CASE REPORT

Crossed fused renal ectopia: case report and review of the literature

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Abstract Crossed fused renal ectopia is a rare congenital anomaly, and is mostly detected incidentally. A 45-year-old man, during investigation for recurrent abdominal pain, was found to have an empty left renal fossa and right crossed renal ectopia with fusion on ultrasonography. In the present case, there were no abnormalities and/or alteration of the renal function. Abdominal tomography scan with contrast medium confirmed the diagnosis. Ectopic kidney is often associated with other abnormal situations such as agenesis, vascular malformation, incontinence, a palpable abdominal mass, urinary tract infection, high incidence of stone formation, and genital anomalies. Through this case report, we emphasize the importance of adequate imaging.

Keywords Congenital kidney anomalies · Ectopic kidney · Ultrasound

Sommario L'ectopia renale crociata con reni fusi è una malformazione congenita molto rara. Asintomatica con riscontro casuale. Riportiamo la storia clinica di un uomo di 45 anni giunto al nostro ambulatorio per dolori addominali ricorrenti, senza alcuna alterazione della funzionalità renale. L'ecografia addominale B-mode ha evidenziato a sinistra la loggia renale disabitata dal rene e occupata da anse intestinali; a destra, a livello

dell'ipocondrio, la presenza di due reni situati in sequenza longitudinale. Il completamento diagnostico è stato effettuato con una TAC addome con mezzo di contrasto che ha confermato la presenza dell'ectopia renale con reni fusi tra loro. L'imaging ultrasonografico risulta sia una buona indagine di screening per lo studio morfologico e vascolare delle malformazioni renali sia nel follow-up, considerando le possibili complicanze (infezioni, pielonefriti, massa palpabile, reflusso vesicoureterale, ostruzione, idronefrosi ed urolitiasi) associate alla patologia malformativa.

Introduction

Congenital anomalies of the kidney are rare diseases, which constitute approximately 10–12 percent of all anomalies identified in adulthood and comprise a wide range of pathologies that include abnormal number (agenesis, supernumerary kidney), positional anomalies (malrotation, ectopia), shape and fusion (horseshoe kidney, cross-fused ectopia), and urinary tract anomalies [1].

Ectopic kidney describes a kidney placed outside the normal renal fossa. The simple ectopia presents a kidney ectopic on its own side; instead, crossed ectopia, fused or unfused, presents when the kidney is located across the midline with its contralateral kidney.

Case report

A 45-year-old man came to the Nephrology Department complaining recurrent abdominal pain, for about the past 6 months, and feeling of heaviness in the right iliac fossa, usually in the morning. No family history of congenital

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anomalies was reported. Physical exam was within normal limits. There was no bacterial growth on urine culture. Routine laboratory examination showed no abnormal values.

Abdominal ultrasound B-mode supine, prone, and side, with front longitudinal scans, coronal, transverse, and oblique subcostal was performed.

Ultrasound showed the left renal loggia uninhabited by the kidney and occupied by intestinal loops (Fig. 1); to the right, in hypochondrium area, with the presence of two kidneys located in the longitudinal sequence. There was clearly evident an indentation (or incision on an edge) at the fusion point between the two kidneys (Fig. 2). The bipolar diameters of the two kidneys were standard (one 105 mm, the other 110 mm albeit with an indicative measurement due to melting bridge): both kidneys showed normal parenchymal thickness (19–19.5 mm), no hydronephrosis, the absence of stones, and normal bladder.

ColorDoppler sampling at the level of vesical trigone showed the jet phenomenon, evidence of the arrival of urine in the bladder bilaterally.

In addition, ultrasound examination can give some information about vascularization; in our patient, it was normal; in fact, there were evident two renal arteries, and there were no abnormal venous vessels.

Indeed ColorDoppler ultrasound imaging was carried out for the samplings of the two renal arteries: for one kidney arranged above and the other arranged below the kidney, both having intraparenchymal resistance indices in the interlobar arteries of both kidneys with standard flow (Fig. 3).

Abdominal tomography scan with contrast medium confirmed the presence of renal ectopia with kidneys having been fused with each other, both being located on the right without other associated anomalies. In particular, the lower pole of the first kidney located above gets fused with the upper pole of the second kidney located below, in

the absence of vascular abnormalities and renal pelvis of both kidneys with anterior malrotation.

The ureter of the upper kidney, in the first section of the route, delimits the lateral margin of the lower kidney; in the intermediate portion, the two ureters (right and left) shall run in parallel in the right paraspinal area, and at the level of the 5 lumbar vertebra, the ureter of the lower kidney exceeds the midline, then passes down along the side of the pelvis, and finally arrives at the back of the bladder.

Indeed the ureters terminate in the bladder on the right and left, respectively (Fig. 4).

Concentration and excretion of contrast medium radio-opaque are preserved in fused kidneys.

