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Cross-fused Renal Ectopia – A Rare Congenital Malformation

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ABSTRACT

Anomalies of the kidney and urinary tract are frequent, and they often underlie diseases. A uncommon congenital defect known as cross-fused renal ectopia (CFRE) occurs when one kidney migrates to the other side of the developing baby and fuses with the kidney there. According to reports, crossed renal ectopia occurs 1 in 7,000 times. The instance was discovered during regular foetal autopsy performed at Chandigarh's Government Medical College and Hospital's anatomy department. According to the USG report, a male baby with a gestational age of 13+2 weeks was discovered to have a left ectopic kidney fused to the right kidney's lower pole. Because of stasis, an ectopic kidney often results in a higher incidence of stone development. Because supplementary renal arteries are end arteries, a surgeon must take special care of them during surgical operations because the portion of the kidneys they supply will become ischemic if an accessory artery is injured or ligated. In order to prevent postnatal renal dysfunction and morbidity, it is crucial to have a thorough understanding of the morphological differences of the kidneys. Long-term consequences may be avoided by early diagnosis and acknowledgment of an ectopic kidney.

Keywords

Renal ectopia, Fused kidney, Renal fossa, Congenital anomaly, Developing kidney, Urological anomaly

INTRODUCTION

The kidneys are two bean-shaped organs that are located retroperitoneally on the back between the transverse processes of the lumbar vertebrae 3 and 12 of the thoracic spine. With a little variation in position where the right kidney is somewhat inferior to the left kidney due to the liver, the upper poles of both kidneys are typically orientated slightly medially and posteriorly compared to the lower poles. Between the sixth and eighth weeks of life, the kidney begins to

grow embryologically, and during the ninth week, the kidney ascends [1].

It is believed that CFRE begins to manifest between weeks four and eight of pregnancy. Six distinct structural variants of CFRE have been identified, the majority of which are asymptomatic and are discovered accidentally during imaging tests [2].The kidneys are routinely evaluated on second- and third-trimester obstetric sonographic examinations of the foetus. Cross-fused renal ectopia is the fusion of both kidneys, with at least one kidney on the side opposite to where it should be. However, the ureter of the ectopic kidney crosses the midline and proceeds along the regular path to open in the urine bladder in the normal position. One of the rarest urinary system defects is cross-fused renal ectopia. The horseshoe kidney is the most frequent kind of renal fusion abnormality, with cross-fused renal ectopia coming in second. The majority of CRE cases are asymptomatic and are discovered at autopsy, unintentionally while undergoing radiological testing, or when doing cadaveric dissection [3] [2].

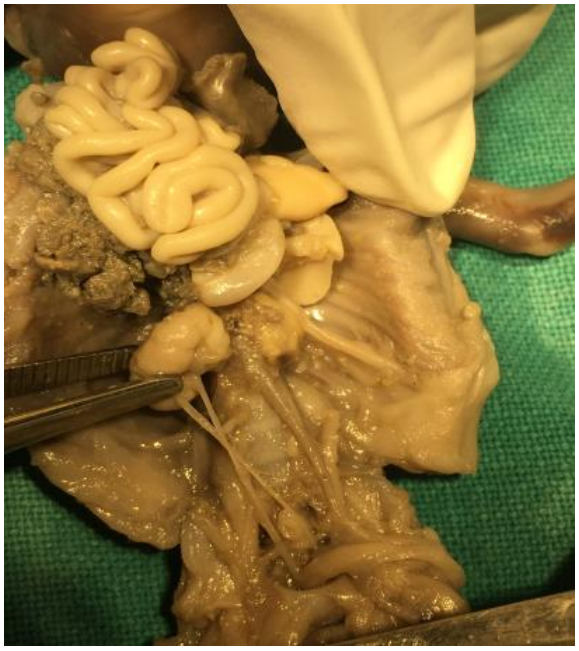
In light of modern surgical procedures like laparoscopic radical nephrectomy, percutaneous nephrectomy, and renal transplant, knowledge of this anatomical variation will therefore be very helpful to the clinician in making a correct diagnosis and providing appropriate treatment [4].

