Left crossed fused renal ectopia L-shaped kidney type, with double nutcracker syndrome (anterior and posterior)

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Abstract
CROSSED FUSED RENAL ECtopIA (CFRE) IS THE SECOND MOST COMMON FUSION ANOMALIES (FAs) OF THE KIDNEYS AFTER HORSESHOE KIDNEY. CFRE RESULTS FROM ONE KIDNEY CROSSING OVER TO THE OPPOSITE SIDE AND SUBSEQUENT FUSION OF THE PARENCHYMA OF THE TWO KIDNEYS. WE REPORT, BY MULTIDETECTOR-ROW COMPUTED TOMOGRAPHY (MDCT) ANGIOGRAPHY, AN EXTREMELY RARE CASE OF A LEFT CFRE (L-SHAPED KIDNEY TYPE), CONSISTING OF MULTIPLE RENAL ARTERIES (ONE MAIN RENAL ARTERY FOR THE UPPER RENAL PARENCHYMA, AND THREE RENAL ARTERIES (ONE MAIN AND TWO ADDITIONAL) FOR THE LOWER RENAL PARENCHYMA) AND TWO LEFT RENAL VEINS, WHICH PRODUCED A DOUBLE NUTCRACKER SYNDROME (BOTH ANTERIOR AND POSTERIOR). THE L-SHAPED LEFT KIDNEY HAS A MAXIMUM LENGTH OF 18.5 CM, A MAXIMUM WIDTH OF 10.2 CM, AND A MAXIMUM THICKNESS OF 5.3 CM. THE UPPER POLE OF THE KIDNEY IS LOCATED AT THE LEVEL OF THE LOWER THIRD OF T12 VERTEbral BODY (4.6 CM LEFT TO THE MEDIOSAGITAL PLAn); THE LOWER POLE IS LOCATED ALONG THE LOWER HALF OF THE L5 VERTEbral BODY (1.5 CM LEFT TO THE MEDIOSAGITAL PLAn). THE FOLLOWING CASE WILL FOCUS ON THE RELEVANT ANATOMY, EMBRYOLOGY, AND THE CLINICAL SIGNIFICANCE OF THIS ENTITY.

Keywords: crossed fused renal ectopia, left L-shaped kidney, additional renal arteries, left renal veins, anterior nutcracker syndrome, posterior nutcracker syndrome.

Introduction

We report, by multidetector-row computed tomography (MDCT) angiography, an extremely rare case of a left CFRE (L-shaped kidney type), consisting of multiple renal arteries and two left renal veins, which produced a double nutcracker syndrome (both anterior and posterior). The following case will focus on the relevant anatomy, embryology, and the clinical significance of this entity.

Patient, Methods and Results
A 26-YEAR-OLD MALE PRESENTED WITH A SIX-MONTH HISTORY OF INTERMITTENT ABDOMINAL PAIN IN THE LEFT FLANK REGION. Medical workup, including urinalysis, was all within normal limits. A plain abdominal radiograph showed an enlarged left kidney shadow. The right kidney could not be visualized. An abdominal ultrasound examination demonstrated the absence of a right kidney and the presence of two kidneys in the left renal fossa, joined by parenchymal tissue. The patient was referred to Neuromed Diagnostic Imaging Center (Timișoara, Romania) for further diagnostic imaging. Using MDCT angiography (64-slice MDCT system; SOMATOM Sensation, Siemens Medical Solutions, Forchheim, Germany), we identified
The aortomesenteric angle is 19° (Figures 2 and 3).

In the aortomesenteric angle, at this level measures 17.5×13.4×9.5 mm. At preaortic level, RV1 is compressed between the inferior vena cava, at the level of T12–L1 intervertebral disc. The ostium of RV1 has an oval shape, edge of the inferior vena cava, at the level of the T12 vertebral body. The diameter of RA1 is 6 mm at the origin and length of 21.5 mm before branching. RA4 originates along the right edge of the AA at the level of intervertebral disc L2–L3. It has a diameter of 4 mm at the origin and extends 40.2 mm before branching. RA3 originates along the right edge of the AA at the level of L2–L3 intervertebral disc. The measurements of R3 include a diameter of 4.6 mm at the origin and length of 21.5 mm before branching. RA4 originates along the right edge of the AA at the level of middle third of L3 vertebral body.

