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Malrotated Ectopic Kidney – A Case Report and Review of Literature

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Abstract **Case Report**

The term is derived from the Greek words ek ("out") and topos ("place") and literally means "out of place." Ectopic kidney is a congenital defect where kidney is in unusual anatomical position. An ectopic kidney is often associated with an increased incidence of stone formation as a result of stasis caused by the altered geometry of urinary drainage [1]. Factors which interfere with development such as the ureteric bud not meeting with the nephrogenicblastema for normal nephrogenisis or metanephric maternal disease, may result in abnormal migration of the kidney resulting in renal ectopia [2, 3]. Diseases in ectopic kidney may present diagnostic problems and unwary surgeon may be tempted to remove it as an unexplained mass [4]. Abnormal position may result in atypical referred pain, misdiagnosed as Appendicitis or Pelvic inflammatory disease in women. Increase in renal transplants, vascular reconstruction and other urologic procedures require thorough knowledge of morphological variants of kidneys and their clinical significance. Here we present a case ectopic kidney noted at our hospital.

Keywords: ectopic kidney, urinary tract anomalies, hydronephrosis, unascended kidney.

INTRODUCTION

During embryonic development, the fetal kidney first appears as buds inside the pelvis at 4th week, near the bladder. As the fetal kidneys develop, they climb gradually toward their normal position near the rib cage in the back by 8th week. The ascent of the kidneys precedes the descent of the gonads into the pelvis. Caudal growth in the embryo appears to assist immigration of the kidneys out of the pelvis into their eventual retroperitoneal location in the renal fossa. Sometimes, one of the kidneys fails to ascend and it may remain in the pelvis. Rarely does a child have two ectopic kidneys. Sometimes the kidneys ascend toward the rib cage, but one may cross over so that both kidneys are on the same side of the body.

Ectopic kidneys may be pelvic, iliac or abdominal, anywhere along the path of their usual ascent or contralateral, referred to as "crossed". There is strong correlation between renal ascension and vascularization. Renal ectopia is associated with anomalies in renal vessels [5]. Anomalies in rotation are seen both in normal and ectopic kidneys [6].

CASE PRESENTATION

A 23 year old male presented with severe right iliac fossa pain extending to the umbilical region since one day. He had no lower urinary tract symptoms. On palpation of abdomen there was a tender palpable mass of about 6x5 cms in Right Iliac fossa extending till umbilicus. His external genitalia were normal. Examination of the chest, cardiovascular and respiratory system had no abnormality. Patient was conservatively managed for pain. Investigations revealed normal Renal function. Ultrasound showed Right Ectopic Kidney with a 2mm stone in PUJ causing Mild hydronephrosis. CT confirmed the diagnosis. It had anteriorly-placed extra-renal pelvis. Patient improved with conservative management. Repeat USG and CT on day 3 revealed no stone in right PUJ. Patient discharged on day 4.

On CT hydronephrosis right iliac kidney was found (Figure-2). CT showed a calculi of size about 2mm in right renal pelvis. It had a dilated, anteriorly-placed extra-renal pelvis with a non-dilated ureter indicating a PUJ obstruction. He was treated conservatively and made an uneventful recovery.

No active urological intervention was needed at the time discharge since both the kidneys were functioning normally, no evidence of obstruction, infection or any complications.

