Case Report

CROSSED RENAL ECTOPIA ASSOCIATED WITH MALROTATION OF INTESTINE- A RARE CASE REPORT

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ABSTRACT

Background: Kidneys and ureters demonstrate a wide range of anomalies due to its complex development. One such anomaly would be the crossed renal ectopia which is fusion of both kidneys on to one side and malpositioned at the pelvis**.** The occurrence of these anomalies, though rare, is due to multifactorial reasons. The renal anomaly was observed in a formalin fixed adult cadaver during the undergraduate medical dissection. The abdominal cavity, on opening, revealed malrotation of the gut with the large intestine on the left side and the small intestinal loops on the right side. The left renal fossa was empty due to fusion of the left kidney with the right kidney forming a lump kidney. The ureter of the left kidney was draining in to its normal position into the urinary bladder. The position of the lump kidney was in the right iliac fossa. It was having arterial feeders from the abdominal aorta and the common iliac arteries and was draining into the inferior vena cava and the right common iliac veins by 5 renal veins. Crossed renal ectopia rarely may be associated with malrotation of the gut. Awareness of such anomalies could be due to incidental finding and helps the surgeons and radiologists in their diagnosis and planning and preventing postoperative complications.

KEY WORDS: CROSSED RENAL ECTOPIA; FUSED KIDNEY; LUMP KIDNEY; MALROTATION; INTESTINE.

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INTRODUCTION

Congenital anomalies of the kidney vary from absence of kidney to abnormal location, shape and the vascular supply. The orientations of the kidneys are also changed. These renal anomalies result due to number of factors like genetic factors, aberrations at molecular levels and environmental reasons. These anomalies are associated with abnormal maturation and failure of differentiation and growth during this line needs change appropriate time during the critical period in the development [1].

The lump kidney or cake kidney is a variety of crossed ectopic kidney and is located on the opposite side of the midline from its ureteral insertion into the urinary bladder. 90% of them are fused generally to their counterpart's inferior pole.

McDonald and McClellan classified ectopic kidneys into six varieties. Of these the lump kidney is very rare, the total kidney mass being irregular and lobulated. In most cases the mass remains in the true pelvis but the ascent may progress only up to the sacral promontory. Both renal pelves are anterior and drain separate areas of the parenchyma. The ureters do not cross but occasionally either the normal ureter or the crossed ureter may have an ectopic orifice

in the urinary bladder [2].

Here we reporting a rare case which was a combination of crossed renal ectopia with Malrotation of intestine.

CASE REPORT

During the routine dissection of abdomen region of a formalin fixed male cadaver of middle age, the following variations were noted in the large intestine and kidneys:

When the abdominal cavity was opened, it was observed that the caecum, vermiform appendix, and the ascending colon were situated on the left side of the abdominal cavity along side of the descending colon. The transverse colon was rudimentary. The descending colon was in its normal place (Figure 1).

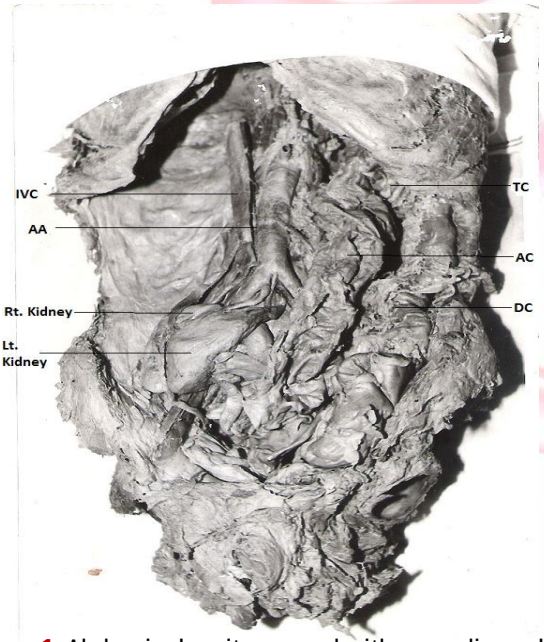

Figure 1: Abdominal cavity opened with ascending colon (A.C.), descending colon (D.C.) on the left side of the abdominal cavity, small transverse colon (T.C.). Both kidneys are on the right side with left kidney medial to the right kidney. Abdominal aorta (AA) and inferior vena cava (IVC) are seen in the posterior abdominal wall.

