World Journal of Pharmaceutical Sciences
ISSN (Print): 2321-3310; ISSN (Online): 2321-3086
Available online at: http://www.wjpsonline.org/
Original Article

A retrospective study of crossed fused pelvic kidney

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Received: 01-02-2019 / Revised Accepted: 25-02-2019 / Published: 28-02-2019

ABSTRACT

Crossed fused ectopic kidney is an unusual congenial malformation of the urinary tract in which both kidneys are located on one side of the midline and are fused together. Usually this condition is identified in the autopsy specimen rather than in general clinical scenario. Workup was done in primigravida antenatal mother's along with regular ultrasound screening during pregnancy to detect any abnormality in antenatal mother and prevent complication. The aim of the workup is to screen asymptomatic antenatal mothers and aid them for a healthy outcome of pregnancy. Study was also conducted in 20 cadavers .A19 years old primigravida diagnosed with crossed fused pelvic kidney. [left side] which is a rare anomaly lead to retrospective study of this condition.

Key Word: Crossed Fused Ectopic kidney, Cake Kidney, Primigravida, CADAVERS

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How to Cite this Article: K. Parthiban M, Vino Victor Jesudas, Nandhini Devi S. A retrospective study ofcrossed fused pelvic kidney. World J Pharm Sci 2019; 7(3): 152-155.

INTRODUCTION

Cake kidney or fused pelvic kidney is a very rare congenital anomaly with a very few cases described in the literature [8,9,10]. The term is used to describe completely fused renal mass located in the pelvic cavity and drained generally by two ureters which do not cross the midline. Cake kidney accounts for only about 2% of all fused kidney types. The estimated incidence is 1/65000 to 1/375000 cases.

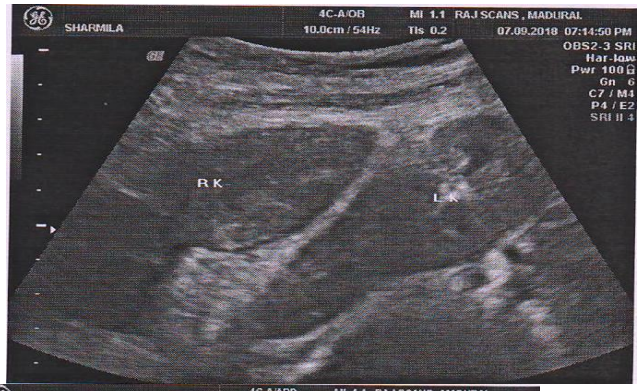
The most common pathologic condition associated with cake kidney is vaginal agenesis, bicornuate or unicornuate uterus, sacral agenesis, caudal regression syndrome, tetralogy of fallot and spina bifida [11,12,13]. Generally patientremain asymptomatic, more common in males [7] with male to female ratio of about 2-3:1. Developmentally, when the renal anlagens fail to ascend and remain in the pelvic cavity extensively fusing with each other. A cake kidney is formed retaining the primitive vascular supply. Vascular supply may be derived from single renal artery and single renal vein. The single renal vascular supply to Cake kidney increases the risk of damage by pelvic trauma, pregnancy or space occupying lesion. This abnormality is usually detected incidentally at autopsy, surgery and radiological investigations. If diagnosed as fused kidney then investigations should be done to rule out other abnormalities. Active intervention is not always needed and patient usually will be asymptomatic.

MATERIALS AND METHODS

The study was conducted in Institute of Anatomy, Madurai Medical College in 20 cadavers and Department of radiology, Government Rajaji Hospital, Madurai Medical College where 60 antenatal cases were screened during the period of June 2018 to Dec 2018. Screening was done in asymptomatic, primigravida with no other co-morbidities with regular antenatal screening. Maternal abdomen was also screened to rule out any congenital abnormities in the mother and to prevent associated complication in pregnancy and for safe delivery. The finding was appropriately documented and ultrasound pictures were taken with consent of the patient. Cadaveric study was also done in this period.