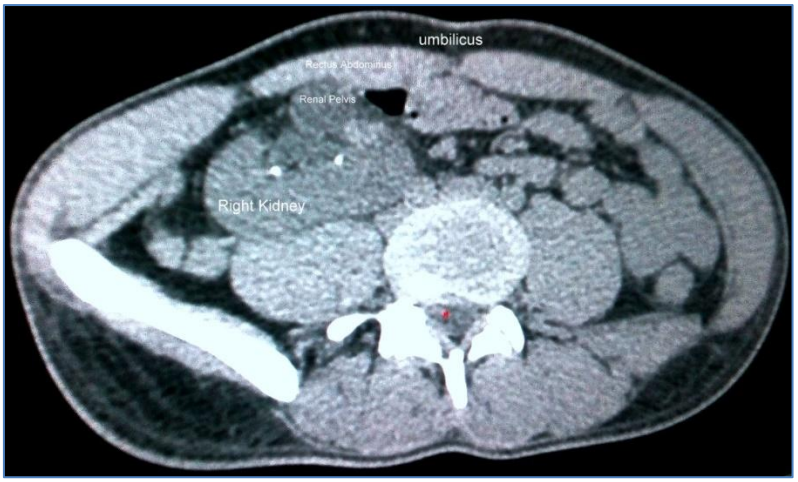

Fig-1: Ectopic Right Kidney with dilated pelvis day 1, of admission

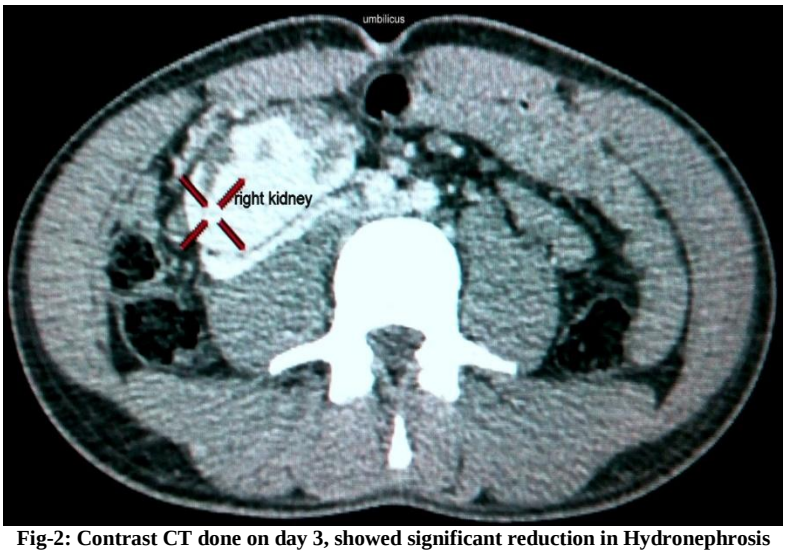

Fig-2: Contrast CT done on day 3, showed significant reduction in Hydronephrosis

Fig-3: Contrast CT of abdomen pelvis, with 3D reconstruction done on day 1

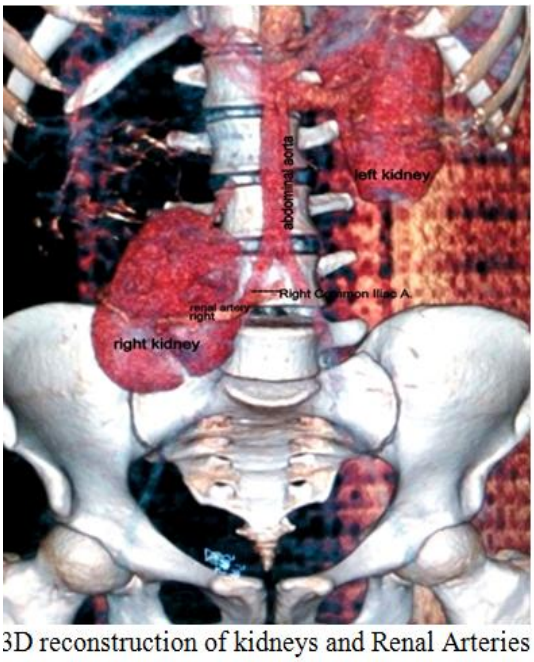

Fig-4: Contrast CT of abdomen pelvis, with 3D reconstruction done on day 1

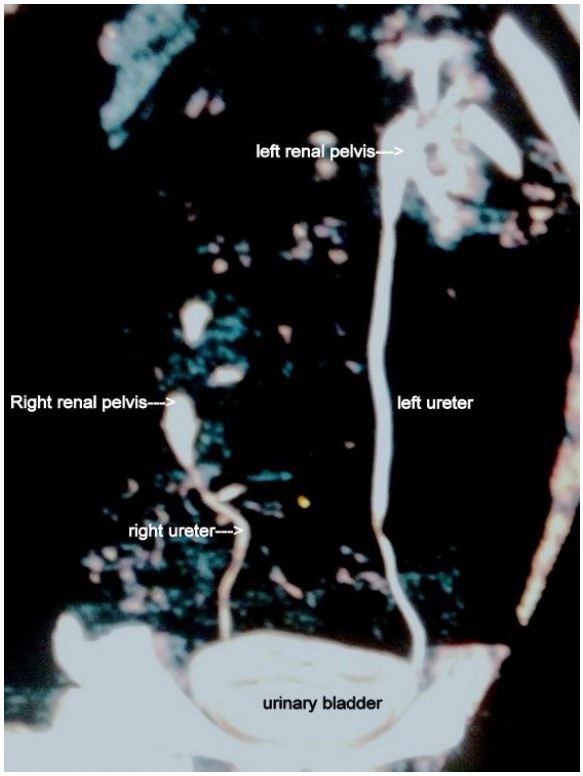

Fig-5: Anteriorly faced Right Renal Pelvis & Left side Normal

DISCUSSION

Ectopic kidney is described as abnormal position of a kidney due to a developmental anomaly and it occurs when migration of the kidney to its normal location during the embryonic period stops at a point. More than one anomaly can occur at the same time. While kidneys ascend through the pelvis, they receive new branches from vessels (iliac and aorta) close to them. When they reach the highest point, they receive new branches from the aorta, and the former branches degenerate. If those vessels do not degenerate in the ectopic caudal kidney, more than one accessory and polar arteries may arise [7, 8]. Hypertension is more frequently encountered in cases with more than one renal artery [9]. In our case Renal artery is arising from Common Iliac Artery.

Incidence of ectopic kidney reported in the literature is 1:500 to 1:1100 [9] Incidence of one normal and one pelvic kidney is 1:800 to 1:3000. [11]Ectopic thoracic kidney is 1:13000, Single pelvic kidney is 1:22000. Crossed renal ectopia 1:7000.

