A RARE CASE OF CROSSED FUSED RENAL ECTOPIA WITH LATERAL FUSION

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Abstract:
Crossed renal ectopia with fusion is relatively uncommon. The reported incidence in literature is about 1 in 2000. We had a 52 year old female presenting with left lower abdominal pain and no other urological complaints. Clinical examination was found to be normal. Ultrasound abdomen showed empty right renal fossa and on left side two kidneys were fused together with gross hydronephrosis with thinned out cortex of one kidney. Contrast enhanced computerised tomography of abdomen and pelvis revealed right to left crossed kidney with fusion of right kidney with convex border of malrotated left kidney. Crossed right kidney had gross hydronephrosis and thinned out cortex. Cystoscopy was done and ureteric orifices were found in normal position. Scintiscan was done, which revealed less than 5 percent function of crossed right kidney. Nephrectomy of ectopic right kidney was done. Specimen confirmed to have right pelviureteric junction obstruction. (PUJO)

Keyword: CROSSED RENAL ECTOPIA, LATERAL FUSION, PUJO

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INTRODUCTION
Crossed renal ectopia with fusion is one of the rare congenital anomaly with autopsy incidence of about 1 in 2000\(^1\). Exact incidence is unknown because of its rarity and different type of fusion anomalies as well as variance in reporting of cases. It has slight male predominance.

CASE REPORT:
We had a 52 year old female who presented with left lower abdominal pain. The pain was dull aching and non radiating. No other urological symptoms were present. Clinical examination was found to be normal. Basic urine and blood investigations were within normal limits. Ultrasound abdomen showed absent kidney in the right renal fossa and on left side, two kidneys were fused together with gross hydronephrosis with thinned out cortex of the crossed kidney.
Contrast enhanced computerised tomography of abdomen and pelvis revealed right to left crossed kidney (fig. 1) with fusion of convex border of right kidney with convex border of malrotated left kidney (fig. 2, fig. 3). There was gross hydronephrosis of right kidney with thinned out cortex (fig. 4, fig. 5) and abnormal blood vessels to the kidneys (fig. 6, fig. 7). Cystoscopy was done and normally positioned bilateral ureteric orifices were confirmed. Scintiscan (fig. 8) was done, which revealed less than 5% function of the ectopic crossed kidney. Radiological findings were confirmed intraoperatively (fig. 9). Right nephrectomy was done through modified Gibson’s incision. Specimen sent for histopathological evaluation which confirmed right PUJO (fig. 10) as cause of hydronephrosis. This case is reported for its rarity and different type of fusion anomaly.
Figure 5 CECT - Fusion of the both convex borders with gross hydronephrosis of crossed right kidney.

Figure 6 CECT - Reconstruction. Fused kidneys at the level of bifurcation. Figure 7 CECT - Abnormal left renal vessel course

Figure 8. Scintiscan showing fusion and poor function of crossed right kidney. Figure 9. Intraoperative picture showing fusion of both convex borders with free poles

DISCUSSION:
Crossed renal ectopia with fusion is one of the rare congenital anomalies with autopsy incidence of about 1 in 2000\(^1\). When a kidney is located on the side opposite from that in which its ureter inserts into the bladder, the condition is known as crossed ectopia. Ninety percent of crossed ectopic kidneys are fused to their ipsilateral mate\(^2\). Left to right crossover with inferior pole fusion is most common.
Embryologically the exact cause of Crossed ectopia is unknown. Wilmer (1938) suggested that crossover occurs as a result of pressure from abnormally placed umbilical arteries that prevent cephalad migration of the renal unit, which then follows the path of least resistance to the opposite side. Cook and Stephens (1977) postulated that crossover is the result of mal alignment and abnormal rotation of the caudal end of the developing foetus, with the distal curled end of the vertebral column being displaced to one side or the other.

The McDonald and McClellan classified the fusion anomalies as (1) unilateral fused kidney with inferior ectopia; (2) sigmoid, or S-shaped; (3) lump or cake; (4) L-shaped, or tandem; (5) disc, shield, or doughnut; and (6) unilateral fused kidneys with superior ectopia. The unilaterally fused kidney with inferior ectopia is the most common type, whereas fusion with superior ectopia is the least common. The vascular supply to the kidneys are variable and unpredictable. Most of the cases are asymptomatic and diagnosed on radiological evaluation for other causes. When patients present symptomatically it is usually in fourth or fifth decade because of complications due to associated anomalies. Associated anomalies in ectopic kidney include cystic dysplasia, UPJ obstruction (29%), reflux (15%), or carcinoma. Asymptomatic patients can be followed up and symptomatic patients treated according to associated anomalies or complications.

Largest series of crossed ectopia is so far reported by Abeshouse and Bhisitkul (1959). They compiled 443 reports of crossed ectopia with fusion. Multiple small numbers of case reports have been published in journals with varied presentations and combinations. Glodny and colleagues (2008) reported on 24 crossed fused ectopia found on a CT scan performed for nonurologic indications.

In our case patient had right to left crossover with fusion at middle convex border of both kidneys with PUJO of right kidney. Right kidney was grossly hydronephrotic and thinned out cortex with poor function, hence treated with right nephrectomy.

**CONCLUSION**

Crossed renal ectopia with fusion is rare anomaly and most patients are asymptomatic and symptomatic patients usually present in their fourth or fifth decade with associated complications. This case is reported here for its rarity and different type of fusion anomaly which was not widely reported.

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