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PICTORIAL REVIEW

CT findings of the main pathological conditions associated with horseshoe kidneys

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ABSTRACT

Horseshoe kidney (HSK) is the most common renal fusion anomaly, with a prevalence of 0.25% among the general population. It consists of kidney fusion across the midline. HSK can be present as an isolated condition in 30%, but there is a wide variety of associated abnormalities. The most frequent include ureteropelvic obstruction, lithiasis and infections. There is also a higher risk of kidney lesions in trauma and an increased incidence of malignancies. Awareness of embryology and anatomy is essential to assess and understand the complications affecting HSK. CT is an excellent method for identification of its main findings.

Horseshoe kidney (HSK) represents one of the most frequent renal malformations, with an incidence of 0.25% among the general population, being twice as frequent in males.[1–3] It consists of a fusion of the kidneys across the midline, joined by an isthmus of renal parenchyma or fibrous tissue.[1,3] There are no known genetic determinant factors, although it has been described on identical twins and siblings of the same family.[4]

CT TECHNIQUE

Multislice CT allows obtaining images with high spatial and temporal resolution over multiple planes and three-dimensional reconstructions of high quality, and therefore the technique of choice for evaluation of urinary tract anatomy and pathologies associated with HSK.[1]

EMBRYOLOGY OF THE NORMAL KIDNEY

An appropriate knowledge of kidney embryology is necessary in order to understand HSK. During the normal embryological development of the kidney, three successive structures are formed: pronephros, mesonephros and metanephros; the first two structures involute, while the metanephros forms the final kidneys. The last metanephric stage starts during the fourth week, with the union of the intermediate mesodermic metanephral mass at the level of the first or second sacral vertebrae, forming the nephrons and ureteral bud (caudal portion of the mesonephric duct), which forms the ureter, pelvis, calyces and collecting tubules.[1] The kidneys are initially found adjacent with the hilum in an anterior position; as the abdomen and pelvis grow, the kidneys ascend gradually towards the lumbar region and separate.[1] They also rotate 90° medially, to end up with the hilum facing anteromedially.[1] At the end of the ninth week of gestation, they reach their definitive position adjacent to the adrenal glands.[1]

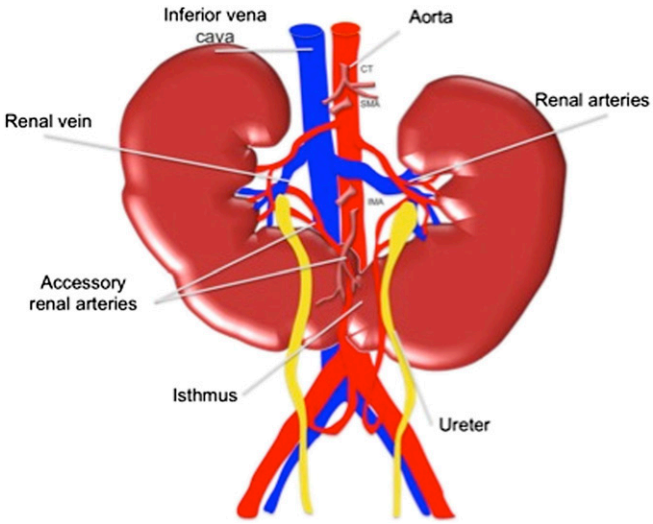
Position and renal fusion abnormalities are the result of an interruption in the normal embryological migration of the kidneys.[5] HSK is the most frequent renal fusion abnormality.

There are two embryological theories depending on the pre-dominant tissue present at the isthmus.[5] When it consists of fibrous tissue, the hypothesis is that between Week 4 and 6 of gestation, after implantation of the ureteral buds, there is fusion of the lower poles, leaving a fibrotic bridge.[5] When the isthmus is parenchymatous (85% of the cases), it has been postulated that the fusion could be the result of a teratogenic process with abnormal migration of the posterior nephrogenic cells joining to form the isthmus.[5] This teratogenic event may be responsible for the higher incidence of congenital abnormalities and for some renal malignancies frequently located at the isthmus.[4]

ANATOMY OF HORSESHOE KIDNEY

HSK consists of a fusion of both kidneys across the midline, joined by a renal parenchymal or fibrous tissue isthmus.[1] In most cases, the fusion occurs at the lower poles of the kidneys.[1] HSK can be located anywhere on the normal

