A Rare Case of Crossed Renal Ectopia without Fusion

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Crossed renal ectopia without fusion is a rare congenital renal anomaly that occasionally presents with abdominal pain and abdominal mass. We present a 65 year old woman who presented with abdominal pain. Renal functions done were normal. Intravenous urogram done showed a crossed left ectopic kidney without fusion. Ultrasound done confirmed left crossed renal ectopia without fusion. The right kidney was normal in location.

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Introduction

Crossed renal ectopia is a congenital anomaly of the kidneys in which the kidney is located on the opposite side of the retroperitoneal space to its ureteric drainage into the bladder. It is an uncommon cause of abdominal pain and abdominal mass. They can be incidental findings in 20-30%. (1)

Case Presentation

A 65 year old para 3+1 known hypertensive and type II diabetic female patient who presented with right lower abdominal pain and palpable mass she had noticed for six months was referred to our unit for an intravenous urogram (IVU). Renal function tests done prior to the IVU were normal.

The scout film of the abdomen showed adequate preparation of the abdomen but while the outline of the right kidney was clearly visualised in the appropriate position, the left kidney was not seen in its normal position. There were no calculi or evidence of nephrocalcinosis. The nephrogram showed a normal sized right kidney in its normal location with a normal outline. No nephrogram was seen in the left renal region. Inferior to the right kidney there was another completely separate nephrogram of normal size and outline.

The ureter of the ectopic kidney inserts into the left base of the bladder
After 10 minutes. Arrow-right kidney. Bold arrow- ectopic left kidney

Left Kidney shows mild hydronephrosis

Normal right kidney

The two kidneys are seen separate from each other with a clear plane between them
Discussion

Crossed renal ectopia was first described in 1964 by Pannorulus. There are 4 types of crossed renal ectopia proposed by McDonald and McClellan; a) crossed renal ectopia with fusion, b) crossed renal ectopia without fusion, c) solitary crossed renal ectopia and d) bilaterally crossed renal ectopia.

It is a rare condition with an incidence of 1:2000 to 1:7000. There is a male predominance with a male to female incidence of 1.4:1. This condition is usually asymptomatic and diagnosed in the third decade. Patients may though present with abdominal pain, a palpable mass, haematuria, recurrent urinary tract infections and dysuria.

More commonly the left kidney will be located on the right side. Crossed renal ectopia may be fused (in 85%) or may be non-fused (less than 15%). The incidence of non-fused crossed renal ectopia is reported to be 1:75,000.

The blood supply of the ectopic kidney usually will arise from the ipsilateral side but occasionally may arise from the contralateral side. Aberrant arterial anatomy may be seen in both the normal kidney and the ectopic kidney. Crossed renal ectopia may occur with other congenital anomalies as part of the VACTREL complex of anomalies.

Common complications that occur are vesicoureteral reflux, recurrent urinary tract infections and renal calculi. Ureteroceles and renal vascular hypertension have also been reported. Diagnosis is commonly achieved by ultrasound, intravenous urogram (IVU), CT urogram and renal scintigraphy.

Embryology

There are three sets of kidneys in the embryo during development; pronephroi, mesonephroi and the metanephroi. In the fourth week, the pronephroi develop and their pronephric ducts open into the cloaca. These pronephric ducts are utilized by the next set of kidneys while the pronephroi degenerate. Late in the fourth week, the mesonephroi develop. These are interim functional kidneys and contain glomeruli and mesonephric tubules which open into mesonephric ducts. The mesonephric ducts then open into the cloaca. The metanephroi function for about 4 weeks prior to the formation of the permanent kidneys.

In the fifth week, the metanephroi develop and begin to function about 4 weeks later. The permanent kidneys develop from the metanephric diverticulum and the metanephrogenic blastema. The primordium of the ureter, renal pelvis, the calices and collecting tubes develop from the metanephric diverticulum. The metanephric diverticulum elongates and penetrates the metanephrogenicblastema. There is repeated branching of the straight collecting tubules to form the major calices and then the minor calices. The collecting tubules are then formed by the remaining generation of tubules. Invagination of glomeruli into the proximal ends of the tubules then occurs.

The process in which the permanent kidneys develop is known as reciprocal induction, whereby there is interaction of the metanephric diverticulum and the metanephrogenic blastema. Initially the kidneys are located in the pelvis ventral to the sacrum and in close proximity to each other. As the abdomen and pelvis grow, the kidneys appear as if the ascend up to their normal adult position. The kidneys also undergo rotation of almost 90 degrees medially during their ascent, thereby bringing the initially ventral hilum to be medially placed.

A likely cause of crossed renal ectopia has been postulated to be due to excessive bending and rotation of the caudal end of the developing embryo. This leads to the ureteric bud being unable to communicate with the more distal ipsilateral metanephros therefore the kidney is then
attracted to the closer contralateral side. The increase incidence of crossed renal ectopia in patients with scoliosis supports this theory.

Conclusion

Renal tract anomalies are a rare cause of sudden onset acute abdominal pain and should be considered as a differential diagnosis. It may be an incidental finding when investigating for other causes of acute abdomen. Although it is normally asymptomatic, complications may arise and treatment of these complications is necessary to prevent compromise renal function.

References