OBSERVATION

A 19 years old primigravida with history of 7 months of amenorrhea, came for routine antenatal ultrasound screening with no other specific complaint. With consent maternal abdomen screened both kidneys were seen in the left side of the pelvis in vertical position and fused together - crossed fused pelvic kidney. Both kidneys were normal in size with regular cortical outline.



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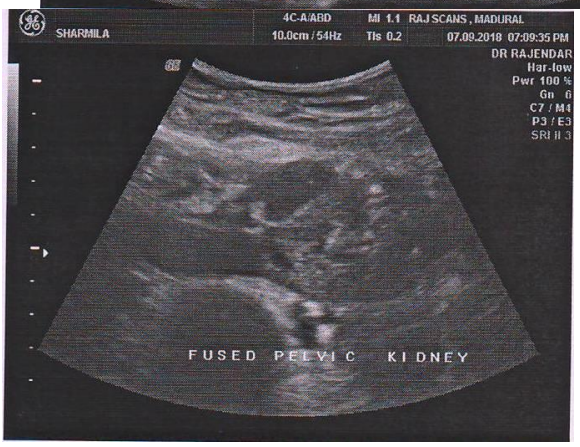
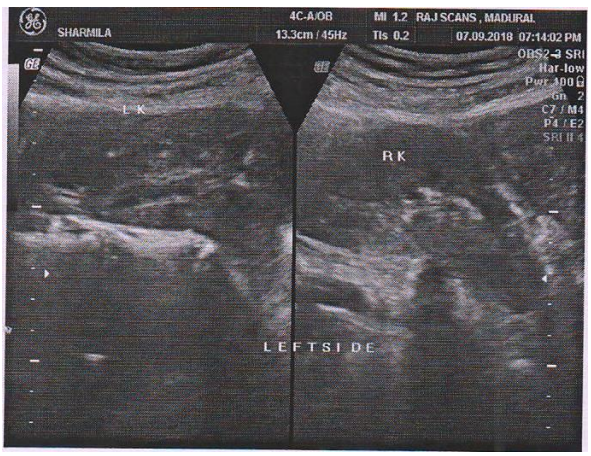




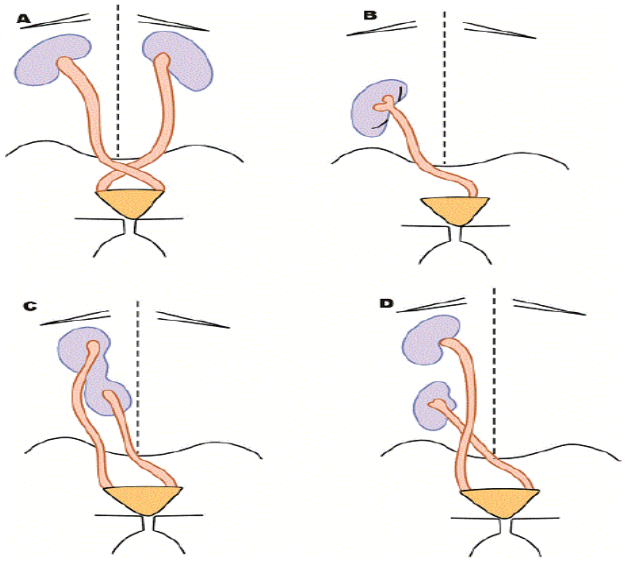
Right Kidney measures – 92*41mm in size.
Left Kidney measures – 97*40mm in size.

Cortical thickness and echoes are normal in both kidneys. No evidence of medical renal disease. No evidence of calculi / hydronephrosis in both kidneys. Both ureters were not dilated. No evidence of focal renal mass lesion/pyelonephritic focus. No Bladder pathology made out. Both kidneys lying at the right paravertebral region at L_4 – L_5 that is at the same level. Superior, mid region and inferior pole were fused. On retrospective study in 60 antenatal mothers and 20 cadavers the anomaly could not be made out which proves it to be the rarest anomaly.

