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Case Report

CROSSED FUSED LEFT RENAL ECTOPIA (CRE) IN A FETUS WITH LEFT SIDED POLYDACTYLY - A CASE REPORT

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ABSTRACT
Crossed fused renal ectopia (CRE) is an uncommon congenital anomaly wherein during development one kidney migrates to the opposite side and fuses with the kidney of that side. On opening the abdominal cavity of a 24 weeks male foetus empty left renal fossa with right sided double hila and ureter with accessory renal vessels were observed. Proper knowledge of morphological variations of kidney and vessels supplying it are essential not only for anatomists but also for urologists. A sound knowledge of these variations assist in the diagnosis of renal anomalies and in transplantation surgery.

Key Words: *Ectopia, Duplex Ureter, Polydactyly*

INTRODUCTION

Congenital anomalies of upper urinary tract include anomalies in number, location, collecting system and renal vasculature. Crossed fused renal ectopia (CRE) is one of the rarest anomalies of urinary system.In CRE both kidneys whether fused or unfused are located on one side. But the ureter of the ectopic kidney crosses the midline following the normal courseto open in its normal position in the urinary bladder.
CRE is the second most common fusion anomaly of the kidney with an incidence of 1 in 7000 at autopsy the first being horseshoe kidney1(Mouriquand, 1998).
Most of the cases of CRE are asymptomatic and are diagnosed at autopsy (Guarino *et al.*, 2004; Sood *et al.*, 2005), incidentally during radiological investigation (Hwang *et al.*, 2002) or during cadaveric dissection (Sukanya Pal *et al.*, 2008). A case of prenatally diagnosed CRE that did not required any postnatal surgical intervention (Kiddo *et al.*, 2005) and a similar case with postnatal nephroureterectomy (Narci *et al.*, 2010) were described in the literature. The aberrant renal vascular supply arises from either right or left side of aorta or from the common or external iliac arteries (Nussbaum *et al.*, 1987).

CASES

On opening the abdominal cavity of a 24 weeks male foetus with polydactyl of left hand, absence of left kidney in the renal fossa with a flattened left supra renal occupying the left renal fossa, enlarged and distorted fused kidney with two hila and uretersfacing medially in right renal fossa were observed (Fig.1).
Two ureters were originating from the fused ectopic kidney on right sideand were opening separately one on each side into the urinary bladder. First or cranial ureter (U 1) was like normal right ureter in its location, course and termination. The second ureter (U 2)was originatingnearer the lower pole of fused kidney and is anterior to the accessory renal vessels.It crossed to the left of mid line at the level of aortic bifurcation and followed the course and termination like a normal left ureter.
The arrangement of structures from anterior to posterior at cranially placed (right) hilum wasrenal vein, renal artery and ureter (VAU). The arrangement of structure from anterior to posterior at the lower pole that appeared as a caudally placed (left) hilum are ureter, accessoryrenal vein and renal artery (UVA). Both the arteries entering the hila originated form abdominal aorta on right side. There is no left renal artery.

DISCUSSION

Development of kidney starts around 4th week of gestation by interaction between ureteric bud and nephrogenic cord at the level of 2nd sacral vertebra in the pelvis. Initially the hila of kidneys face anteriorly with both kidneys being placed close to one another.

