Unascended Left Kidney with Malrotation: A Rare Congenital Anomaly

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Abstract: ***Anomalies of kidney form a significant portion of congenital malformations. Anomalies may occur in the number, position, shape, size and rotation of kidney. Structural, positional and vascular anomalies are the most frequently reported. Rotational anomalies form a rare entity not cited in most of the embryology textbooks. We present a case of unilateral unascended left kidney with malrotation in a male cadaver which was associated with other visceral and vascular variations. An attempt has been made to systematically document these variations and give possible explanations for the same. A detailed knowledge of these morphological variations will be useful for many urological and radiological procedures.***

Keywords: congenital malformations, kidney, rotational malformations, vascular anomalies

1. Introduction

Ectopic kidney is a birth defect in which a kidney is located at an abnormal position. The frequency of ectopic kidney is 1:500 to 1:1100, ectopic thoracic kidney is 1:13000, solitary kidney is 1:1000, single pelvic kidney is 1: 22000, one normal and one pelvic kidney is 1: 3000. (Bergman – EKCP) Ectopic kidneys are most commonly found in the pelvis. Other locations can be iliac region, abdomen, and chest and in some cases contra lateral referred to as "crossed renal ectopia" [1].

Each of the kidneys is situated retroperitoneal in the posterior abdominal wall by the side of the vertebral column and extends from T12 to L3 vertebrae. Right kidney is slightly lower than its left partner due to the presence of liver [2]. Kidney develops between 4th to 12th weeks of intrauterine life. Developmentally at first the metanephric kidney lies in the pelvic cavity opposite the sacral segments. Gradually it ascends and reaches the iliac fossa after crossing the pelvic brim. Finally, it appears on the undersurface of the diaphragm where its further ascent is arrested by the supra renal gland[2]. By 7th week, the hilum points medially and kidneys are located in the abdomen. The thoracic ectopic kidney with partial or complete renal protrusion above the level of the diaphragm into the posterior mediastinum is the rarest form of all ectopic kidneys [3].

During the process of ascent from the pelvis, the kidney derives their blood supply sequentially from vessels that are closest to them- initially median sacral, then common iliac and inferior mesenteric, and finally from the aorta. Simultaneously, there is degeneration of primitive lower vessels. Any failure of degeneration of these primitive lower vessels in ectopic kidney which is caudal in position results in origin of more than one accessory and polar renal artery [4]. During ascent, the hilum of the kidney is directed ventrally. When it reaches the permanent position, kidney undergoes 90 º medial rotations around the vertical axis and the hilum turns medially [2].

Anomalies of structure and position of kidney are commonly reported. Rotational anomalies form a rare entity. Though rare there are wider implications of it in advanced surgical procedures and diagnostic evaluation of kidney donors.

2. Case Findings

During routine, dissection in the department of Anatomy, KIMS Bangalore we observed an unascended left kidney with mal rotation with vascular variations. Location, position and dimensions of the left kidney were analyzed and compared with the right. Later, number of arteries supplying kidney, their origin, length, diameter and distance of these arteries from the aortic bifurcation were noted.

2.1 Findings on Right Kidney

Kidney was normal in position between T12 and L1. It was bean shaped but had compensatory hypertrophy measuring about 15 cm vertically, 10 cm transversely and 3.8cm antero posteriorly. There was a single right renal artery arising from abdominal aorta. Right gonadal vessels were normal.

2.2 Findings on Left Kidney

Left kidney was situated anterior to the bodies of L5, S1, S2, S3 vertebrae. It was oval in shape measuring 10cm vertically, 8cm transversely and 4 cm anterio posteriorly with both ventral and dorsal surfaces smooth. It was almost transversely oriented, upper pole being in the midline and related to left common iliac and internal iliac vessels superiorly. Lower pole,, situated in the true pelvis. Sigmoid colon and mesocolon were displaced to the right side and was related to the right border of the ectopic kidney. Left margin was related to left common iliac and left internal iliac vessels. Hilum faced laterally instead of medial, showing 90º degree malrotation. The left ureter crossed over the ventral surface of the left kidney to open into the urinary bladder. There were five renal arteries supplying left kidney. [Fig 1, 2, 3]. The source of their origin, their distance from the aortic bifurcation, length and diameter of each one it is mentioned in the table 1.

