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Case Report

Percutaneous nephrolithotripsy for renal pelvis stone in a crossed fused ectopic kidney: a case report

Rajeev Sarpal*, Tarun Chaudhary, Manoj Biswas, Shikhar Agarwal, Ankur Mittal

Himalayan Institute of Medical Sciences, SRH University, Jolly Grant, Dehradun, Uttarakhand, India

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***Correspondence:**
Dr. Rajeev Sarpal,
E-mail: rajeev_sarpal@hotmail.com

ABSTRACT

Crossed renal ectopia is the second most common fusion anomaly of the kidney after horseshoe kidney. The incidence of both fused and unfused cases is 1 in 7000 in autopsies. Percutaneous nephrolithotripsy (PCNL) is a well-established technique in the surgical management of nephrolithiasis. The conventional fluoroscopic guidance of PCNL will be of limited value in ectopic anomalous kidneys due to the abnormal anatomical landmarks with consequent compromise of the procedure's safety. A 30-year old male patient, previously healthy, presented to our urology outpatient clinic complaining of dull flank pain of six month duration with tenderness in the right renal area and an enlarged right kidney and 1 episode of gross haematuria. Radiological investigations showed left side crossed ectopia with nephrolithiasis. Patient was managed successfully by fluoroscopic guided percutaneous nephrolithotripsy. The position of the stone-containing ectopic kidney can make it easy to reach the pelvis of the target kidney without injuring any adjoining structure. The superimposition of the different soft tissue densities in the radiologic view may limit the ability of the operator to distinguish between different tissue identities. Laparoscopic guidance can represent as a practical solution to this technical problem.

Keywords: Crossed renal ectopia, Nephrolithiasis, Percutaneous nephroltihotripsy, Hematuria

INTRODUCTION

Crossed renal ectopia is the second most common fusion anomaly of the kidney after horseshoe kidney. The incidence of both fused and unfused cases is 1 in 7000 in autopsies. Abeshouse and Bhistkul estimated its occurrence at 1 in 1000 live births.[1] The unfused crossed renal ectopia is much rare with an incidence of 1/75000 in autopsies.[2] It consists of transposition of a kidney generally with left-to-right crossover. Ninety percent of ectopic kidneys are fused to their ipsilateral mate and the ureter of the ectopic kidney inserts into its normal position in the bladder at the contralateral side.[3] The mechanism of failure of ascent and complete rotation is also not clear. Poor development of a kidney bud, a defect in the kidney tissue responsible for prompting the kidney to move to its usual position, genetic anomalies and exposure to teratogen drugs is thought to be the cause of this anomaly.[4]

Pamarolus defined the first case in 1654. In 1957 McDonald and McClellan described the types of crossed renal ectopia (i) crossed renal ectopia with fusion, (ii) crossed renal ectopia without fusion, (iii) solitary crossed renal ectopia, (iv) bilaterally crossed renal ectopia. The anomaly is generally diagnosed at paediatric age group during the investigation of accompanying malformations.[4,5] Possible complications are listed as infection, kidney damage due to reflux of urine, trauma and nephrolithiasis. Here the authors are presenting a male patient, with a right sided unfused crossed renal ectopia anomaly without any complaints until this last emergency department admission. Their case is worth to

report for the demonstration of the possible benign nature of this type of anomaly.

The urinary system can suffer from different congenital anomalies like renal agenesis, multiple kidneys, renal ectopia, and fusion defects. Procedural difficulty and compromised safety are expected when surgical intervention is indicated in patients with anomalous kidneys. The abnormal renal anatomy may contraindicate certain interventions or require modification of their classical procedural aspects.

Percutaneous nephrolithotripsy (PCNL) is a well-established technique in the surgical management of nephrolithiasis. The conventional fluoroscopic guidance of PCNL will be of limited value in ectopic anomalous kidneys due to the abnormal anatomical landmarks with consequent compromise of the procedure's safety. In this group of patients, laparoscopic assistance will provide direct visual guidance through the abnormal tissues during the percutaneous technique.