CASE

An autopsy was done on a male foetus with gestational age 13^{+2} weeks sent by obstetrics and gynaecology dept) to the Department of Anatomy, Government Medical College and Hospital, Chandigarh. The indication of medical termination was a USG finding that was suggestive of cross-fused renal ectopia. An informed consent was signed by the parents. They were made aware of the autopsy procedure. Then the foetus was kept in formalin so that fixation could occur and an autopsy could be performed easily. The foetus was examined grossly, followed by photography. Various measurements were taken, like CR Length, Hand length, Crown heel length, foot length, biparietal diameter, etc. Internally, the foetus was examined according to autopsy norms.

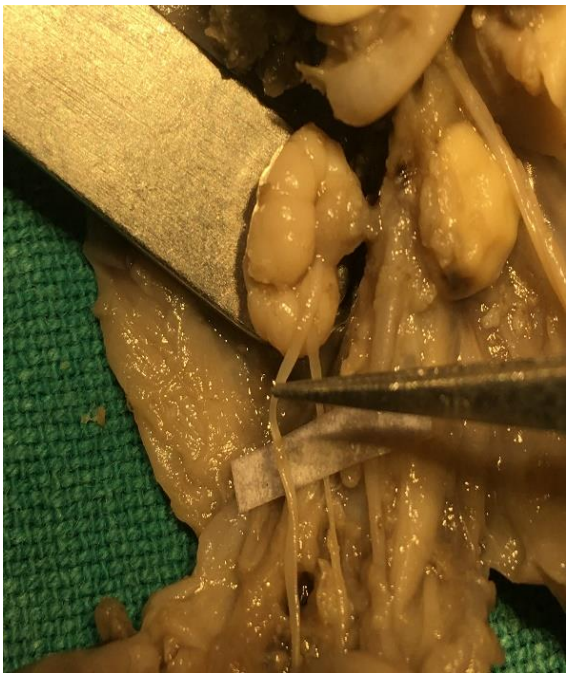
On examination, it was found that the foetal kidneys were fused at the lower end, confirming the presence of cross-fused renal ectopia (CFRE). The type of CFRE observed in our case was classified as L-shaped (Figure 1).

Figure 1: In-situ location of foetal kidneys



A closer view revealed the fused right and left kidneys and ureters, which were opening on their respective sides of the urinary bladder (Figure 2).

Figure 2: Closer view of the right and left kidneys showing ectopia



The mother was 24 years old with a $P_1G_1L_0A_1$ obstetrical history. Maternal Family history indicated no chromosomal abnormality, but a history of diabetes mellitus was seen. Even maternal histories of drug intake related to infertility treatment are seen. Paternal family history of hypertension There was no history of alcohol intake or smoking among both parents.

Ultrasound Findings

Uterus showing intramural fibroids, the cervical cavity measured 3.80 cm in length, the internal os is closed, and no fluid is seen in the cervical canal. Both kidneys with fused poles are imaged in the right renal fossa, suggesting a cross-fused right renal ectopia. Externally, the left foot shows persistent medial rotation (possibly an evolving Tallipes deformity).

External Examination

No congenital malformation was found externally. The foetus was photographed, and autopsies were performed thereafter.

Internal Examination

No other associated anomaly except agenesis of the right lung and presence of macerated liver.

DISCUSSION

Renal abnormalities that are present at birth are among the third most typical birth malformations [1]. Males are more likely to experience it than females [5]. The ureteric bud, which will eventually become the collecting portion of the human kidney, and the nephrogenic cord, which will eventually give rise to the excretory tubules during the fourth week of IUL and is located in the pelvis at the level of the second sacral vertebra (metanephros), communicate as the definitive human kidney develops. The embryonic kidney rises to its typical location, which is in the lumbar region next to the still-developing suprarenal gland, during the sixth to eighth week of pregnancy due to differential development of the abdominal wall. Both kidneys' hilas face anteriorly at first, but eventually turn to face medially as a result of rotation [3]. Ectopic kidneys arise as a consequence of migration stoppage or route deviation during the embryological development of the metanephros [6].