The left kidney was absent in its normal position. The right kidney was found near the pelvic brim lying in the right iliac fossa and appeared like an irregular lobulated mass. This was because the left kidney had fused with the right kidney. The hila of both kidneys were facing anteromedially (fig.2 & 3). The left ureter was crossing from right to left side to open into the urinary bladder in the normal position (Figure 3). The renal arteries of both kidneys were multiple and arising from the lower abdominal aorta and right common iliac artery. There were five renal veins draining into the right common iliac vein and inferior vena cava (Figure 2, 3). The right part of the abdominal cavity was occupied by coils of small intestine. Hence the anomaly was a crossed ectopia of kidney associated with the malrotation of the intestine.

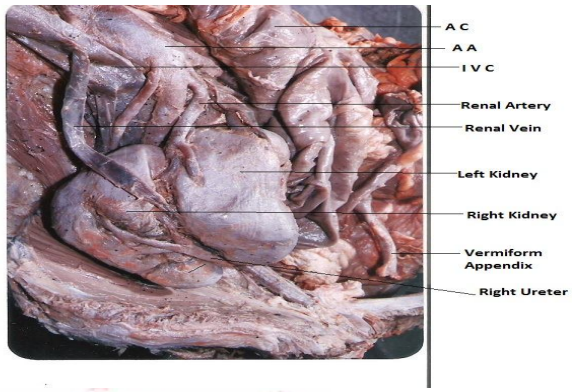

Figure 2: The kidneys are seen near the pelvic brim on the right side. The Right kidney is on the right side and medial to the left kidney. Both kidneys receiving multiple renal arteries from abdominal aorta (AA) and right common iliac artery. Renal veins are draining into inferior vena cava (IVC). The ascending colon, caecum and vermiform appendix on the left side of the abdominal cavity.

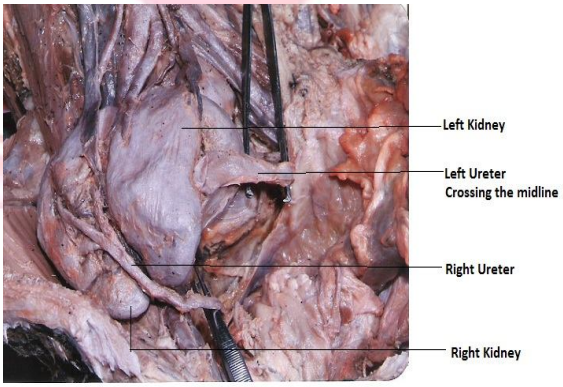

Figure 3: Shows the renal ectopia with the ureters. The left ureter is running horizontally, crossing the midline to go to the left side of the urinary bladder to open at the normal site.

DISCUSSION

The kidneys develop in the true pelvis and gradually ascend into the abdomen with the hila lying at the level of body of first lumbar vertebra about 5 cm lateral to the midline. The congenital anomalies of the kidney and ureter are seen to vary from complete absence of kidneys to the abnormal locations, orientation, size and shape. The congenital anomalies also are commonly seen in the ureters and vascular supply to the kidneys. The reasons for such anomalies are multifactorial and could be genetic [1].

Renal ectopia results from failure of normal ascent of the embryonic kidney. This may arise as a result of abnormalities of the ureteric bud or metanephros, an abnormal vascular supply or genetic abnormalities. The incidence of renal ectopia in postmortem studies varies from 1 in 500 to 1 in 1290. It occurs slightly more frequently on the left side and 10% of cases are bilateral. The male to female ratio is equal. Around 50% remain unrecognized throughout life. In parts of Kenya there is a 3 times increased incidence in unrelated subjects, suggesting an as yet another undefined environmental factor [3,4,5].