CASE REPORT

A 30-year old male patient, previously healthy, presented to our urology outpatient clinic complaining of dull flank pain of six month duration. The patient was a healthy looking male. His abdominal examination revealed tenderness in the right renal area and an enlarged right kidney could be palpated. There was no tenderness or mass in the left renal area. It was associated with 1 episode of gross haematuria. A kidney, ureter, and bladder (KUB) X-ray was ordered and showed a 2x3 cm radio-opaque shadow in the right upper quadrant zone, consistent in density with a stone. The right kidney was laterally and superiorly placed from its expected location close to lumbar spine. In addition, no normal soft tissue shadow of left kidney was seen in left renal area (Figure 1).

Figure 1: X-Ray (KUB) showing a 2x3 cm radio-opaque shadow in the right upper quadrant zone, consistent in density with a stone.

His lumbar spine was normal in appearance and there was no scoliosis or any spinal dysraphism and showed the opacity to be at the level of third lumbar vertebra (Figure 2). A reconstructed contrast enhanced urinary tract CT was done and showed left ureter crossing the midline to the right side and reaching the renal pelvis of the ectopic kidney which contained the stone. Both ureters were seen in the right paramedian zone down to the lower border of L5 where the left ureter starts crossing leftwards to enter the bladder normally. Both kidneys were fused together with the native right one lying slightly anterior (Figure 3).

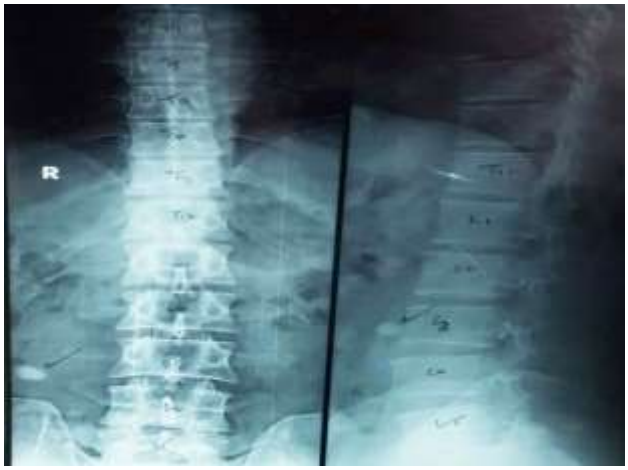

Figure 2: X-Ray lumbar spine showing opacity at the level of third lumbar vertebra.

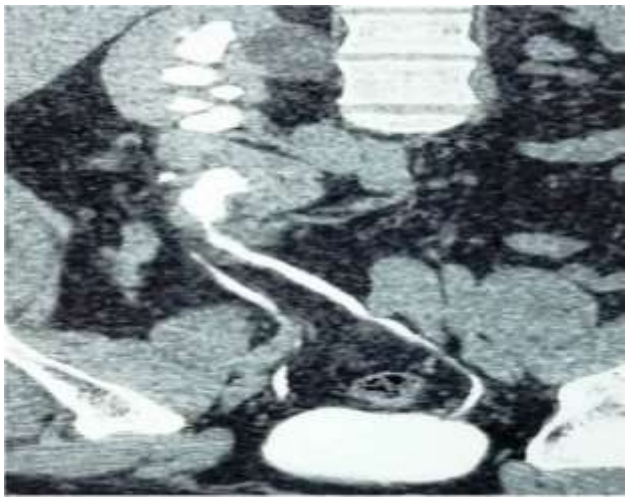

Figure 3: CECT showing left ureter crossing the midline to the right side and reaching the renal pelvis of the ectopic kidney.

The anatomic abnormality of the ectopic kidney was explained to the patient in addition to the technical difficulty imposed by it. A plan was formulated to do PCNL. The patient's consent was obtained and he was admitted for the procedure. After the induction of general anaesthesia with muscle relaxation and endotracheal intubation, the patient was put first in the lithotomy