Left renal arteries	*Distance of the artery from aortic bifurcation in cms*	*Origin of the arteries*	*Length in cms*	*Diameter in cms*
LRA 1	0.4	Left common iliac artery	5.9	0.5
LRA2	3.6	Left common iliac artery	5.1	0.5
LRA3	7.1	Left internal iliac	0.8	0.2
LRA4	11.3	Posterior division of left internal iliac artery	2.2	0.4
LRA5	-	Lateral sacral artery	0.5	0.1

Table 1: LRA1 – Left Renal Artery 1, LRA2 – Left Renal Artery 2, LRA 3 – Left Renal Artery 3, LRA4 – Left Renal Artery 4, LRA5- Left Renal Artery 5

3. Discussion

Unilateral ectopic kidney is common. Congenital pelvic kidney is commoner on right than on the left side. Muhammad Asghar and Fidaullah Wazir in their study for the prevalence of renal ectopia by diagnostic imaging found renal ectopia in 25% (0.2 %) patients out of total 12,000 patients with abdominal complaints. Out of 25 renal ectopia, 7 were on right side, 5 were left ectopic pelvic kidneys. Bilateral ectopic pelvic kidneys were found in only 2 cases [5].

Belsare, et al reported a similar case of ectopic pelvic kidney on the left side but in a female cadaver with enlarged uterus, displaced left ovary, and multiple renal vessels on both the sides, variations in the aorta, inferior venacava and in the gonadal vessels [6].

Banner concluded that in case of females, the pelvic kidney may result in obstetric complications. Thus, in the present case most of the findings are in accordance with the findings of Belsare et al expect that features of ectopic kidney with malrotation with vascular variations were found in a male cadaver [6].

Sometimes excessive cranial ascent of kidney prior to diaphragmatic closer or delayed closure and maldevelopment of pleuroperitoneal membrane can lead to a ectopic thoracic kidney [3].Campbell found 22 cases of ectopic kidney in 15,919 autopsies of children among which only one was intrathoracic kidney [7].

Anomalies of rotation may be seen in normal as well as renal ectopia [8]. Four types of rotational anomalies have been identified. In non- rotation, renal pelvis faces ventrally. In incomplete rotation, it presents ventromedially. In rare case, reverse and excessive rotation presents hilum faces laterally and prevents itself in a position depending upon the degree of rotation [8]. In the present case, it is reverse and 90^{o} malrotation, with hilum of ectopic kidney facing laterally. [Fig 1]

Renal artery vascular variations are categorized into 2 types: “early branching” and “extra branching”. In early branching main artery is proximal to the hilum but in extra branching, arteries are grouped into 2 sets, hilar (accessory) and polar (aberrant) arteries. Hilar arteries, with main renal artery enters the kidney through the hilum, polar arteries enters the kidney directly through the capsule from outside the hilum. In the present case, it was early branching type, where the branched arteries were proximal to the hilum [1] [Fig 2,3].

During ascension each kidney is vascularised by its neighboring arterial branching originating from external and internal iliac arteries. Later as the kidney ascends higher up, branches originate directly from the abdominal aorta [9]. The initial lower branches are not permanent; they regress and are replaced by newer ones with higher origin. Thus, the definitive renal arteries will originate in the upper lumbar region. There is good interrelation between kidney ascent and the level of origin of renal arteries. Thus, any anomaly in the renal vascular development or regression may delay the kidney migration, leading to ectopia [9].

Important to note that, renal rotation takes place before definitive vascularisation. Rotational anomalies are often may be due to aberrant renal vessels [8].

Most peculiar feature of the present case is the vascularisation of left ectopic kidney through five renal arteries which originated in series from, lateral sacral artery, posterior division of left internal iliac artery, left internal iliac artery, two proximal branches from the left common iliac artery. All of them differed in their length and diameter.

In this case there was an additional extra renal hilar (accessory) artery originating from the lateral sacral artery.