Another study compiled 443 reports of crossed renal ectopia with fusion and estimated its occurrence to be 1 in 1000 live births. In crossed renal ectopia usually the left kidney presents the anomaly [6].

According to Wein the renal anomalies have been classified into various types. They are as follows: renal agenesis (unilateral or bilateral), supernumerary kidney, simple renal ectopia, crossed renal ectopia without fusion and with fusion. The crossed renal ectopia with fusion are further classified into sigmoid or S- shaped kidney, Lump kidney or cake kidney, L- shaped kidney or Tandem kidney, Disc or shield or Doughnut or Pancake kidney, Superior or inferior ectopic kidney, Horse shoe kidney, solitary crossed ectopia and bilateral crossed ectopia without fusion. The present case report showed a crossed renal ectopia with fusion of lump kidney variety according to this classification.The unilateral fused kidney with inferior ectopia is very common, with male dominance and left to right cross over is frequent [1].

The lump kidney or cake kidney is a relatively rare form of fusion. Extensive joining has taken place over a wide margin of maturing renal anlage. The total kidney mass is irregular and lobulated. Usually ascent progresses only as far as the sacral promontory, but in many instances the kidney remains in true pelvis. Both renal pelves are anterior and drain separate areas of parenchyma. The ureters do not cross.

Retrospective analysis of six patients with crossed renal ectopia revealed that they were asymptomatic and diagnosed when the patients came for other investigation. They could be asymptomatic. These anomalies are associated with other urological problems which may need surgical treatment [7].

The factors responsible for the change in the position of kidney during development are uncertain. The cross over occurs as a result of pressure from abnormally placed umbilical arteries which prevent cranial migration of kidney and the kidney ascends towards the least resistance site i.e., towards the opposite side [8].

Strong and undetermined forces are responsible for ascent of the kidneys and these forces attract one or both kidneys to their final destination on the opposite side or midline [9].

It has been postulated that cross over is the result of malalignment and abnormal rotation of the caudal end of the developing fetus with distal curled end of the vertebral column being displaced to one side or the other. As a result either the cloaca or the mesonephric duct derivatives lie to one side of the vertebral column allowing one ureter to cross the midline and enter the opposite nephrogenic blastema or the kidney and ureter are transplanted to the opposite side of the midline during the normal renal ascent [10].

Ascent of kidney influences the development of the renal fascia layers and its planes. If there is some disturbance in this development, the looseness of the renal fascial tissue layers favours easy malposition of the intestines into the empty renal fossa while returning back into the abdominal cavity [11].

There has been reporting of a case of crossed fused ectopic kidney which was associated with a gastrointestinal anomaly of high caecum. Ultrasonography of this patient did not reveal the renal anomaly because the fused kidney on the right side appeared as a single mass mimicking a normal kidney and the displaced colon was mistaken for a normal kidney on the left. These anomalies and its associations can be better identified by using contrast roentgenography [12].

Renal anomalies may lead to malrotation of the intestines leading to change in position of the same. This may lead to errors in diagnosis such as appendicitis, diverticulitis and neoplasms of

the colon when the patient comes with history of lower abdominal pain. Renal anomalies in females such as pelvic kidney, crossed ectopia may lead to obstetric complication and hinder normal vaginal delivery [13].

CT angiography of 64 yr old male revealed ectopic kidneys with malrotation of the intestines. In another case crossed renal ectopia was observed in the fetus with polydactily. These may be asymptomatic and coincidental findings. Knowledge of these occurrences is essential as they may lead to complication and technical difficulties during surgical intervention [13, 14].

CONCLUSION

Crossed renal ectopia can be associated with malrotation of the gut. The rationale could be due to many factors such as the abnormal placement of the umbilical arteries, anomalies in ascent of kidney leading to laxity of the renal and extra peritoneal connective tissue layers and timing of the ascent of kidney before the gut rotation. Awareness of such anomalies could be due to incidental finding and helps the surgeons and radiologists in their diagnosis and planning and preventing postoperative complications.

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