CASE REPORT

Renal Calculus in an Ectopic Pelvic Kidney – A Case Report and Review of Literature

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Pelvic kidneys are anatomical abnormalities that occur when the kidney does not rise from the pelvis during embryogenesis. The majority of cases are asymptomatic, though they are associated with higher risks for traumatic injury, infections, renal calculi, and other urological issues.

Because of its advantages of flexion and deflection, retrograde intrarenal surgery (RIRS) employing flexible ureterorenoscopy (fURS) is an alternative treatment method for small- to medium-sized calculi in anatomically aberrant kidneys. Presented here is a case of a 43-year-old male with a renal stone in a pelvic left kidney with the ureter crossing the midlineand is located at the prevertebral region at the level of L4 to S1.

Key words: pelvic kidney, ectopic kidney, retrograde intrarenal surgery, flexible ureteroscopy

Introduction

Congenital renal anomalies are among the most common birth deformities, exceeded only by cardiac and skeletal defects.[1] An ectopic kidney's vascular supply is not consistent, and may receive vascular access from a range of vessels as the fetal blood supply can be retained. Multiple vascular sources may supply the ectopic kidney; the iliac arteries, direct branches from the aorta, mid sacral vessels, or the hypogastric arteries have all been found supplying ectopic kidneys. Surgery on an ectopic kidney requires thorough knowledge of this anatomy.[2]

Ectopic kidneys are also associated with several other congenital abnormalities. In females, this may be in the pelvis, such as Mullerian agenesis or unicornuate uterus. Ectopic kidneys can be a feature of multisystem congenital syndromes such as CHARGE syndrome (coloboma, heart disease, atresia choanae, retarded growth, genital hypoplasia, and ear abnormalities) or VACTERL malformations (vertebral, anal, cardiac, tracheal, esophageal, renal, and limb anomalies).[3] With an ectopic kidney, the kidney may become malrotated or the ureter may cross the midline as it approaches the kidneys. This may prove a challenge on performing retrograde ureteroscopic procedures such as diagnostic ureteroscopy and retrograde intrarenal surgery. The aim of this report is to present a case of a pelvolithiasis on a pelvic kidney with ureter crossing the midline that underwent retrograde intrarenal surgery as well as the challenges the surgeons encountered as well as tips on how to successfully clear the stone.

The Case

A 43-year-old male initially presented with periumbilical pain radiating to the epigastric area, described as colicky, dull, associated with vomiting episodes. Consult with private physician was done, wherein he was managed as a case of Acid Peptic Disease (APD) and was prescribed Omeprazole 40mg/tab 1 tab once daily. However, progression of symptoms and intermittent low-grade fever were noted. Work-up included urinalysis which showed microscopic hematuria. Serum Creatinine was within normal limits, and WBC count was slightly elevated (12.3).

Non-contrast Enhanced Helical CT Scan showed a left pelvic kidney located at the prevertebral region, at the level of L4 to S1, with a lithiasis in the interpolar/pelvic region measuring 0.7cm x 1.3cm x 1.3cm (AP x T x CC), 550HU, Moderate dilatation of the upper and mid calyces. The patient was then scheduled for elective definitive surgical intervention, and underwent Retrograde Intrarenal Surgery (RIRS). Intra-operatively, retrograde pyelography was performed and an S shaped ureter was noted from the left ureterovesical junction insertion crossing the midline then back to the ipsilateral renal pelvis at the pelvic kidneys (Figure 3). A 0.35 Sensor wire was placed and an F11 ureteral access sheath placement was attempted. With the configuration of the ureter, the F7.5 flexible ureteroscope was inserted through the guidewire under C-arm guidance. Since the stone migrated to the inferior pole, there was diffulty in the visualization and access of stone. The flexible scope was fully deflected and lithotripsy was performed. The procedure took around 2.5 hours for both fragmentation and dusting of the stone. Some less than 5 mm stone fragments were left for medical dissolution and expulsion. The patient tolerated the procedure well, and postoperative course was uneventful.

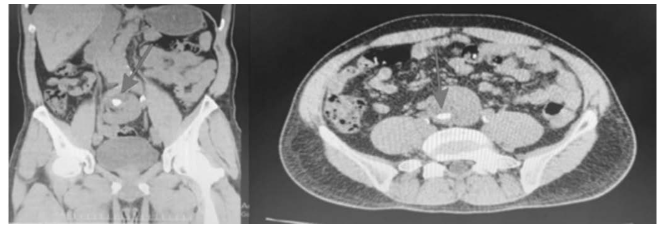

Figure 1. Non-contrast Enhanced Contrast Helical CT scan
Left: Coronal view. Right: Axial view. Arrows indicate the lithiasis.

Figure 2. Intraoperative findings – Flexible Ureteroscopy
Left: Guidewire inserted into the left ureteral orifice.
Middle: Fragmentation of the nephrolithiasis using LASER lithotripsy. Right: Post-fragmentation.