Various congenital anomalies with the urogenital system have been described. These are: multicystic dysplasia in a fused or unfused crossed kidney [12, 13], ureterocoele, patent urachus [14] hydronephrosis, ectopic ureteric orifice, vesicoureteric reflux, vaginal agenesis [15], hypospadias etc. Pelviureteric junction obstruction, as in the current case, has also been noted.

Anomalies of Rotation may be seen in normal or ectopic kidneys. Four rotational anomalies have been identified. Non Rotation-Renal Pelvis facing ventrally, Incomplete Rotation-Ventromedially, Reverse and Excessive Rotation-Hilum faces laterally. In our case it's incomplete rotation.

Urinary tract anomalies form a long list. Congenital anomalies of the urinary tract are often the underlying causes of pathologies. According to Guiterrez, 40% of pathologic conditions of the urinary system are due to these variations of number, position, shape and size or in rotation of kidney(s), calyces, ureter(s) or bladder. Usually these anomalies are associated with anomalies of the vertebral column, lower gastrointestinal tract, genital tract or spinal cord and meninges.

Even though it is not in its usual position, An ectopic kidney may not cause any symptoms and may function normally. Many people have an ectopic kidney and do not discover it until they have tests done for other reasons. Sometimes, a doctor may discover an ectopic kidney after feeling a lump in the abdomen during an examination. In other cases, an ectopic kidney may cause abdominal pain or urinary problems.