Examination of the osteoarticular system demonstrated hypoplasia of the 12th ribs bilaterally (Figure 1).

The upper renal parenchyma (identified by a longitudinal–vertical axis) was located in the normal position, is vascularized by a single renal artery (RA1) with origin at the left edge of the AA at the level of middle third of L1 vertebral body. The diameter of RA1 is 6 mm at the origin and has a length of 35.3 mm to the branching level. Venous drainage of the renal parenchyma is through a single renal vein (RV1) with preaortic trajectory. This drains into the left edge of the inferior vena cava, at the level of the T12–L1 intervertebral disc. The ostium of RV1 has an oval shape, with 13.4×9.5 mm. At preaortic level, RV1 is compressed in the aortomesenteric angle; at this level measures 17.5×4.6 mm. The aortomesenteric angle is 19° (Figures 2 and 3).

The pyelocaliceal system is arranged vertically in the renal sinus and has three major renal calyces. The left ureter descends vertically, with a slight curve to the right. It lies anterior to the sacral foramina and ends in the urinary bladder wall in the normal anatomic location (Figures 1 and 4).

Lower renal parenchyma (identified by a horizontal longitudinal axis) is ectopically located in an inferior lumbar position. It is vascularized by three renal arteries: one main renal artery (RA2) and two additional renal arteries (RA3 and RA4). RA2 has an origin at the left edge of the AA at the level of intervertebral disc L2–L3, it has a diameter of 4 mm at the origin and extends 40.2 mm before branching. RA3 originates along the right edge of the AA at the level of L2–L3 intervertebral disc. The measurements of R3 include a diameter of 4.6 mm at the origin and length of 21.5 mm before branching. RA4 originates along the right edge of the AA at the level of middle third of L3 vertebral body.
It has a diameter of 3.4 mm at the origin and measures 44.5 mm before penetrating the renal parenchyma. The venous drainage of the renal parenchyma consists of a single renal vein (RV2), which lies posterior to the AA and drains into the left edge of the inferior vena cava (IVC), at the level of the upper third of L2 vertebral body. The ostium of RV2 has a cross-sectional area measuring 10.2×10.0 mm. RV2 becomes compressed between the AA and the L2 vertebral body; at this level, the compressed venous lumen has a cross-sectional area measuring 8×4.1 mm (Figures 2 and 3).

The pyelocaliceal system is oriented horizontally within the renal sinus and has three major renal calyces. The right ureter crossed the mediolateral plane at the level of promontorium, and finally penetrates in the bladder wall in normal position (Figures 1 and 4).

| Figure 4 – Multiplanar reformatting (MPR) (A and C) and maximum intensity projection (MIP) (B and D) images in coronal (A and B) and sagittal (C and D) planes of the left ectopic crossed fused kidney demonstrates the anterior orientation of the renal pelves, and the passage of the both ureters in contact with the anterior surfaces of the upper and lower renal parenchyma. |

Discussion

Renourinary system anomalies include renal agenesis, multiple kidneys, renal ectopia, and fusion defects [10]. According to Khan et al. [11], urinary tract system anomalies account for 3.0% of all congenital anomalies. CFRE, described for the first time by Pamarolus in 1654 (quoted by [12]), is the second most common fusion abnormality of the kidney [13]. In the general population, CFRE occurs more frequent in men than women, from 3:2 [10] to 2.3:1 [14]. The crossover of the left kidney to the right side is the most common form for CFRE [10]. In our case, the patient was a male with a crossover from right to left.

Classification

In 1927, Papin and Eisendrath [15] performed an extensive classification of renal and ureteral abnormalities and grouped them into 10 renal anomalies and four ureteral anomalies. The ten types of renal anomalies include: (1) anomalies of number, (2) anomalies of volume, (3) anomalies of form, (4) anomalies of location, (5) median fusion, (6) anomalies of rotation, (7) reduplication of the pelves and ureters, (8) anomalies of the pelvis (other than reduplication), (9) anomalies of the vessels, and (10) non-classifiable anomalies. For anomalies of location (4), the authors describes three entities: (a) simple or ordinary unilateral ectopia, (b) simple or ordinary bilateral ectopia, and (c) crossed ectopia with or without fusion. The median fusion type (5) includes four entities: (a) horseshoe kidney, (b) L-shaped kidney, (c) cake kidney, and (d) sigmoid kidney. McDonald and McClellan [16] classified the crossed ectopic kidney into four types: crossed renal ectopia with fusion (CFRE), crossed renal ectopia without fusion, solitary crossed renal ectopia, and bilaterally crossed renal ectopia. Subsequent studies [2, 3] led to the establishment of a CFRE classification of six types (according to the degree of fusion, location or rotation of the fused renal mass). These consist of (in decreasing order of frequency): (1) inferior crossed fused ectopia, (2) sigmoid or S-shaped kidney, (3) unilateral lump kidney, (4) unilateral disc kidney, (5) L-shaped kidney, and (6) superior crossed fused ectopia.