The ureteral hypothesis and mechanical theory are the most often cited reasons of the genesis of crossed fused ectopia. The ureteric hypothesis postulates that severe bending and embryonal caudal end rotation cause the ipsilateral metanephric blastema and ureteric bud to turn away from one another and away from touch. On one side, two kidneys develop as a result of a normal migrating ureteric bud inducing the mesonephric blastema twice. Mechanical hypothesis states that an improperly positioned umbilical artery is to blame for the growing kidney's aberrant ascent, which migrates to the opposite side according to the route of least resistance [7].

Depending on the orientation of the fusion kidney and ureter, McDonald and McClellan classified crossed fused renal ectopia into six types [8]. In descending order of occurrence, they are

- **Unilaterally fused kidney inferior ectopic:** where the lower pole of the orthotopic kidney fuses with the upper pole of the crossed ectopic kidney.
- **Sigmoid or S-shaped kidney:** where the normal side kidney is present superiorly and the direction of the pelvis is medial, the crossed kidney is positioned inferiorly and the direction of its pelvis faces towards the lateral side. As kidneys fuse after complete rotation on the vertical axis, both renal pelvises lie in the correct orientation.
- **Unilateral lump kidney:** where fusion occurs over a wide margin, both renal pelvis are facing anteriorly and placed more inferiorly.
- **L-shaped or tandem kidney,** in which the crossed kidney lies inferiorly and transversely, fused with the lower pole of the normal kidney.

- **Unilateral disc kidney,** in which kidneys fuse along medial borders
- **Unilaterally fused kidney superior ectopia,** the ectopic kidney lies superiorly, and its lower pole fuses with the upper pole of the normal kidney. The renal pelvis of both kidneys are directed anteriorly [7].

The associated ureters of these kidneys drain orthotopically into the bladder according to the categorization given above. individuals are often asymptomatic, therefore the diagnosis is nearly always an accidental discovery. However, 30% of these individuals report with intermittent flank discomfort, dysuria, hematuria, and fever because of blockage or infection. CFRE is often accompanied by nephrolithiasis, hydronephrosis, and blockage of the pelviureteric junction. Imperforate anus (4%), skeletal malformations (4%), and cardiovascular septal abnormalities are further related defects. A reliable method for detecting fused renal ectopia is USG. Its results often point to fused kidneys on the ipsilateral side and a missing kidney in the contralateral renal fossa or pelvis. In order to design surgical operations, contrast-enhanced CT with delayed film is helpful in illustrating the architecture of the draining ureter and renal vascular supplies. The crossing fused renal ectopia has a fairly excellent prognosis if no further defects or problems exist [7].

CONCLUSION

When conducting clinical assessments, imaging tests, and surgical treatments, these renal abnormalities should be taken into consideration [9]. When both kidneys are missing from their usual lumbar location (renal fossa), it is challenging for a radiologist to make a prenatal diagnosis of cross-fused renal ectopia. When identified, one should examine the embryo for further congenital defects that are related [3]. It may be accompanied with renal cysts and stone disease, although when the stone is tiny and the cyst is straightforward, just conservative care and follow-up may be necessary [2]. If there are no other anomalies, women should be advised to continue their pregnancies without fearing the results since the baby after birth will likely be symptom-free and may even have normal kidney function. Additionally, early discovery of renal abnormalities may aid in the meticulous postnatal examination of the infant and can reduce needless fatalities brought on by aberrant renal functioning [3]. Modern techniques of research, including as Ultrasonography, computer tomography (CT), magnetic resonance imaging (MRI) scan, etc., are extremely helpful to identify the ectopic kidney in addition to the standard contrast X-rays, i.e., intravenous pyelogram and ascending pyelogram [10].

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