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CASE SERIES ILLUSTRATING SPECTRUM OF RENAL ANOMALIES IN CONGENITAL ANOMALIES OF KIDNEY AND URINARY TRACT (CAKUT)

Radio-Diagnosis

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ABSTRACT

Congenital anomalies of kidneys and urinary tracts (CAKUT) are disorders caused by defects in development of kidneys and their outflow tracts. The formation of kidneys begins at 3 weeks of gestation and nephrogenesis continues until 36 weeks. In severe cases of CAKUT, when the kidneys do not form, the fetus will not survive. However, in less severe cases, the baby can survive with combined kidney and outflow tract defects or they may only be identified in adulthood (1). CAKUT occur in 3-6 per 1000 live births and is the leading cause of end-stage renal disease (ESRD) in children and also cause subsequent renal problems in adulthood like stone formation, infection, hypertension, and renal failure (2). CAKUT may be rarely associated with syndromes. This case series present the imaging findings in various modalities mainly CT urography in evaluation of renal anomalies.

KEYWORDS

Horseshoe Kidneys, Pancake Kidneys, Ectopic, Malrotation, Duplication Of Ureters.

INTRODUCTION:

Ultrasonography (US) is usually the first imaging performed as it is easily available, non-invasive and radiation free. Computed tomography (CT) and magnetic resonance imaging (MRI) are useful to confirm the USG detected abnormality, characterization of complex malformations, demonstration of collecting system and vascular anatomy and for early detection of complications like renal calculi, infection and malignancies (3).

Non-contrast CT can detect renal stones and nephrocalcinosis. CT urography is useful in duplex system, their complex course, distal opening and associated other genitourinary malformations. MRI has the advantages of being non-ionizing form of imaging, better soft tissue contrast and detection of collecting system abnormalities however disadvantage is requirement of sedation in small children, cost and availability (4)(5)(6).

CLASSIFICATION:

Renal anomalies can be broadly classified in the following manner:

1) Structural which includes renal agenesis, renal dysplasia, renal hypoplasia, supernumery kidneys.
2) Renal fusion anomalies which includes cross fused renal ectopia, horseshoe kidney, pancake kidney.
3) Renal position which includes renal malrotation, ectopic kidney, crossed renal ectopia.

Ureteric anomalies which includes vesico-ureteric reflux, ureteropelvic junction obstruction, congenital megaureter, duplex collecting system, ectopic ureter.

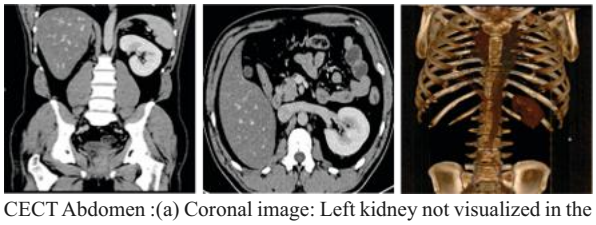
Lower urinary tract anomalies which includes bladder exstrophy, bladder agenesis, posterior urethral valves.

CASE SERIES:

Renal agenesis:

Complete absence of one or both kidneys indicates renal agenesis. Bilateral agenesis is incompatible with life and is associated with pulmonary hypoplasia and limb defects. It is often asymptomatic with compensatory hypertrophy of other kidney. Unilateral renal agenesis is mostly incidentally detected in adults in US or CT performed for other reasons. Associated abnormalities can be seen like ipsilateral seminal vesicle cyst, ipsilateral absence of seminal vesicle in males or mullerian abnormalities in females (7)(8).

Case 1 : A 11-year-old boy presented with recurrent urinary tract infections and poor weight gain. No h/o fever or hematuria at present. No significant past surgical history. Clinical evaluation advised imaging. CECT abdomen was taken.

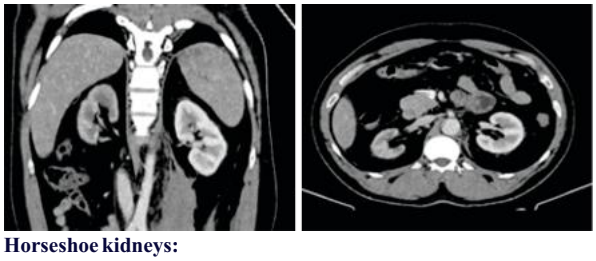


CECT Abdomen :(a) Coronal image: Left kidney not visualized in the renal fossa. (b) Axial image: Confirms absence of left kidney with compensatory hypertrophy of the right kidney.

