Case Report

Cross fused ectopic kidney a rare congenital malformation with review of literature

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ABSTRACT

Crossed renal ectopia is one of the rarest urinary system anomalies. It is more often seen at autopsy than in clinical practice. Most cases of renal ectopia remain asymptomatic during life and are diagnosed incidentally. Renal ultrasonography, intravenous pyelography, computerized tomography, and renal scintigraphy (technetium-based tests) are the imaging methods used for diagnosis. Demonstration of crossed renal ectopia is important because it is a predisposing factor for obstruction, infection, and neoplasia of the urinary system. We report male patient who had unremarkable clinical findings. He had left-to-right inferior crossed renal ectopia with fusion. Surgeons should be aware of ectopic and fused kidneys to minimize perioperative complications because of the uncertain anatomy.

Keywords: Crossed fused renal ectopia, Urinary system anomalies, Ureter, Embryology, Renal anomaly

INTRODUCTION

Crossed fused renal ectopia is a rare congenital malformation, which is reported to be usually asymptomatic but may have varied presentations. Urinary system anomalies include renal agenesis, multiple kidneys, renal ectopia, and fusion defects. Demonstration of a congenital renal anomaly is important in the treatment of patients with renal infection and in surgery of the urinary system (Bhatnagar et al., 2004; Stimac et al., 2004). Crossed renal ectopia, in which one of the kidneys lies on the contralateral side but whose ureter passes to the ipsilateral side, is the second most common renal fusion anomaly following horseshoe kidney (Kwon et al., 2004), and in different autopsy series it is detected in 1 in 1000 to 1 in 7000 (Hwang et al., 2002, Yano et al., 2003; Guarino et al., 2004). It is more often seen in postmortem studies than in clinical practice. Most patients with renal ectopia are asymptomatic during life and the number found clinically is estimated to be only 1 in 10000 patients (Guarino et al., 2004). When a kidney is located on the side opposite from which its ureter inserts into the bladder, it is defined as crossed renal ectopia and if it is fused with the opposite kidney then it is defined as crossed fused renal ectopia. Ninety percent of crossed ectopic kidneys are fused to their contralateral mate. Crossed ectopia is always combined with abnormally located ureters, and the ureter or ureters cross the midline at the level of the distal aorta or its bifurcation to enter the bladder at the normal position (Yano et al., 2003; Kwon et al., 2004; Sood et al., 2005). We report a patient with crossed fused renal ectopia who was admitted for observation post Road traffic accident.

CASE REPORT

A 38 years old male came to emergency ward with history of fall from bike. Clinically patient’s GSC was 15/15 with mild abrasion on chest and abdomen with mild pain in abdomen. Investigations like X-ray chest and...
abdomen were within normal limits, ultrasonography and CT scan (Figure 1 & 2) revealed crossed left ectopic kidney fused with the normal right and left kidney. Patient was observed for 24 hours post trauma and was discharged next day as he had no fresh complaints.

DISCUSSION

Crossed fused renal ectopia is thought to result from the abnormal development of the ureteric bud and metanephric blastema during the fourth to eighth weeks of gestation. After horseshoe kidney, crossed fused ectopia of the kidneys is the most frequent fusion abnormality of the urinary tract with a male predominance of 3:2. Wilmer, in 1938, first categorized the fusion anomalies of the kidney and McDonald and McClellan, in 1957, included crossed ectopia with fusion, crossed ectopia without fusion, solitary crossed ectopia and bilateral crossed ectopia in a modified classification. The currently accepted classification, which helps in understanding the embryology, renal ascent and rotation, is depicted in Figure 2. These abnormalities are clinically significant because approximately half the patients manifest complications e.g., hydronephrosis, infections and nephrolithiasis. Left to right crossover occurs more frequently and the upper pole of the crossed ectopic kidney is fused to the lower pole of the normally located kidney in most instances. In the present study left to right crossover was seen.1,4 The characteristic ultrasonographic findings in crossed fused renal ectopia include an anterior and/or posterior notch with difference in orientation of the 2 collecting systems in the fused kidneys.5,6 In addition, ultrasonography can give vital information on the arterial supply and venous drainage, which can be grossly abnormal. Calyceal dilatation and distortion, hydronephrosis and urolithiasis can also be diagnosed with ultrasonography.7,8 The other imaging modalities that can be used for anatomical imaging include intravenous pyelography and contrast-enhanced CT scans; the magnetic resonance imaging. The exact incidence of crossed fused renal ectopia is not known because a large majority of patients are asymptomatic; the estimated prevalence in autopsy series is 1:2000.2 Anecdotal cases have been reported with hypertension.9 These malformations may be associated with symptoms related to infection (pyelonephritis), obstruction (hydronephrosis due to pelviureteric junction obstruction) and urolithiasis.1,8 Entrapment of the ureter in the isthmus leading to hydronephrosis, as seen in case 3, has not been reported before. Vesicoureteral reflux is also commonly associated;10,11 this may be responsible for the high incidence of pyelonephritis. A micturating cystourethrogram is required whenever vesicoureteral reflux is suspected. These associations also place demands on the functional assessment of the upper tracts; we have preferred radionuclide scans for such assessments because of lesser radiation exposure, easy availability, recurrent usage and effective diagnostic accuracy. The cortical scans, using DMSA, help in the detection of scars and the renal dynamic scans, using IIEC, help in the evaluation of differential renal function and the drainage pattern (obstructive or non-obstructive) of the upper tracts. Thus, the investigations need to be tailored to patient's specific needs. There are no specific guidelines for the management of crossed fused renal ectopia. The fused renal units do not need to be separated. The treatment is guided toward the associated problems that lead to either symptoms or the deterioration of the upper tracts e.g., a pelviureteric junction obstruction would require a pyeloplasty or vesicoureteral reflux would require either injection of a bulking agent or reimplantation of ureter(s).
CONCLUSION

In conclusion, crossed fused renal ectopia is mostly detected incidentally during investigation for other problems. It is more common in boys. The left moiety crosses over to the right in the majority of cases. And, when urological problems are associated, they require appropriate surgical management. So it's important for surgeon to know about such renal anomalies.

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REFERENCES
