An Unusual Case of Non-Functioning Crossed Renal Ectopia Without Fusion

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Citation

Abstract
Crossed renal ectopia occurs either in fused or non-fused form. Only 10% are of the non-fused variety. The majority of these go unnoticed as they remain asymptomatic and are discovered incidentally. Very rarely they present with grossly hydronephrotic kidney. Our patient presented with a huge lump in the abdomen which was a grossly hydronephrotic ectopic kidney with multiple stones within it. We would like to report such an unusual case at our institute.

CASE HISTORY
A 55-years-old Hindu male residing in Mumbai came with chief complaints of pain in the right side of the abdomen and a huge lump in the right side of the abdomen since 3 months ago. The patient was asymptomatic before. There was no history of trauma or fever. He had a history of nausea and intermittent vomiting in between. There was no history of dysuria, pyuria, lithuria or hematuria. There was no bowel disturbance. The patient had no history of any major medical illnesses or any surgery done in the past. The patient had an addiction towards tobacco and betal nut.

On examination, the patient’s vital parameters were within normal limits. On per abdominal examination, there was a large swelling occupying the entire right lumbar and iliac fossae, measuring 20 x 15 x 7cm. The swelling was non-tender and not inflamed. It was ballotable. Hernia orifices were normal and so was the rest of the general examination.

On investigating, Hb was 15.1g%, WBC count 9300/cumm, serum creatinine 1.3mg%, urine routine was within normal limits. Ultrasonography of abdomen and pelvis showed crossed renal ectopia with the rest of the viscera being normal. The patient’s IVU showed crossed renal ectopia of the left kidney towards the right side below a normal right kidney. This ectopic kidney was grossly hydronephrotic, non-functioning and studded with multiple calculi. A Tc-99m DTPA scan was performed to assess exact kidney function. It showed a non-functioning left ectopic kidney with loss of the cortico-medullary ratio and a normally functioning right kidney.

A decision to perform open left nephrectomy was taken. At exploration, the following findings were confirmed:

- Grossly hydronephrotic ectopic kidney measuring 20 x 15 x 8cm.
- Paper-thin parenchyma of the ectopic kidney.
- Right kidney away from this ectopic one (non-fused) and within normal limits.
- Multiple adhesions on Gerota’s fascia over the ectopic kidney.
- Multiple renal calculi within the ectopic kidney.

A drain was put in the right paracolic gutter. Post exploration, the patient was asymptomatic and recovered well. He had minor wound infection, which was sutured after healing, and he was discharged after one week. At follow-up, the patient was asymptomatic.

At pathology, gross examination showed loss of the corticomedullary ratio, multiple renal calculi and minimal whitish parenchyma near the hilum.
Microscopy showed thyroidization of calyces and features of chronic calculous pyelonephritis.

DISCUSSION

When the kidney is located on the side opposite from which its ureter inserts into the urinary bladder, the condition is known as crossed renal ectopia. Of crossed ectopic kidneys, 90% are fused to their ipsilateral mate. Except for horseshoe anomaly, they account for the majority of fusion defects.

Fusion anomalies of the kidney were first categorized by Wilmer (1938), but McDonald and McClellan (1957)(1) refined and expanded this classification to include crossed ectopia with fusion, crossed ectopia without fusion, solitary crossed ectopia and bilaterally crossed ectopia.

The fusion anomalies have been designated as

- Unilateral fused kidney with inferior ectopia.
- Sigmoid or S-shaped.
- Lump or cake.
- L-shaped or tandem
- Disc, shield or doughnut, and
- Unilateral fused kidney with superior ectopia.

The first reported case of crossed ectopia was described by Pamarolus in 1654. A total of 62 patients with crossed ectopia without fusion have been reported (2). This constitutes 10% of all crossed ectopic kidneys. The anomaly occurs more commonly in males with a ratio of 2:1 and left-to-right ectopia is seen three times commoner than right-to-left ectopia.

The factor responsible for the change in kidney position during gestation is still undetermined (3), the reason for crossed renal ectopia is similarly uncertain. In 1938, Wilmer suggested that crossover occurs as a result of pressure from abnormally placed umbilical arteries that prevent cephalad migration of the renal unit, which then follows the path of least resistance to the opposite side.

Of crossed ectopic kidneys, when they are not fused, the unfused kidney usually resides in its normal dorsolumbar location and is oriented properly. The ectopic kidney is inferior with either diagonal or horizontal position with an anteriorly placed renal pelvis. A variable but definitive distance usually separates the two kidneys and each one is surrounded by its own Gerota’s fascia. In every case of crossed ectopia without fusion, the ureter from the normal kidney enters the bladder on the same side, whereas that of the ectopic kidney enters the bladder on the contralateral side.

In general, the occurrence of associated anomalies in crossed renal ectopia excluding solitary crossed ectopia is low. Most frequent are imperforate anus (4%), orthopedic anomalies (4%), and skeletal and septal cardiovascular defects.

Most of these patients do not have symptoms and are diagnosed incidentally. Gross hydronephrosis(4) and renal calculi (5) have been discovered in few patients, but uncommonly. Occasionally, an asymptomatic abdominal mass may be the only presenting symptom as seen in our patient.

For diagnosis, along with intravenous urography and ultrasonography of abdomen and pelvis, renal CT angiography is now considered as gold standard (6). However, a DTPA renal scan is a must when considering nephrectomy in non-functioning ectopic kidney. Prognosis is
usually good except when patients are having severe urinary tract infection and/or calculi.

SUMMARY
Our patient presented with crossed renal ectopia without fusion, with gross hydronephrotic non-functioning kidney, along with multiple renal calculi, with left-to-right ectopic pattern, and underwent nephrectomy.

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