A RENAL CONGENITAL ANOMALY - UNILATERAL EXTRACALYCEAL ECTOPIC RIGHT KIDNEY WITH MULTIPLE ABERRANT RENAL VESSELS

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INTRODUCTION

Kidneys are a pair of excretory organs situated on the posterior abdominal wall, one on each side of the vertebral column, behind the peritoneum. Kidney develops from metanephros, through pronephros and mesonephros appear to disappear. Kidney starts developing in the sacral region then it ascents upward into renal fossa and migrates with the help of some factors (Krishna garg, PS mittal, Mrudula chandrupatia.2013).

Migration of the right ureteric bud to the left, inducing the left metanephric blastema twice to form the two kidneys on the left side, could be the probable cause of this anomaly. According to Bailey SH, Mone MC, Nelson EW 2002, the incidence of this rare congenital malformation was 1 in 1000 live births, with slight male preponderance (M:F = 3:2), left side being more commonly affected; the incidence in autopsy series being 1 in 2000 (as it is often undiagnosed throughout life, presenting as a silent anomaly). Familial incidence of crossed fused renal ectopic, horse shoe kidneys and dysplastic kidneys were reported by Rinat C Farakas A, Frishberg Y 2001, who suggested that an autosomal dominant inheritance pattern and an improper timing of induction of metanephric blastema by ureteric bud could be the possible etiology of this anomaly. Though mostly undiagnosed throughout life, crossed fused ectopic kidneys with extracalyceal multiple renal artery could provide a formidable challenge during surgery (De Virgilo C, Gioviczi P, Cherry KJ, Stanson AW, Bower TC, Hallet JW Jr., Pairolero PC 1995).

The present case report not only aims at explaining the possible embryological basis of this rare congenital malformation, but also the risk factors associated with the surgical interventions in crossed fused ectopic kidney. Hence, the knowledge of the uncertain anatomy of such atypical renal anomaly is of equal interest to both the anatomist as well as the surgeon (especially for successful donated cadaveric kidney transplants).

Case Report:

Crossed ectopia is an uncommon condition in which a kidney is located on the opposite side where its ureter inserts into bladder (Rodrigo GV, Beltran AJR, Sala A A 2001) Crossed ectopic kidneys are fused to their ipsilateral mate in more than 90% of cases. Crossed ectopia without fusion is rare (1 in 75,000 autopsies) as compared to the fused crossed ectopia with fusion (Nursal GN, Buyukdereli G 2005). We here by present a case of crossed fused renal ectopia-inferior type found in Routine dissection of the abdomen of a 63 year old male cadaver in the Dept. of Anatomy, Hi-tech medical college rourkela. Revealed the presence of a rudimentary ectopic right kidney fused to the lower pole of the left, with multiple aberrant renal arteries. The ureters were normal, the left kidney (upper) had a normal pelvis while the right (lower) had an extra renal pelvis (extra calyx). The two ureters entered the bladder normally, Right side showed empty renal fossa with an intact adrenal gland. The possible embryological causes of this anomaly have been discussed here. Even though this type of condition can be presents as a silent renal anomaly (it can be undiagnosed throughout life and revealed through autopsy or dissection) the surgical implications are noteworthy, which too have been highlighted in this report.

Keywords: Crossed fused renal ectopic, Multiple aberrant renal vessels, Extra renal pelvis of ureter, Malrotation and migration of ureteric bud.

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A Renal congenital Anomaly - unilateral extra calyceal Ectopic Right Kidney with Multiple Aberrant Renal Vessels

fusion anomalies have been often done due to shortage of compatible live donor pools.

Pic. 1 congenital anomaly of kidney showing upper pole of right kidney fused with lower pole of left kidney with right extra calyx and multiple aberrant renal artery within the abdomen

Pic. 2 congenital anomaly showing 1st, 2nd and 3rd renal arteries.

Pic. 3 Congenital anomaly of kidney showing upper pole of right kidney fused with lower pole of left kidney with right extra calyx and multiple aberrant renal artery outside the abdomen

DISCUSSION

Crossed fused renal ectopia is a unique situation where the two kidneys are seen lying on one side of the spine, the right kidney being fused to the lower pole of the left (as in this case) with normal literature so far. The various fusion anomalies are designated as (i) unilateral fused kidney with inferior ectopia (ii) sigmoid or S shaped kidney; (iii) lump or cake; (iv) L-shaped or tandem; (v) disc, shield, or doughnut; and (vi) unilateral fused kidneys with superior ectopia (Stuart B B, Alan D, Perlmutter, Alan B R 1992). This classification lends some order to an understanding of the embryology of renal ascent and rotation. The present case is unilateral fused kidney with inferior ectopia type, this condition often presents as a silent anomaly, remaining undiagnosed throughout life (Boyan N, Kubat H, Uzum A 2007). However, diagnosis by renal USG, IVP, CT scans (with contrast) and renal scintigraphy (technetium scans) is important, as the fused ectopic kidney may be an important predisposing factor for PUJ obstruction, recurrent UTI, calculous disease and renal neoplasms. Severe hydronephrosis from PUJ obstruction secondary to aberrant renal arteries obstructing extrarenal pelvis of ectopic kidney is quite common. The anomalous extrarenal pelvis of the ectopic kidney (due to incomplete ascent and malrotation of ureteric bud) may lead to recurrent UTI and stag horn calculi. Incidence of struvite, newberyite and carbonated-apatite calculi requiring ESWL and an atrophic nephrolithotomy (Stubbs AJ, Resnick MI 1977). Location of the ectopic kidney close to the pelvic cavity often causes the pain of renal colic in calculus disease to mimic PID (not the typical loin to groin pattern), thus making the diagnosis of renal stones quite difficult. Such ectopic fused kidneys often present with renal neoplasms like renal cell
carcinoma, Wilms' tumour or invasive transitional cell carcinoma (involving extrarenal pelvis). These cases may require laparoscopic guided hemi-nephrectomy or nephroureterectomy, preserving adequate renal perfusion (Gur U, Yossepowitch O, Baniel 2003) Present case was similar with the Sukanya Palit, Asis Kumar Datta, Arunabha Tapadar 2008 studied ,they observed that the presence of a rudimentary ectopic right kidney fused to the lower pole of the left, with multiple aberrant renal arteries arising at variable levels from the abdominal aorta. and Rajesh Sharma, Rupali Bargotra 2009, found Crossed Fused Renal Ectopia-Inferior Ectopia Type through the ultrasonography and also O. Hochwald, R. Shaoul observed crossed fused ectopic left kidney in his study14. Knowledge of ectopic fused kidneys with aberrant renal vasculature and normal ureters with pelvicalyceal system (as highlighted from this cadaver) is equally important in cadaveric renal transplants, keeping in mind the uncertain anatomy of anomalous blood vessels at the renal pedicle (Uzzo RJ et al, 2002). Successful renal transplants from such cadaveric fusion anomalies have been often done due to shortage of compatible live donor pools.

CONCLUSION

The present case report showing the ectopic rudimentary right kidney fused to the lower pole of the left with multiple aberrant renal vessels, aims at highlighting the uncertain anatomy and possible embryological explanation of this rare congenital malformation. Even though the anomaly is revealed only on autopsy series and may remain undiagnosed throughout life the clinical implications are immense on proper diagnosis. Hence the anatomical knowledge of such renal anomalies would minimize the perioperative and postoperative morbidity related to surgical interventions and cadaveric transplantation procedures

REFERENCES