position. Cystoscopy showed that both ureteric orifices were in their normal anatomical site in the bladder. A left access ureteric catheter was inserted over a guide wire (Teromo's 0.025'') under fluoroscopic guidance and vision. The patient was then positioned prone. Upper calyceal puncture was made under fluoroscopic guidance by using 'Bull's Eye technique (Figure 4). The creation of the PCNL sheath tract was then done under fluoroscopic guidance using serial metallic dilators (Figure 5) and stone was fragmented. All stone fragments were removed and an antegrade double J stent was placed, a Nelaton's catheter was placed on nephrostomy. The procedure took about one and half hour. At the completion of the surgery, the patient was extubated in the supine position and sent to the post-anaesthesia care unit. After a smooth one hour postoperative course with a mild to no pain score, he was discharged to the ward. The nephrostomy tube was removed in the ward after 48 hours. The patient was discharged home on the postoperative day 3. His outpatient follow-up was uneventful. Post-operative X-ray and ultrasound showed complete clearance (Figure 6).

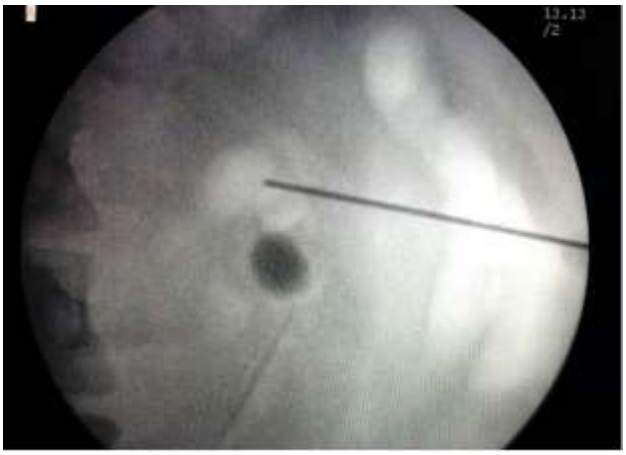

Figure 4: Upper calyceal puncture under fluoroscopic guidance by using 'Bull's Eye technique'.

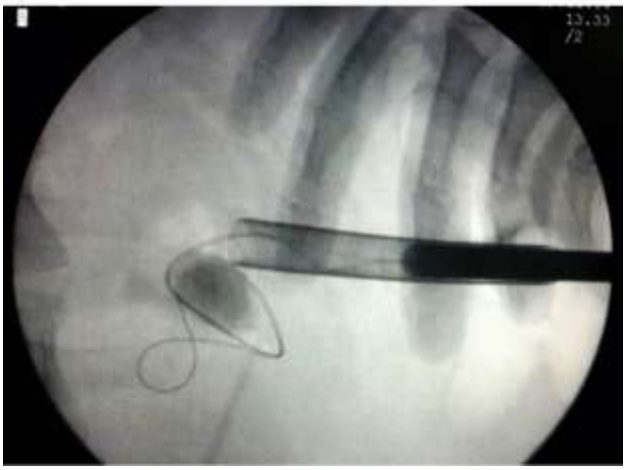

Figure 5: PCNL sheath tract made under fluoroscopic guidance.

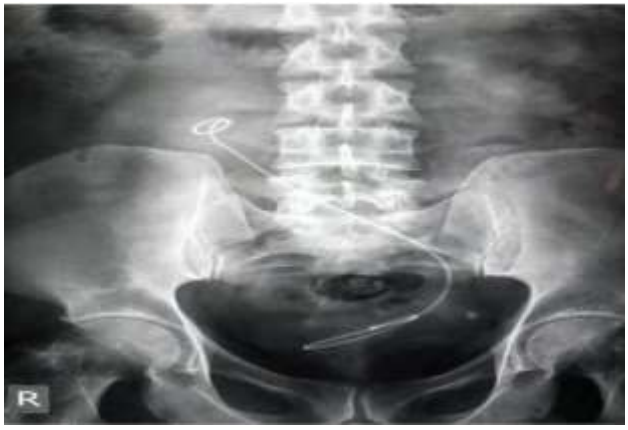

Figure 6: Post-operative X-ray showing complete clearance of stone.