Development

The definitive kidney (the metanephros) originates during the fourth and fifth weeks of intrauterine life, at the level of the first or second sacral segment [14]. At this level, the ureteral bud (originating from the mesonephric duct) extends toward the metanephric blastema. These entities join to form the definitive kidney [14]. From the pelvic cavity, the two masses of metanephrogenic tissue ascend from the pelvis between 6th and 9th week [14]. Initially, the kidneys lie close to each other with the hila oriented anteriorly. To reach the definitive position in the lumbar region involves a well-orchestrated series of movements, including ascent, lateral migration, axial deflection, and medial rotation (almost 90°) [2, 11, 14]. According to Khan et al. [11], the ascent of the kidney is caused by the diminution of body curvature and by the growth of the body in the lumbar and sacral regions. In the 9th week of development, the kidneys attain the adult position, adjacent to the adrenal gland [14].

The pathophysiological mechanisms of CFRE are not fully understood [7]. At the beginning of the cranial migration of the ureteral buds, the nephrogenic blastemas are squeezed together between the umbilical arteries, which may ultimately cause their fusion. If the metanephric masses are at the same level, the result is a cake or horseshoe kidney [17]. Complete fusion gives rise to a cake kidney, while fusion localized to one renal pole (most commonly in the lower pole) causes horseshoe kidney. The ascension of the cake and horseshoe kidneys are limited to the origin of the inferior mesenteric artery from the abdominal aorta (AA). It is postulated that if the compression factor of the umbilical arteries persist at the beginning of the cranial migration in the presence of two unequal metanephric masses, the result will be crossed ectopia [17]. Ashley and Mostofi [18] suggested that the persistence of caecum in the right lumbar region...
(before reaching its final position in the right iliac fossa, last 180° rotation of midgut around the axis of superior mesenteric artery) may favor the migration of the right renal unit to the left.

Cook and Stephens [19] suggested an alternative pathogenetic explanation for the development of a CFRE in which the growth of the ureteric buds (arising from both wolffian ducts) occur in a single nephrogenic cord. According with Rina et al. [5], this process may result in the inability of the ureteric bud to communicate with the more distant ipsilateral metanephric blastema. The nephrogenic tissue from the side that does not receive a ureteric bud will completely regress [20].

During its normal migration, the metanephros acquires its vascular supply from pelvic branches of the iliac arteries and from branches of the dorsal aorta [21]. According to Felix, in an embryo of 18 mm (with complete degeneration of mesonephros), a number of nine lateral mesonephric arteries were counted from the 10th thoracic to the 5th lumbar segment [22]. These nine pairs (located in the angle between the mesonephros and the metanephros), have been divided into the cranial (the 1st and the 2nd pair), middle (the 3rd to the 5th pair) and the caudal (the 6th to the 9th pair) groups. The renal arteries develop from a single pair, from the middle group [6], at the level of L2 vertebral body. During the transition phase from mesonephros to metanephros, if more than one mesonephric arteries from the middle group persists, it results the additional renal arteries. The additional renal arteries must be considered persistent mesonephric arteries [22]. In rare situation in case of complete ascent of the kidneys, some additional (superior or inferior) renal arteries may arise through persistent extreme groups (cranial and caudal) of the mesonephric arteries.

In the developing kidney, the vascular supply is reestablished progressively as it migrates superiorly. If migration is arrested, the temporary blood supply will become permanent [9]. According to Türkvat et al. [6] and Khan et al. [11] the blood supply to the ectopic kidney most frequently arises from the vessels on the ipsilateral side, but may occasionally arise from the contralateral side. In the CFRE, the superior kidney is usually supplied by only one renal artery (branch of the AA), while the remaining kidney gains its blood supply from the middle group of the mesonephric arteries. Türkvat et al. [6] demonstrated that the total number of arteries ranges from one to six, most commonly there are two to four major arteries to the two kidneys that form the CFRE.

