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Case Report

Single ureter draining crossed fused renal ectopia with single renal vein: case report

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ABSTRACT

We present very rare case of incidentally detected crossed fused renal ectopia (CFRE) with a single ureter, single renal vein in a 14-year-old boy who presented as peri umbilical pain and chronic constipation. His contrast enhanced computed tomography (CECT) showed the left kidney was fused to the lower pole of the right kidney with single ureter draining both the moieties into the urinary bladder on ipsilateral side and single renal vein draining into the inferior vena cava. CFRE is usually asymptomatic and undetected in the absence of renal pathologies. Reporting such cases will help in diagnosis and management when symptomatic.

Keywords: Cross, Fused, Renal ectopia with single renal vein, Single ureter

INTRODUCTION

Crossed fused renal ectopia is a very rare congenital anomaly in which both the fused kidneys lie on one side of the patient's body and usually drained by double pelvis and ureters which ultimately drain into the urinary bladder bilaterally.[1,2]

Majority cases are asymptomatic and diagnosed as an incidental finding and approximately 30% patients present with on and off flank pain, dysuria, hematuria, and fever when associated with pathology.[3]

We encountered an unusual case of incidentally detected crossed fused renal ectopia with single ureter, single renal vein, and two renal artery came for mild pain due to chronic constipation. Only one case was reported of this variant of crossed fused renal ectopia to the best of our knowledge. Reporting such cases with various vascular and ureteric anomalies will help in diagnosis and planning management in future.

CASE REPORT

A 14-year-old male patient presented with chronic constipation with mild, intermittent and dull peri umbilical pain. His complete blood count, renal function and urine microscopy examination was normal. An abdominal ultrasound (US) revealed absence of left kidney in the left renal fossa and contrast enhanced computerised tomography with 3 D reconstruction shows left kidney on right side which is fused at lower pole of right kidney, having single ureter draining both the kidneys into the urinary bladder on the right side and single renal vein draining the renal parenchyma into the inferior vena cava (Figures 1a-e and 2). The fused kidneys were supplied by two renal arteries originating one from aorta and other from the right common iliac artery (Figure 2b). There are no associated other congenital abnormalities. His constipation was treated with laxative and low fibre diet which relives his pain. However, the patient is under periodic follow-up as such cases may show malignant transformation.

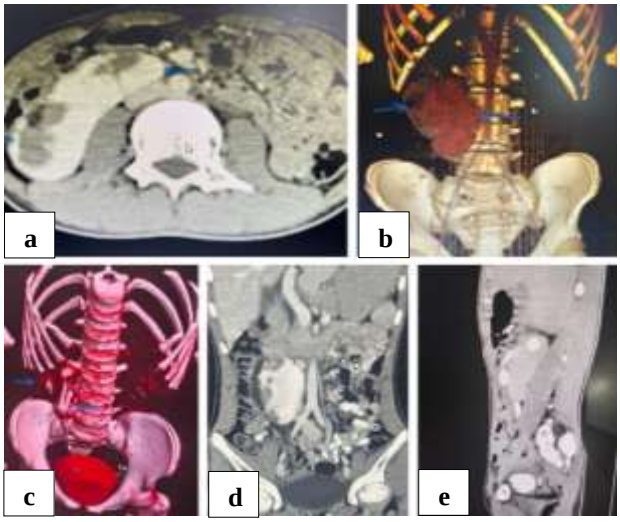

Figure 1: (a) Axial section, (b) and (c) volume rendered 3D reconstruction showing crossed fused renal ectopia with pelvicalceal system of both kidneys (shown by arrows), (d) coronal view of venous phase, and (e) sagittal view of excretory phase in contrast CT scan showing single ureter draining both kidney on ipsilateral side.