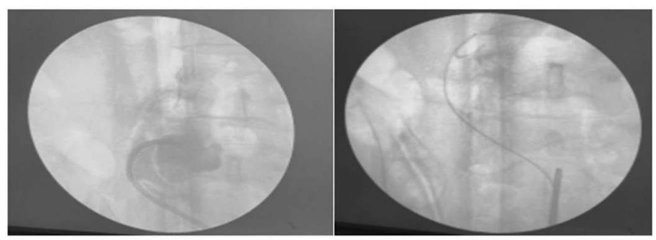

Figure 3. Intraoperative findings - Fluoroscopy
Left: Retrograde pyelography with tip of fURS within the renal pelvis
Right: Ureteral stent placement with coiling of the distal tip noted

Discussion

The kidney develops between weeks 6 and 8 after conception, and the embryologic kidney rises from the pelvis into the lumbar region in the 9th week. If the kidney fails to pass above the fork of the umbilical arteries, the blood supply degenerates, or there are other factors inhibiting renal migration, then the kidney fails to rise to its normal anatomical location and instead becomes ectopic. The exact location can be varied, with most cases being in the contralateral pelvis, but in the cases of crossed renal ectopia, both kidneys can be on the same side of the spine or, more rarely, the kidney can be outside the pelvis or retroperitoneal space entirely and even become located within the thorax. Ectopic renal units are usually a unilateral condition, but there are documented cases of bilateral ectopic kidneys.[4]

The incidence quoted is variable worldwide but is often approximately 1 in 1000 births.[4] A retrospective study of 13,701 antenatal scans in Turkey found an incidence of pelvic kidneys of 1 in

571, although this study only included scans with a normal amniotic fluid volume.[4] A Taiwanese study screening 132,000 school children found a lower incidence of only 1 in 5000, but it is thought this may have underestimated the actual incidence due to the screening method used.[5] An ectopic, pelvic kidney is the most likely finding in a fetus where the prenatal ultrasound finds an absent or missing renal fossa, but the amniotic fluid is normal.[6]

Even in asymptomatic patients, the ectopic kidney often has reduced function relative to the contralateral kidney. Most patients with ectopic kidneys are asymptomatic, and if recognized at all, the diagnosis tends to be an incidental finding while investigating other pathology or on routine antenatal ultrasonography. However, urinary tract complications can develop, and patients may present with a range of pathologies, including an increased incidence of urinary tract infections, ureteropelvic junction obstruction in the ectopic kidney, or increased risk of renal calculi. The most common associated abnormality is vesicoureteral reflux, which occurs in 30% of patients with simple renal ectopia.[6]

Patients with ectopic pelvic kidneys are more likely to develop urolithiasis compared with the normal population. This is linked to impaired drainage of the kidney due to altered anatomy, as well as metabolic reasons. Therefore, treatment of pelvic kidney stones remains challenging for the urologist because of the structural and architectural anomalies of such kidneys. The exact incidence of renal calculi in pelvic kidneys is unknown, but is thought to be higher than the general population due to altered anatomy and impaired urinary flow rates. The altered anatomy does have significant implications in the operative management of renal calculi, and the risk of vascular injury is increased relative to the general population.[7]

Extracorporeal Shockwave Lithotripsy (ESWL) in the prone position was first recognized as a non-invasive method for treatment of patients with pelvic kidney stones in 1988. Several studies have indicated that ESWL can be recommended as a first-line treatment option for anomalous kidney stones because of the high rates of stone clearance. However, Demirkesen et al. reported that ESWL for normal kidneys had a higher stone-free rate than that for aberrant kidneys (78% vs. 56%, respectively) and that aberrant kidneys had a higher rate of clinically insignificant residual fragments than did normal kidneys (37% vs. 18.5%, respectively).[8] Consequently, ESWL was been viewed as the first-choice non-invasive treatment modality with a relatively poor success rate for pelvic kidney stones.[9]

Flexible ureteroscopy has emerged as an alternative treatment method for small- to medium-sized renal calculi because of its advantages of flexion and deflection. Several studies have shown that the success rates of flexible ureteroscopy in patients with pelvic kidneys range from 75.0% to 84.7%.[10] Nonetheless, anatomic alterations including a tortuous ureter and malrotated kidney have been suggested as significant factors that increase the difficulty of the procedure and influence the stone clearance rate in treatment by flexible ureteroscopy. The use of a ureteral access sheath has been noted to assist in performing the procedure efficiently and safely.[11]

Other treatment options for stone disease in ectopic kidneys include Pyelolithotomy [open vs laparoscopic (transperitoneal vs retroperitoneal vs trans-mesenteric) vs robot-assisted], as well as Percutaneous Nephrolithotomy (PCNL). Although some authors have suggested the use of ultrasound or computed tomography guidance to achieve percutaneous access in patients with pelvic kidney stones, there is still a high risk of injuring the surrounding viscera and major vessels.[12]

A 11/13Fr ureteral access sheath was used for this patient, and flexible renoscopy was done under C-arm guidance. Stone fragmentation was done using Boston Scientific® Auriga™XL LASER Lithotriptor using Dusting method (high frequency, low energy, low pulse duration). A 6Fr x 24cm ureteral stent was placed post-procedure. Indwelling urethral catheter was removed first post-operative day, and patient was discharged on the second post-operative day. Imaging on follow-up one month post-procedure showed no lithiasis, and DJ stent was subsequently removed.

Conclusion

Although fURS in patients with anomalous kidneys can be technically challenging, advancements in endourological techniques have made it a safe and effective procedure. In these patients, the stone-free rates are good with a low risk of major complications.

Patients with stone disease in anomalous kidneys need individualized management and probably should involve an interdisciplinary treatment with interventional radiology colleagues with interventions carried out in high-volume endourology centers. Although randomized trials between treatment modalities would be difficult given the rarity of this condition, perhaps large prospective multicentric studies with long-term follow-up and standardized references would be able to provide with high-quality insightful data.

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