Renal hypoplasia:

Renal hypoplasia is a congenital condition in which the kidney is smaller than normal but has structurally normal parenchyma, with reduced numbers of calyces, lobules, and papillae (9). This differs from renal atrophy, where the kidney initially develops normally but later decreases in size due to acquired pathology. In adults, distinguishing between hypoplasia and atrophy can be challenging. Renal hypoplasia may be unilateral or bilateral, and in unilateral cases, compensatory hypertrophy of the opposite kidney is commonly seen.

Case 2: A 27-year-old male presents with long-standing hypertension detected 2 years ago. No h/o diabetes. Occasional h/o recurrent urinary tract infections in childhood. No fever or hematuria currently. CECT abdomen was taken.

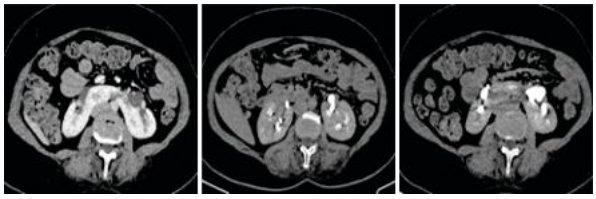


Horseshoe kidneys:

Horseshoe kidney is the most common congenital anomaly (10). It is formed by fusion across the midline of two distinct functioning kidneys, one on each side of the midline. They are connected by an isthmus of renal parenchyma or fibrous tissue. In the vast majority of cases the fusion is between the lower poles. In rest of the case, superior or both the superior or inferior poles are fused. The normal ascent of kidneys are impaired by inferior mesenteric artery which hooks over

the isthmus resulting in a lower abdominal location and abnormal rotation.

Case 3: A 35-year-old female presented with difficulty in passing urine for 2 days. H/o loin pain with vomiting 1 week back, which subsided with IV analgesics. No h/o fever, chills, or hematuria. CECT abdomen was taken.

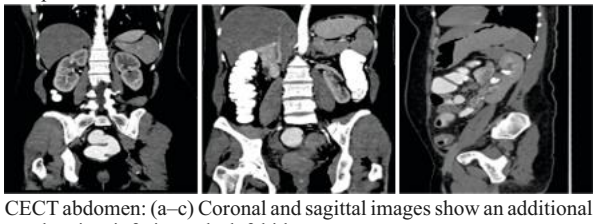


CECT abdomen: (a–c) Axial images show fusion of the lower poles of both kidneys across the midline forming an isthmus, consistent with horseshoe kidney.

Supernumery kidneys:

A supernumerary kidney is an uncommon urogenital anomaly. In this one or more accessory kidneys are seen often on the left and caudal to the native kidney. Mostly the accessory kidney is smaller in size with reduced function. Supernumary kidneys commonly have associated various urogenital and non urogenital anomalies.

Case 4: A 38-year-old female presented with abdominal pain for 2 days. K/c/o ulcerative colitis, ?infective exacerbation. No h/o urinary complaints or hematuria. CECT abdomen was taken.



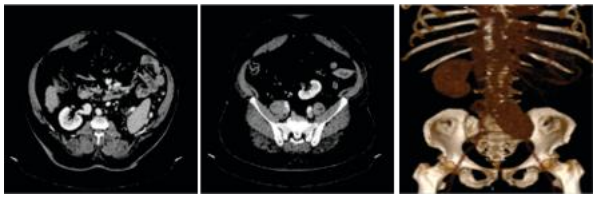
CECT abdomen: (a–c) Coronal and sagittal images show an additional renal moiety inferior to the left kidney

Ectopic and cross fused ectopic kidneys:

Renal ectopia is a congenital renal anomaly characterized by abnormal location of kidneys outside the flank region. The ectopic kidney can be located on the same side as its ureter or crossed ectopia where it is located opposite from its ureteric orifice. The location may be pelvic, iliac, abdominal or chest. Ectopic kidney is usually smaller with varying degree of malrotation.

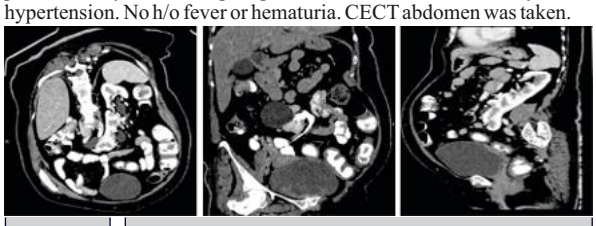
Crossed renal ectopia is often incidentally detected. 85% of the cases the ectopic kidney fuses with the orthotopic kidney (5). Commonest fusion pattern is fusion of lower pole of orthotopic kidney to the upper pole of crossed ectopic kidney. Often there is associated other genitourinary malformations (12).