In our case, the superior kidney found in the normal position, is served by a single artery (the main renal artery) originating in the AA at the level of middle third of L1 vertebral body. The inferior kidney is served by three renal arteries, one main renal artery originating from the left edge of AA at the level of intervertebral disc L2–L3 vertebral body, and by two additional renal arteries originating from the right edge of the AA at the level of L3 vertebral body.

According to Nam et al. [23], the development of the renal veins is a part of the complex developmental process of the IVC. Between the 4th and 8th week of development, the posterior cardinal veins, the subcardinal veins, and the supracardinal veins develop a vast network that build the IVC. During the development process of the IVC the anastomotic communications between the subcardinal and supracardinal channels form a venous collar that encircling the AA. The ventral part of this collar generally persists as the normal left renal vein (preaortic left renal vein). If the dorsal part of the collar persists, then the left renal vein is located posterior to the AA (retroaortic left renal vein) [24]. If both the dorsal and the ventral portions persist, there will be a circumaortic venous collar in the adult [24, 25]. Venous drainage of the crossed ectopic kidney is usually into the distal IVC or the common iliac veins. In our case, both renal veins drain into the IVC (at the level of middle third of L1 vertebral body and upper third of the L2 vertebral body, respectively). The vein draining the superior kidney passes anterior to the AA (in the aortomesenteric angle), while the vein draining the lower (ectopic) kidney passes posterior to the AA.

In the early stage of development, while the kidneys are located in the pelvic cavity, the hilum and the renal pelvices are oriented anteriorly. After the complicated series of movements during ascent, the renal pelvices will come to rest medially [2, 11, 14]. In case of CFRE, the renal pelvis of the superior kidney is usually vertical with an anterior or anterior-medial orientation [16]. The inferior kidney (and the renal pelvis) position and orientation are variable, depending on the morphological type of CFRE [2, 6, 9]. In our case, both renal pelvices have an anterior orientation. The superior renal pelvis is placed vertically while lower renal pelvis has a pronounced skew.

**Clinical implication**

Most cases of CFRE are asymptomatic during life and are diagnosed incidentally [2, 10, 13]; the patients renal function is usually normal [4]. When symptoms do occur, however, the most common symptoms reported are flank and lower abdominal pain, a palpable abdominal mass, hematuria, dysuria, urinary tract infections, renal failure, fever and hypertension [3, 10]. Pathologies associated with this anomaly include: nephrolithiasis, ureteropelvic junction obstruction, hydronephrosis, urinary stasis, reflux, ectopic ureteroceles, calculus formation, hydroureter, and renal or urothelial tumors [3, 4, 10]. Congenital anomalies, especially vertebral and skeletal anomalies of the bony pelvis, are commonly associated with CFRE [2] and may include cardiovascular anomalies [3, 10], annular pancreas, multicystic dysplasia [4], VACTER syndrome [7, 4], caudal regression, trisomy 21, and Turner syndrome [7].

Renal arterial stenosis secondary to CFRE may cause renovascular hypertension [26]. In addition, the presence of additional renal arteries may lead to prolonged operative time or surgical interventions in the presence of the CFRE.

The left renal vein (LRV) lies either anterior or posterior to the AA. The compression of the LRV between the superior mesenteric artery and the AA produce the anterior nutcracker syndrome [23], while compression of the LRV in the decreased space between the AA and the vertebral bodies may cause a posterior nutcracker syndrome [23]. Rudloff et al. [27] demonstrated
that the compression of the LRV leads to hematuria. This is thought to be secondary to the increased pressure in the LRV, which results in congestion of the left kidney and the venous communications.

Conclusions

CFRE is the second most common FAs of the kidneys after HK. CFRE results from one kidney crossing over to the opposite side and subsequent fusion of the parenchyma of the two kidneys. Usually, CFRE is diagnosed as an incidentally, when patient is examined for other reasons, unless it is complicated by infection or obstruction. MDCT examination can provide good delineation of vascular and urinary tract anomalies. In CFRE, the arterial supply is usually normal, the venous communications.

References


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