DISCUSSION

During foetal development, the urinary bladder enters the metanephric blastema adjacent to the anlage of the lumbosacral spine. During the next 4 weeks, the developing kidney comes to lie at the level of the L1-L3 vertebrae. Because the mechanisms responsible for normal complete ascent of the kidney during gestation are unknown, the cause of crossed renal ectopia is also unknown. It is 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 part of least resistance to the opposite side.[4]

There are different theories about the phenomenon. First theory is that crossed ectopia is strictly a ureteral phenomenon, with the developing urinary bladder wandering to the opposite side and inducing differentiation of the contralateral nephrogenic anlage. Some authors supported the opinion of the strong but undetermined forces are responsible for renal ascent. According to them these forces attract one or both kidneys to their final place on the opposite side of the midline. Another point of view is that the crossover is the result of the malalignment and abnormal rotation of the caudal end of the developing foetus, with the distal curled end of the vertebral column being displaced to one side or the other. As a result, either the cloaca lies 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 normal renal ascent.[4] It is important to note that renal ectopia is frequently associated with congenital anomalies of other organ systems. Genetic factors may also play a role.[6,7]

An ectopic kidney may not cause any symptom and may function normally, even though it is not in its usual position. Possible complications include vesicoureteral reflux, infection, stone, and kidney damage. Signs and symptoms include vague lower abdominal pain,

haematuria, fever, urinary tract infection, hypertension, renal failure, and a palpable abdominal mass.[8,9]

On ultrasonograms, overlying abdominal gas may obscure a portion of the fused kidneys, making precise diagnosis difficult. This diagnosis should be considered when two separate kidneys cannot be identified. The sonographic appearance of this entity consists of a characteristic anterior and/or posterior notch, difference in orientation of the collecting systems in the fused kidneys, and absence of a kidney in the contralateral renal fossa, or elsewhere in the body, such as the pelvis.[10]

Review of literature showed only 6 cases of stone disease treated in crossed renal ectopic kidneys.[11]

The altered geometry of the urine drainage system in CFRE can lead to urinary stasis which favours the development of renal stones.[12] In a study from Japan, 15 of 166 cases of crossed renal ectopia were associated with urinary tract stones (Iwasaki 1988).

Thorough preoperative radiologic investigation of the renal system is recommended in the surgical management of these cases. This includes the use of CT scanning, intravenous pyelography, and angiographic studies. Good patient selection is also important for the outcome of laparoscopic assistance in these patients. Those with history of extensive prior abdominal surgeries are not good candidates due to the laparoscopy-hostile peritoneal environment present in these patients.[13]

Various interventional modalities, including open surgery, percutaneous procedures, extracorporeal shock wave lithotripsy (ESWL), and ureteroscopy, have been used in the management of nephrolithiasis in anomalous kidneys. For stones smaller than 2 cm in diameter, ESWL is currently the preferred method of treatment. For lager stones and for those ones resistant to ESWL, percutaneous nephrolithotomy-if applicable-is the method of choice.[13] PCNL offers advantages over open surgical approaches in terms of the lesser degrees of bleeding, post-operative pain, and consequently a shorter hospital stay. Scarring of the kidney will also be less due to a lesser degree of trauma.

The renal access difficulty due to complex renal configurations imparted by the anomalous renal anatomy will make the management of nephrolithiasis difficult and challenging to the urologist. Fluoroscopic guidance for the insertion of equipment of PCNL is likely to be of limited value because anatomically abnormal radiolucent tissues may interpose between the skin and the target location of the lithotome leading to tissue injury during the insertion of the access needle. These tissues can be of vital importance like blood vessels and renal tissue, depending on the merits of the case. In our patient, the position of the stone-containing ectopic kidney made it easy to reach the pelvis of the target kidney without injuring any adjoining structure.

The superimposition of the different soft tissue densities in the radiologic view may limit the ability of the operator to distinguish between different tissue identities. Laparoscopic guidance can represent as a practical solution to this technical problem. The leading part of the insertion needle in PCNL can be guided a traumatically under vision into the target kidney in a safe way. The added physiological aberrations imposed by laparoscopy are likely to be minimal and of no clinical significance since the laparoscopic intervention serves a quick purpose and is short lived.

CONCLUSION

The renal access difficulty due to complex renal configurations imparted by the anomalous renal anatomy will make the management of nephrolithiasis difficult and challenging to the urologist. The position of the stone-containing ectopic kidney can make it easy to reach the pelvis of the target kidney without injuring any adjoining structure.

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