Figure 1. Anatomy of horseshoe kidney. CT, celiac trunk; IMA, inferior mesenteric artery; SMA, superior mesenteric artery.



renal embryological ascending path; nevertheless, most are localized in a low position at the level of the third to fifth lumbar vertebrae, because the isthmus prevents them from ascending beyond the inferior mesenteric artery.[1,6] The fusion also prevents normal renal rotation, leaving the lower poles facing medially, with the renal pelvis located anteriorly and a high ureteral insertion.[1,6] The ureters usually cross in front of the isthmus as they descend towards the bladder.[1] Their blood supply is variable; the renal arteries may originate from the abdominal aorta, iliac arteries or inferior mesenteric artery.[1,6] Venous drainage may occur through supernumerary veins, directly or indirectly draining to the inferior vena cava[1] (Figure 1).

ASSOCIATED COMPLICATIONS

HSKs are asymptomatic in up to 30% of the cases and are an incidental finding during routine examinations; nevertheless, there is a wide variety of genitourinary and extragenitourinary pathologies affecting patients with HSKs.[1]

Complications most commonly seen are ureteropelvic junction obstruction, lithiasis and renal infections. There is also higher risk of renal lesion on abdominal trauma and increased incidence of certain renal malignancies.[1,5] Association with other genitourinary, cardiovascular, gastrointestinal and skeletal malformations, and congenital syndromes such as trisomy 18 and Turner syndrome have been described.[1,5]

Nephrourological complications

Ureteropelvic junction obstruction

There is a higher incidence of obstruction at the ureteropelvic junction in 35% of the patients.[4] Given the abnormal rotation of the kidney, the proximal ureter is oriented higher and medially[1,4] (Figure 2). Ureteral path above the isthmus can also contribute to the obstruction.[1,4]

Renal lithiasis

Kidney stones are found in 20–60% of patients with HSKs.[1,4] This may be owing to ureteropelvic junction obstruction (Figure 3) and to the orientation of the calyces, which determine urinary ectasis and hinder the passage of the stones.[4] Metabolic factors have also been suggested to contribute to stone formation.[4]

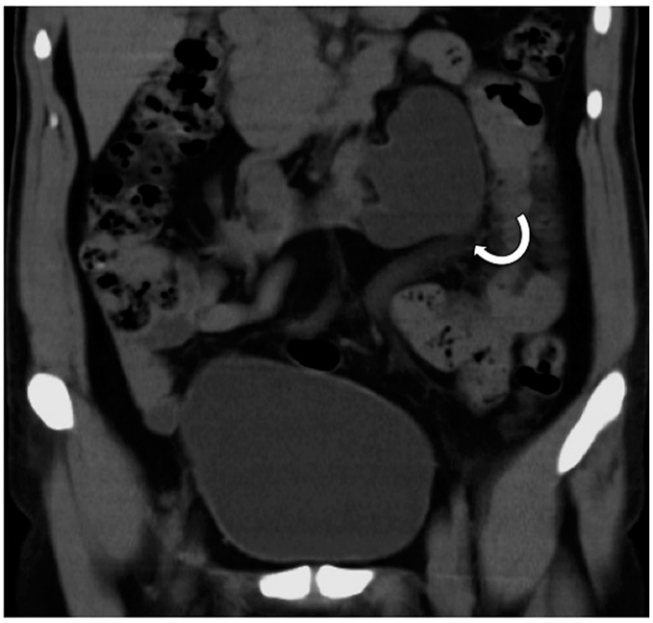
Renal infections

Infections are frequent in HSKs affecting 27–41% of the patients[4] (Figure 4).

Multiple suggested factors include urinary ectasis, vesicoureteral reflux and increased stone formation.[4] These can complicate with abscess formation (Figures 5 and 6), owing to the anatomic and functional characteristics of HSK.[4]

Xanthogranulomatous pyelonephritis has been described in HSKs[7] (Figure 7). This is an infrequent secondary complication

Figure 2. CT of a 53-year-old female with renal failure. (a) Coronal reformatted and (b) CT axial view without contrast. Left ureteropelvic junction obstruction (curved arrow) and marked cortical atrophy with dilation of the calyces and renal pelvis (straight arrow) can be seen.