Therefore, abdominal ultrasound performed after some symptoms that lasted for several months has been very useful for diagnosis of crossed kidney ectopic with fusion, without associated abnormalities.

Discussion

Renal ectopy and fusion are common congenital anomalies of the kidney and urinary tract (CACUT). Crossed fused kidney ectopia is a very rare congenital malformation. The exact incidence of crossed fused ectopia is unknown, because most of the patients are asymptomatic; it occurs in about 1 in 1000 live births. After horseshoe kidney, crossed fused ectopia is the most frequent fusion abnormality of the urinary tract in which one kidney migrates abnormally to the opposite side and fuses with the other kidney during development. This abnormality arises as a consequence of abnormal renal ascent in embryogenesis with fusion of the kidneys within the pelvis. It is thought to occur in the first trimester, at around the 4th–8th week of fetal life (In a normal situation, the kidney reaches its appropriate position at L2 level at the end of the 2nd month). An abnormal development of the ureteric bud and metanephric blastema between 4 and 8 weeks' gestation with the increased prevalence in scoliotic patients.

It has a 3:2 male-to-female predominance.

Left to right is more common than right to left (3:1).

In this abnormality, as in our patient, both the kidneys are located on the same side, with the most cases in fused condition, with two separate ureters arising from the respective kidneys. The ureter arising from the crossed-over kidney travels back to the opposite side and inserts in the bladder. Crossed fused kidney ectopia was detected in patients during physical or radiological investigation for other genitourinary issues.

Imaging continues to occupy a key role in the congenital kidney diseases for facilitating a careful diagnosis. Especially, ultrasound is often used for its detection and

Fig. 1 Longitudinal section: left renal loggia uninhabited by the kidney and occupied by intestinal loops

Fig. 2 Longitudinal section. **a** Overview reconstruction: the presence of two kidneys in the longitudinal sequence on the right side, normal parenchymal thickness. **b** and **d** both the kidneys are located on the right side and show fusion at their poles; the lower pole of the first

kidney located above is fused with the upper pole of the second kidney located below. **c** Complete parenchymal hypoechoic bridge indicating, probably, the “notch” between the two kidneys

Fig. 3 **a** Jet phenomenon **b** B-mode and color: aorta, renal artery, and msa (mesenteric superior artery). **c** and **d** ColorDoppler ultrasound imaging was carried out for the samplings of both renal arteries

evaluation in order to take advantage of its rapid scanning time, lack of radiation, low cost, and easy feasibility, and Doppler ultrasound helps in accurate diagnosis through the study of vascularization. This is a great help, because

ectopic kidneys usually contain numerous small vessels and reflect the constant changes in blood supply during the developing stage, until the kidneys reach their final position.

Fig. 4 Abdominal tomography scan with contrast medium: the presence of renal ectopia with kidneys fused with each other, both located on the right with anterior malrotation and normally enhancing parenchyma

Also, tomography scan and magnetic resonance imaging provide additional information.

Often, this anomaly presents some complications like hydronephrosis and recurrent urinary tract infections, abdominal mass, and a high incidence of stone formation. There are also reported cases of renal cell carcinoma and Wilm's tumor associated with crossed fused kidney ectopia [2, 3].

In ultrasound, there may be observed a characteristic “notch” between the two fused kidneys [4].

The literature has reported a few cases of renal ectopia with fused kidneys, and the clinical profile has been studied, and the management of this entity sometimes associated with other abnormalities of the genitourinary tract, such as hydronephrosis, vesicoureteral reflux, gallstones, dysplasia renal, and hypospadias [5–7]. Diagnosis is made in the course of investigations for abdominal pain, abdominal mass, anorectal malformations, vesicoureteral reflux, and urinary tract infection. In rare cases, this fault is signaled during prenatal ultrasound or during surgical exploration [8, 9].

Riaz Ahmad also described a rare association, never reported in the literature, of ectopia with fused kidneys in a young woman with thrombocytopenia and bilateral radial aplasia, radially deviated hands, the presence of thumbs (TAR syndrome). The ultrasound, performed for recurrent urinary tract infections, has not revealed the kidney in the right renal loggia, and the diagnosis was completed with magnetic resonance imaging and renal scintigraphy [10].

Conclusion

This paper highlights the role of imaging in a rare case of cross-fused renal ectopia with normal function and without specific symptoms except for a recurring abdominal pain in the right side.

Ultrasound imaging and echocolor Doppler are very useful with regard to morphological and vascular study of the kidney malformations and for the follow-up, considering the possible complications associated therewith (infections, pyelonephritis, palpable mass, vesicoureteral reflux, obstruction, hydronephrosis, and urolithiasis).

There are no specific guidelines available for the management of crossed fused kidney ectopia; crossed fused ectopia usually does not require any primary treatment, and the kidney units do not need to be divided. The treatment is related to the presence of symptoms and/or complications [11, 12].

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all individual participants included in the study.

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