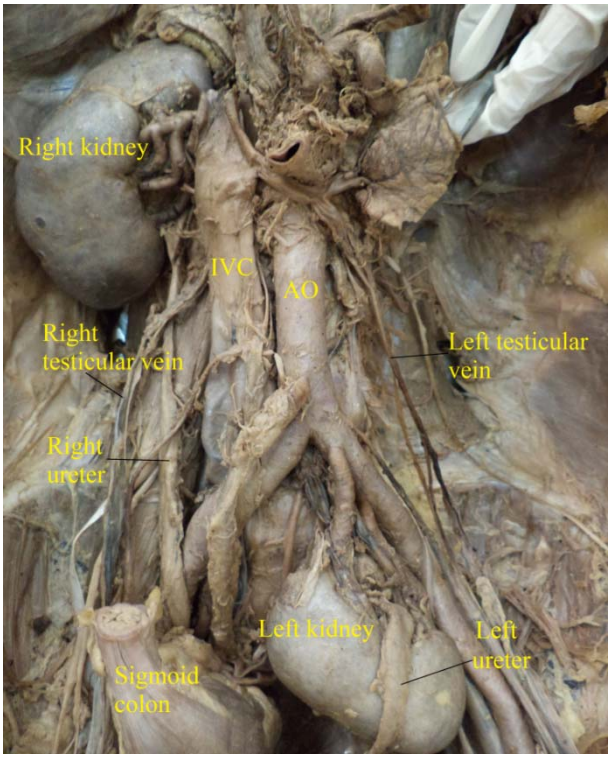

Figure 1: Photograph showing normally located hypertrophied right kidney, left ectopic pelvic kidney, sigmoid colon being pushed to the right side, left ureter curving over the ventral surface of left kidney, left testicular vein joining with the left supra renal to form a common venous channel, to finally drain into inferior venacava

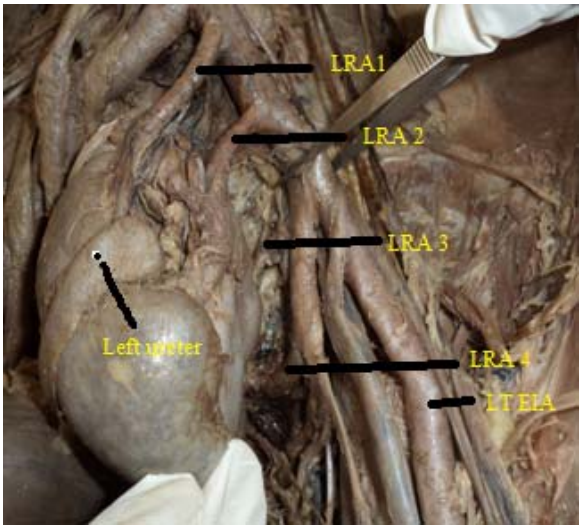

Figure 2: Four Renal Arteries supplying left ectopic pelvic kidney. LRA 1 – Left Renal Artery 1, LRA 2- Left Renal Artery 2, LRA 3 – Left Renal Artery 3, LRA 4 – Left Renal Artery 4, LT EIA- Left External Iliac Artery

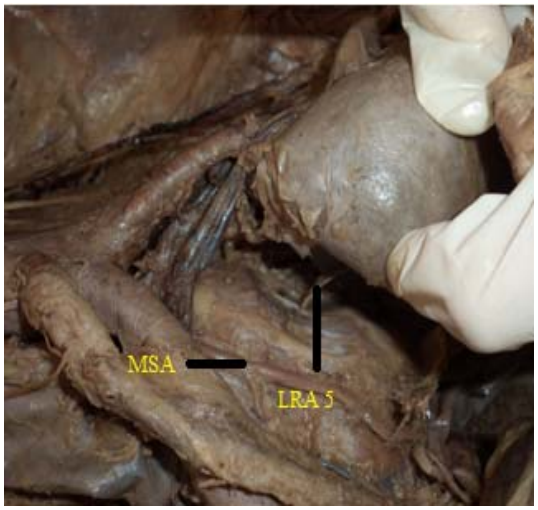

Figure 3: LRA 5 – Left Renal Artery 5 branching from Lateral Sacral Artery supplying left ectopic kidney from beneath, MSA – median Sacral Artery shown for reference

4. Conclusion

Ectopic kidneys with their atypical locations, mal rotation and vascular peculiarities have a major clinical significance. In addition to the routine contrast X rays i.e intravenous pyelogram and ascending pyelogram, the modern methods of investigations such as ultrasonography(USG), computer tomography (CT) and magnetic resonance image (MRI) scan etc are very useful to diagnose the ectopic kidney. Hence, the knowledge regarding these anatomical variations is not only important for anatomists but also for urologists in kidney transplantation, percutaneous nephrectomy and other surgical procedures. It is also important for interventional radiologists in investigatory procedures.

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