Case 5: A 46year old male patient, k/c/o chronic liver disease with portal hypertension, presented with upper gastrointestinal bleed. No specific urinary complaints. CECT abdomen was taken.



CECT abdomen: (a–c) Axial and volume-rendered images show a malpositioned kidney located in the pelvis.

Case 6: A 62 year old female patient presented with acute abdominal pain for 4 days and multiple episodes of loose stools. K/c/o systemic hypertension. No h/o fever or hematuria. CECT abdomen was taken.

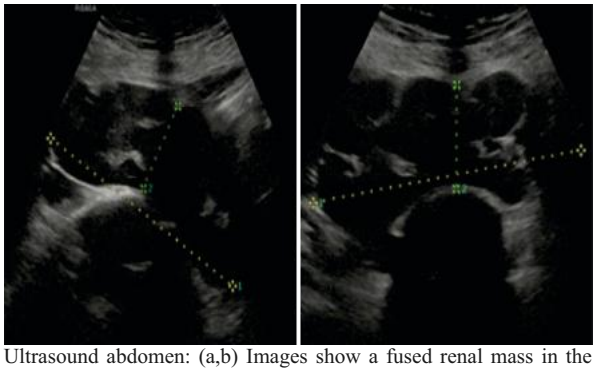


CECT abdomen: (a–c) Axial, coronal and sagittal images show both kidneys located on the left side with fusion.

Pancake kidneys:

Pancake kidney is one of the rarest types of renal ectopia. Pancake kidney is a rare fusion anomaly of the kidneys characterized by the presence of a displaced, lobulated pelvic renal mass of dual parenchymatous system. It is joined at the medial borders of each pole to produce a doughnut or ring-shaped mass without intervening septum. Also known as disc, shield or doughnut kidney is that which has. The pelvis is anteriorly directed and the ureters remain uncrossed.

Case 7: A 32-year-old female presented with lower abdominal pain and urinary frequency for 3 days. No h/o fever, hematuria, or prior similar episodes. USG abdomen was taken.

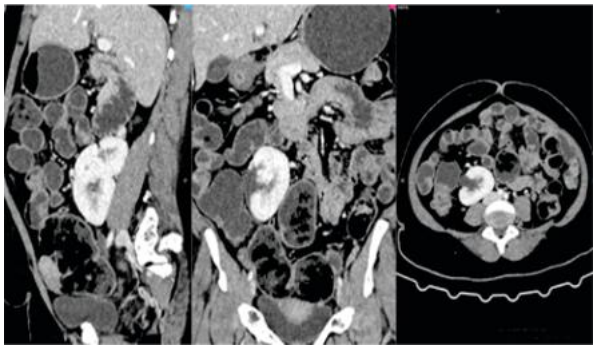


Ultrasound abdomen: (a,b) Images show a fused renal mass in the pelvis with no separation of renal moieties, consistent with a pancake kidney.

Malrotated kidneys:

Malrotation is most commonly associated with an ectopic or fused kidney, but may also occur in kidneys that undergo complete ascent. The condition may be unilateral or bilateral. The most common type is an incomplete rotation, or non-rotation (11). The renal pelvis is directed anteriorly or somewhere in between anterior and normal medial position in adults. Reverse rotation and hyperrotation are major types of malrotation. Anomalies of rotation may produce partial PUJ obstruction.

Case 8: A 40-year-old male presented with non-specific abdominal pain for 1 week. No urinary complaints, fever, or hematuria. Incidentally detected renal anomaly on imaging. CECT abdomen was taken.



CECT abdomen: (a–d) sagittal, coronal, axial and volume-rendered images show a low-lying left kidney with altered hilar orientation.

CONCLUSION:

Timely diagnosis is crucial in selected anomalies to minimize renal damage, prevent or delay the onset of end stage renal disease (ESRD) and provide supportive care to avoid complications of ESRD.

As CAKUT are one of the leading causes of end stage renal disease, it is important for the radiologists to be familiar with the varying imaging appearances of CAKUT on US, CT and MRI, thereby helping in prompt diagnosis and optimal management.

Imaging helps in the early diagnosis, follow-up, surgical planning, detection of complications and associated renal and extra-renal malformations.

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