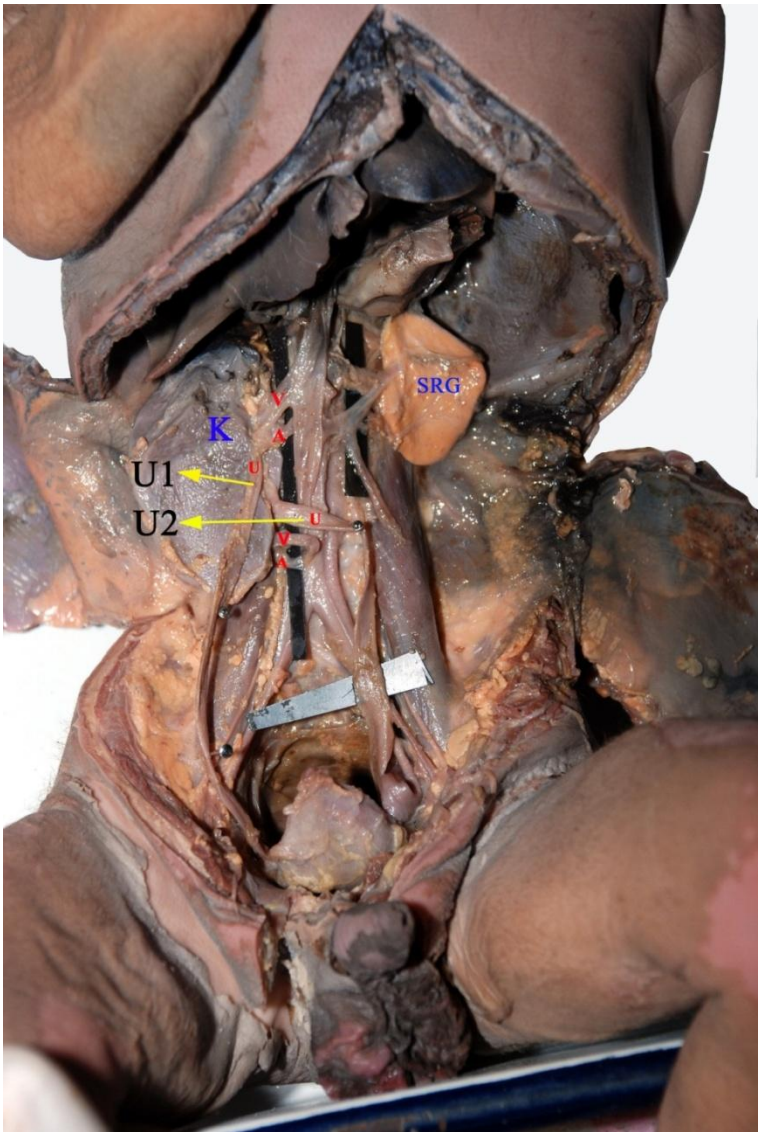

Figure 1: Empty left renal fossa, fused kidney on right side with right ureter (U 1) and left ureter (U2)

During 6^{th} to 8^{th} week of development the foetal kidney ascends along posterior abdominal wall from pelvis to its normal position in lumbar region close to the developing supra renal gland. During its ascent it undergoes 90 degrees axial rotation from horizontal to medial resulting in rotation of hilum from anterior to medial side.

Renal fusion anomalies are of two types i.e. horseshoeing kidney and crossed fused renal ctopia. During their ascent if developing kidneys fuse in the midline it results in horseshoe kidney. If one kidney advances slightly ahead of the other inferior pole of advancing kidney fuses with the superior pole of lower kidney and results in crossed fused renal ectopia.

Pathophysiology of CRE remains unknown. Several theories have been proposed for the embryological explanation for the CRE. CRE was thought to result from defective development of ureteric bud and metanephricblastema during 4^{th} to 8^{th} week of gestation, vascular obstruction during ascent of kidney, pressure from abnormally placed umbilical arteries, malrotation of caudal end of fetus, teratogenic, genetic or environmental factors (Boyan *et al.*, 2007).In the present case the left kidney fused with the lower part of right kidney while in pelvis and followed it during ascent and rotation resulting in medially directed hila.

During ascent the kidneys derive their blood supply from the vessels adjacent to them. Absence of left kidney in its normal location in the present case is associated with absence of left renal vessels and presence of an accessory renal artery for the crossed fused kidney.The arterial supply and venous drainage of CRE are challenging to the nephrologist, radiologist and surgeon.

Mc Donald and McClellan (1957) classified crossed ectopic kidney into 4 types

1. Crossed renal ectopiawith fusion – 90%.
2. Crossed renal ectopia without fusion –uncommon.
3. Solitary Crossed renal ectopia-very rare.
4. Bilateral Crossed renal ectopia – extremely rare.

CRE without fusion is due to mechanical factors. CRE with fusion is of different types depending on the extent and nature of fusion.Male female sex ratio of its incidence is 2:1 and left to right CRE incidence is three times more common than that of right to left CRE (Lee, 1949; Win ram and Ward-Mcquaid, 1959). In the present case the patient was male and it is the left kidney that crossed to the right and fused with the lower part of right kidney resulting in double pelvis. These observations are consistent with that reported in the literature.

CRE with other congenital anomalies of skeletal system, GIT, Cardio-pulmonary are common (Nursal and Büyükdereli, 2005). In the present case it is left upper limb polydactyly was observed.Renal anomalies present a left sided predominance and the exact mechanisms leading to this lateralization can be vascular development, differential gene expression, or susceptibility to environmental factors such as hypoxia (Schreuder, 2011).

The objective of the case report is to highlight the importance of prenatal diagnosis by ultrasonography and postnatal follow-up in the evaluation and management of renal anomalies. CRE rarely causes significant clinical problems. Treatment of cross fused renal ectopia should be targeted to the complications of the anomaly and associated malformations and not the anomaly itself (Felzenberg *et al.*, 1999).Early identification of this anomaly is clinically important for long term follow-up as it is associated with recurrent urinary tract infection, renal stones, uroepithelial tumours at a later stage of life (Patel and Singh, 2008)

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