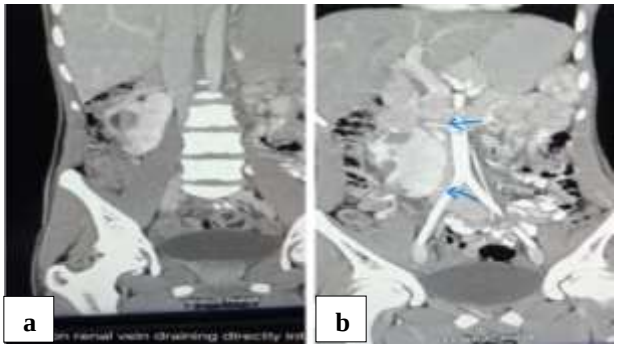

Figure 2: (a) Coronal view of venous phase in contrast CT scan showing single renal vein draining both kidneys, and (b) coronal view of arterial phase of contrast CT scan showing two renal arteries arising from aorta and common iliac artery.

DISCUSSION

Glenn et al first used the term to describe renal fusion into one mass which is rare congenital anomaly stand second after horseshoe kidney and usually supplied by a single renal artery and drained by two separate ureters.[2]

The most commonly accepted embryological basis of crossed fused renal ectopia are ureteral theory and mechanical theory. Six variations of crossed fusion have been reported: type 1, inferior crossed fused ectopia; type 2, sigmoid or S-shaped kidney; type 3, unilateral lump kidney; type 4, unilateral disc kidney; type 5, L-shaped kidney; and type 6, superior crossed fused ectopia.[1,3] However, drainage by a single common ureter is an unusual variant till time reported only in seven cases and drainage with single ureter and single renal vein very rare reported only one case.[1-3]

Crossed-fused renal ectopia, which can remain asymptomatic with male predominance throughout life and either went undetected or diagnosed incidentally.30% of patient associated with pathology like obstruction or infection and presents with flank pain, dysuria, hematuria, or fever occasionally be palpated as abnormal abdominal masses.[1,4]

Anomalies frequently associated with crossed ectopia are imperforate anus (4%), skeletal abnormalities (4%), and cardiovascular septal defects. Index patient is incidentally detected as came for periumbilical pain due to chronic constipation.[1,3]

Crossed fused renal ectopia has high clinical significance due to its arterial and venous abnormal course. It is essential for clinicians and surgeons to understand such anomalies, as they could impact patient care.[5]

USG is a good modality to demonstrate the presence of fused renal ectopia. The sonographic findings usually suggest an absent kidney in the contralateral renal fossa or pelvis and fused kidneys on the ipsilateral side.[3]

Contrast-enhanced CT with three-dimensional reformatted images helps make an accurate diagnosis of crossed fused renal ectopia with visualization of the number of draining ureters and its vascular supply for better surgical management. It is helpful to screen for the presence of stones, hydronephrosis or masses. Therefore, MDCT urography enables a comprehensive evaluation of patients with renal fusion anomalies in a single examination.[1]

Our case is very rare having crossed fused ectopia which is drained by single ureter on right side, venous drainage is by single renal vein which drains in vena cava and supplied by two renal arteries one arising from right lateral wall of aorta and other just from right common iliac artery and there are only one similar reports in literature.

Surgeons should be aware of this type of kidney anomaly as it may affect the position and course of the ureter, renal vessels, and kidneys themselves if surgical intervention required to avoid the complication.[4] Regular follow-up visits are recommended in such patients due to the risk of calculi, infection, obstructive nephropathy, renal cell carcinoma, transitional cell carcinoma, and Wilms' tumor have been reported in crossed fused renal ectopia cases.[1,2]

If there are no associated anomalies and complications, then the prognosis of the crossed fused renal ectopia is very good as like in index patient.[3]

CONCLUSION

Cross fused renal ectopia with single renal vein and ureter is very rare and reporting of which will help in the future

understanding and management when associated with pathology to avoid the complication as such condition may have various vascular and ureteric anatomical variation.

Multi-detector CT is preferred investigation for comprehensive evaluation of patients with renal fusion anomalies in a single examination.

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