DISSCUSION

CFRE results from over bending and rotation of the caudal end of the developing embryoleading to the inability of the ureteric bud to communicate with the more distant ipsilateral metanephros. The kidney is then attracted to the closer contralateral side.

MC Donaldand Mc Clellan classified crossed fused renal ectopia into six types (1). (A) Unilateral fused kidneyinferior ectopia with the upper pole of the crossed ectopic kidney fusing with the lower pole of the orthotopic ipsilateral mate. (B) Sigmoid or S-shaped kidney in which the crossed kidney lies inferiorly with the renal pelvis directed laterally.(C) Unilateral Lumpkidney with fusion occurring over a wide margin and both renal pelvis directed anteriorly; located more inferiorly. (D) L-Shaped or Tandem kidney in which the crossed kidney lies inferiorly and transversely fusing with the lower pole of the normal kidney. (E) Unilateral disc kidney fusion occurs along the medial border .(F) Unilateralf used kidney superior ectopia type is the least common type the ectopic kidney is placed superiorly with its lower pole fusing with the upper pole of the normal kidney.



CFEK is sporadically reported in the literature because this anomaly may remain as a silent clinical entity without producing any signs and symptoms. Sharma V, reported two cases of L-shaped kidney in females and one case of inferior ectopia in a male (2). Tur K vatan at al reported four cases, of which two were inferior ectopia (both females) and two L –shaped tamdem kidneys (both males) and hydronephrosis was noted in two cases (3). Yin et al described a right to left CFEK of superior ectopia type in male patient with thoracic scoliosis. (4). Sigmoid type of kidney which is second common type of CFEK associated with stayhorn calculus was also reported. (5)

Solanki et al evaluated 5 boys and 1 girl, all having CFEK and noted left to right ectopia in 4 cases and right to left cross over in 2 cases. (6)

Kaufman and find later reported a cake or lump kidney with left to right ectopia located in the right lumbar region in 81 years old female cadaver.(4) Crossed fuse renal ectopia is one of the rare congenital abnormalities. The prevalence of the crossed renal ectopia with fusion was estimated to the 1 in 1000 live births. The incidence of autopsy can vary from 1 in 2000 to 1 in 7500 (2,3].

CREK are generally located in abdomen at a lower level or in pelvic cavity. Crossed fusion variety is most common and present in 90% of cases. Many Theory like influence of genetic factor, teratogenic factor or malignant and abnormal rotation of caudal end of embryo that will lead to aberrant development of metanephric blastema and ureteric during the 4^{th} to 8^{th} week of gestation. Hence both kidneys could not achieve normal position. But the

cause of crossed ectopia in still not known and the shape and site of crossed kidneys depend upon the time and amount of fusion and extent of rotation. Mostly patients are asymptomatic. If symptomatic then the most common presenting symptoms are abdominal or frank pain, a palpable mass, dysuria or hematuria. Ureteral orifices are usually orthotopic. Only 3% have ectopia ureteric orifices. Vesicoureteric reflux (VVC) ureteropelvic junction (UPJ) obstruction, ureterocele, nephrolithiasis and very rarely carcinoma are the common associated anomaly that can lead to pyelonephritis. Investigation like USG, CECT, MRI, IVP, MDCT, UROGRAPHY are the investigation of choice Rarely temporary episodes of urinary pathway obstruction can lead to acute abdominal pain. Our patient had no complaints, finding was only accidental. Only regular follow up needed. If symptomatic patients can be easily managed only by conservative measures. Our patient delivered an alive preterm male baby by Cesarean section[obstetric indication] and neonatal screening of baby abdomen by ultrasound had no abnormalities.

CONCLUSION

Mostly detected incidentally commonly detected in autopsy specimen. The cadaveric study done also proves that this is a rare anomaly. This can be observed in various clinical forms and management should be planned according to the clinical presentation and anatomical abnormality because most of the patients remain asymptomatic throughout their life.

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