Abstract:
Ectopic kidney is a rare congenital abnormality and is usually asymptomatic and found incidentally. Giant hydronephrosis is also an uncommon lesion in children and adolescents and is usually due to ureteropelvic junction obstruction. We report a case of 14-year-old male presenting with abdominal fullness, suprapubic pain and decreased urine output with biochemical evidence of renal impairment. The findings from the radiologic investigations were suggestive of an ectopic pelvic kidney with giant hydronephrosis due to ureteropelvic obstruction causing contralateral obstruction to the kidney and ureter - an extremely rare presentation.
INTRODUCTION: An ectopic kidney is a rare congenital developmental anomaly wherein there is the failure of the kidneys to ascend and migrate to its normal renal location. In the embryogenesis of the kidneys, cranial migration of the embryonic metanephrons occurs during the second month of gestation. It may become arrested at any site above its original point in the pelvis (1). Given the rarity of a pelvic kidney, it often is an unsuspected finding in patients presenting with vague or atypical abdominal symptoms. Only on further evaluation with radiographic examination is an ectopic kidney identified as the cause. Because of structural and architectural anomalies that can accompany a pelvic kidney, conditions such as chronic obstruction and nephrolithiasis are common (4). The variation in anatomy of a pelvic kidney creates anomalous vascular patterns and altered spatial relations with adjacent pelvic organs. CASE REPORT: 14 years old male patient presented to the emergency department with 4 days history of lower abdominal swelling and suprapubic pain with anuria for the past 8 hours. His medical and surgical histories were unremarkable and he was not any medications. A physical examination revealed dull suprapubic mass arising from the pelvis and extending into right lumbar region and just above umbilicus with mild tenderness. Blood investigations revealed elevated blood urea (65 mg %) and serum creatinine level (4.7mg %). Abdominal sonography revealed right ectopic pelvic kidney with grossly dilated pelvicalyceal system occupying whole of the pelvis with thinned out cortex. Left kidney normal in size and location with moderate hydroureteronephrosis. CT KUB plain confirmed these findings. It was postulated that ectopic right kidney was causing obstruction of the left ureter. On ureteral catheterisation only 30 ml of urine drained.
A nephrostomy tube was inserted into the right pelvic ectopic kidney under ultrasonographic guidance and 2.7 litres of straw coloured fluid drained. On drainage of the right ectopic kidney the pain settled, and satisfactory amounts of urine drained from the urethral catheter; his renal function improved (serum creatinine 0.8mg %). A nephrostogram revealed marked dilatation of the right with abrupt narrowing of PUJ. Delayed film showed no contrast entry into the ureter. The left kidney was normal on renography but the right pelvic kidney showed reduced function with poor excretion (GFR: 10ml/min). Assessment of the creatinine clearance of the PCN fluid done showed the creatinine clearance of 8ml/min. pelvicalyceal system. The right ectopic kidney was then explored: at operation, the right kidney was in the pelvis and was grossly hydronephrotic with minimal renal cortex and narrowing at the PUJ. There was no other intra-abdominal pathology and a right nephrectomy done. The patient had an uneventful recovery and was discharged after 7 days. At 2 months follow-up ultrasonography confirmed a normal left kidney, the serum creatinine level was within normal limits.

DISCUSSION: Ectopic kidneys were described as early as the century by anatomists and may be found in a variety of locations, i.e. pelvic, iliac, abdominal, thoracic, contra lateral or crossed. The actual incidence varies among autopsy series from 1 in 500 to 1 in 1200 but average occurrence is about 1 in 900. There is no apparent difference between the rates in male and females at autopsy but, because of the higher frequency of uroradiological investigation in females, ectopia has been more readily recognised in them. Left-sided ectopia is reported slightly more than the right and the incidence of pelvic ectopia is estimated at 1 in 2100–3000 at autopsy (1). Most ectopic kidneys are asymptomatic and are discovered incidentally.

The present case appears to have been an acute presentation of congenital abnormality, which had otherwise been of no concern to the patient until presenting with oliguria, loin pain and biochemical evidence of renal impairment (3). An ectopic kidney is more susceptible to the development of hydronephrosis or urinary calculus formation than a normally positioned kidney. 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 (4). Because of its positioning it is more prone to blunt abdominal trauma. There are five reported cases where a pelvic ectopic kidney was removed as it was thought to represent a pelvic malignancy, with disastrous results. With the availability of modern imaging, a pelvic ectopic kidney should be more readily diagnosed from amongst the differential diagnoses of a pelvic mass. Treatment is mainly based on the functional capacity of the kidney; nephrectomy being done on non-functional kidneys and corrective procedures forming the mainline of treatment for the functional kidneys. This is an unusual and interesting set of circumstances and when faced with a complex situation such as this, the input of radiological colleagues is of great value (2).

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