When a kidney is out of the normal position, drainage problems are likely. Sometimes, urine can even flow backwards from the bladder to the kidney, a problem called vesicoureteric reflux, or simply "reflux". Abnormal urine flow can set the stage for some of the problems associated with ectopic kidney:

1. Infection: Normally, urine flow prevents bacterial growth in kidneys and urinary tract. When a kidney is out of the normal position, urine may get trapped in the ureter or in the kidney itself. This gives bacteria a chance, to grow and spread. Symptoms of a urinary tract infection include frequent or painful urination, back or abdominal pain, fever, and chills. The urine may be cloudy or have an unusual odour.
2. Stones: Urinary stones form from substances found in the urine, such as calcium and oxalate. Urine that remains too long in the urinary tract increases the risk that these substances will have time to form stones. Symptoms of urinary stones include extreme pain in the back, side, or pelvis; blood in the urine; fever or chills; vomiting; and burning feeling during urination (dysuria). This was noted in our case, however didn't cause obstruction.
3. Renal failure: If urine backs up all the way to the kidneys, damage to the kidneys can occur. As a result, the kidneys can't filter wastes and extra fluid from the blood. Symptoms of kidney failure include swelling in the legs or abdomen, weakness, headaches, or nausea. If total renal failure occurs, a renal transplant will be necessary. Total renal failure happens only when both

kidneys are damaged. One ectopic kidney, even when it has no function, won't cause renal failure if there is additionally a normal kidney.

4. Trauma: If the ectopic kidney is in the lower abdomen or pelvis, as in the current case, it may be susceptible to injury from blunt trauma. People with ectopic kidney who want to participate in body contact sports may wish to wear protective gear.

Contrast-enhanced CT scan can clearly differentiate vascular and non-vascular structures and has remained the diagnostic method of choice [16]. Knowledge of the vasculature is important when operating on anomalously placed kidneys.

The treatment options vary with the presence of symptoms or complications. If the urinary function is normal with no evidence of urinary tract blockage, no treatment for ectopic kidney is needed. Monitoring of the patient's condition can be justified in case a change occurs. If IVU shows that obstruction is present, surgery may be need to correct the position of the kidney to allow for better drainage of urine. To correct reflux, the ureter is reimplanted into the bladder. If extensive renal damage has occurred, nephrectomy is indicated. As long as the other kidney is working properly, losing one kidney is not a problem. Fifty-six percent of ectopic kidneys have a hydronephrotic collecting system. Half of these cases result from obstruction at the ureteropelvic or the ureterovesical junction (70% and 30%, respectively), 25% from reflux grade III or greater, and 25% from the malrotation alone. Vesicoureteral reflux has been found in 30% of children with ectopic kidneys. Most ectopic kidneys are clinically asymptomatic, except in cases of associated ectopic ureter [20, 17]. The blood supply of an ectopickidney can vary. There can be more than one aberrant artery and aberrant arteries can originate fromthe abdominal aorta, common iliac artery, externaliliac artery or inferior mesenteric artery [18].

Prognosis: The ectopic kidney is no more susceptible to disease than the normally positioned kidney, except for the development of hydronephrosis or urinary calculus formation [20, 21]. This may be due to the anteriorly placed pelvis and malrotation of the kidney, which may lead to impaired drainage of urine from a high insertion of the ureter to the pelvis or an anomalous vasculature that partially obstructs one of the major calyces or the upper ureter. In addition, there may be an increased risk of injury from blunt abdominal trauma, because the low-lying kidney is not protected by the rib cage. Renovascular hypertension secondary to an anomalous blood supply has been reported, but a higher-than-normal incidence is yet to be proved. A recent study by van den Bosch and colleagues 2010 examined the urologic and nephrologic consequences of both simple and crossed renal ectopia. They found no adverse effects on blood pressure or kidney function during childhood [19].

CONCLUSION

Ectopic kidney is a congenital anomaly where the kidney is in abnormal anatomical position and tend to retain vascularity from nearest vessel. Asymptomatic, non-complicated cases are managed conservatively but percutaneous nephrolithotomy or flexible uretrorenoscopy is necessary if there are complications such as recurrent stones and failure of conservative line management. Nephrectomy is considered only when poorly functioning kidney with recurrent stone formation and recurrent infection. Thorough understanding of morphological variants of kidneys plays an important role in kidney transplantation, percutaneous nephrolithotomy and other surgical procedures.

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