply during the renal ascent may explain the high incidence

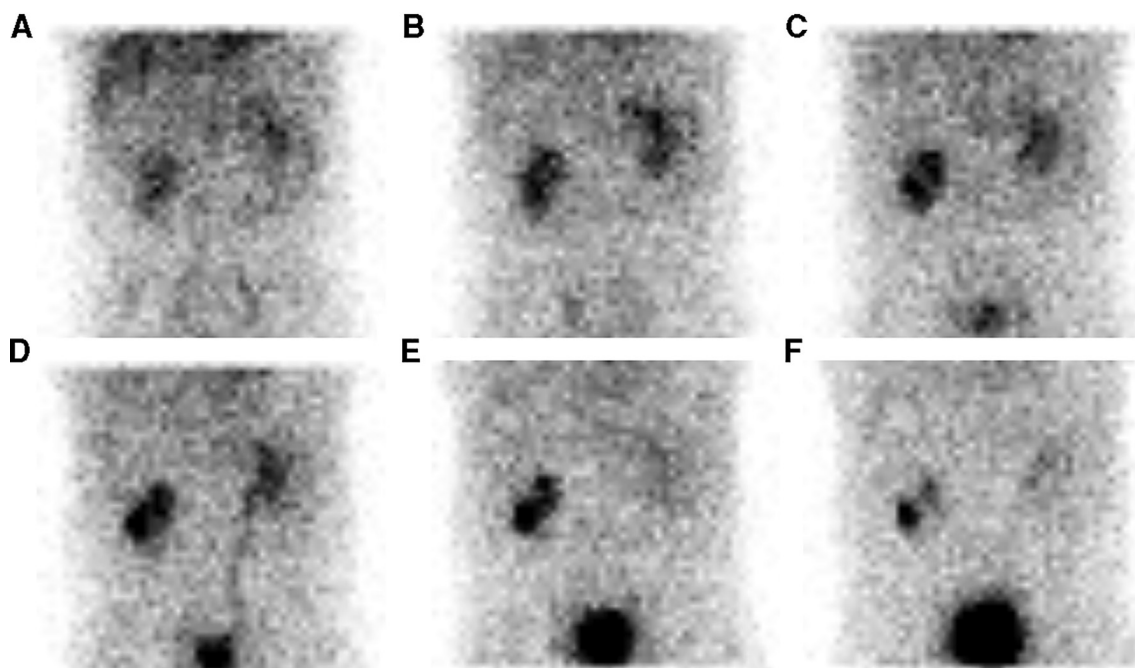


Fig. 3 – Dimercaptosuccinic acid (DMSA) scintigraphy shows stasis of radioactive urine in lower right calyceal group with normal concentration and excretory phase in the left kidney.

of the variations in terms of number, shape, size, position, rotation, and vascularization.

The frequency of multiple renal arteries ranges from 9% to 76% with an incidence rate of 28%-30%, depending on gender and race [5,6]; in our case, the patient has 4 renal arteries and the recurrence of this variation is of about 0.4% [7].

Frequently, ectopic rotated kidney with vascular aberrations predisposes patients to renal or ureteral obstruction, with consequent stone formation, infections, and abdominal or colicky pain; other common symptoms are hypertension and renal disorders, or even renal failure [8]. Urine in some cases may flow backward from the bladder to the kidney, leading to vesico-ureteral reflux and kidney scarring. Kidney malrotation may also increase the risk of hydronephrosis: in our patient case, the right kidney was ectopic and his renal pelvis was facing laterally, resulting in a slower urine excretion and in a stasis in the lower calyceal group.

However, these renal anomalies could remain asymptomatic for a long time and be accidentally discovered during imaging examination.

Treatment options may depend upon the presence of symptoms or complications. At the same time imaging plays a pivotal role in early diagnosis and proper treatment. No therapy is required if urinary function is normal, with no obstruction of the urine flow [9]. If an obstruction is present, surgery may be required in order to rectify kidney location and to allow better drainage of urine. If vesico-ureteral reflux is present, ureter course can be corrected by surgery. In case of extensive renal damage, nephrectomy may be indicated.

Ectopic reversed rotated kidneys due to malrotation and vascular anomalies increase the chance of developing long-term complications that can lead to hematuria and/or hydronephrosis, with colicky pain.

The knowledge of these anomalies and their correct description in radiological reports (especially kidney location, number and origin of arterial supplies, and ureteral malformations) may prove very useful to urologists and radiologists in making accurate diagnosis and choosing the right medical treatment [10].

As matter of a fact, clinical, biochemical, and radiological follow-up are of the utmost importance for urologists, interventional radiologists, nephrologists and surgeon to exclude complications like calculi and hydronephrosis.

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