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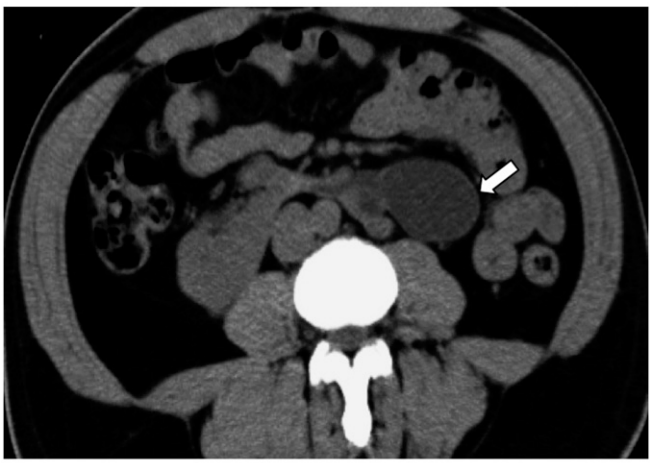


(b)

Figure 3. CT of a 35-year-old male with repeated episodes of renal colic. Coronal reformatted contrast-enhanced CT in corticomedullary phase. Left ureteropelvic obstruction is seen with multiple superior calyceal stones (straight arrow).

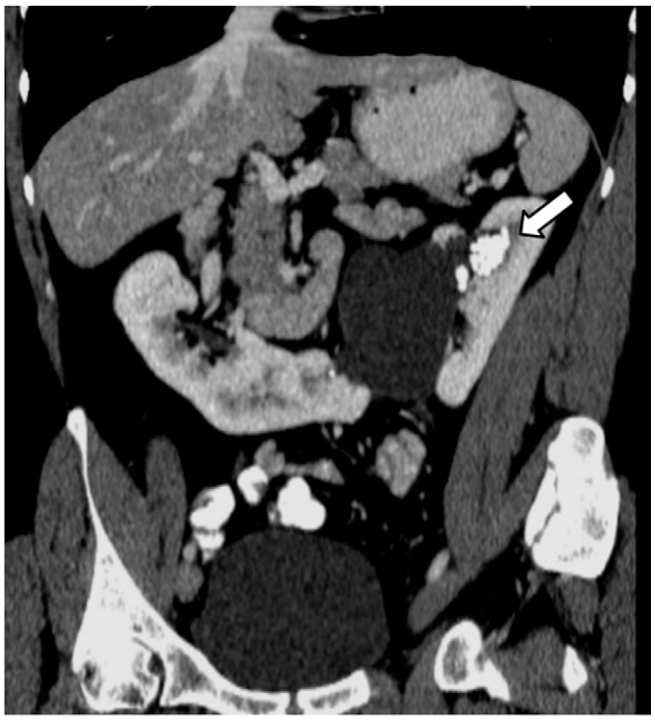
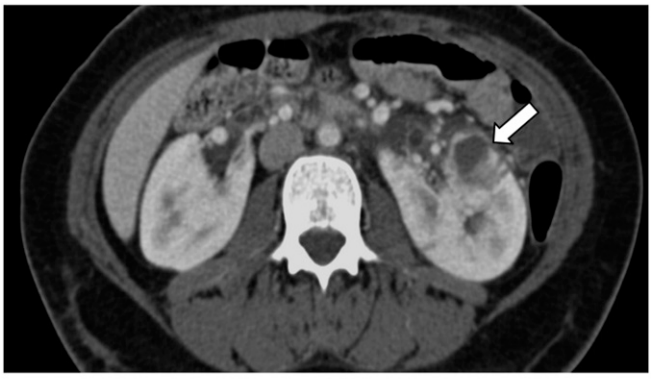


Figure 4. CT of a 45-year-old male with fever. (a, b) Contrast-enhanced CT in corticomedullary phase, axial views at different levels show an anterior abscess on the left side (straight arrow).



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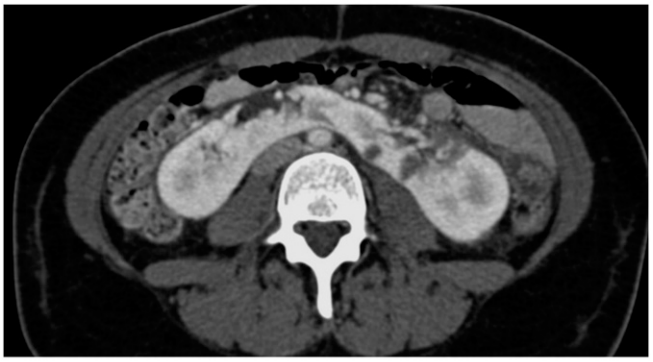
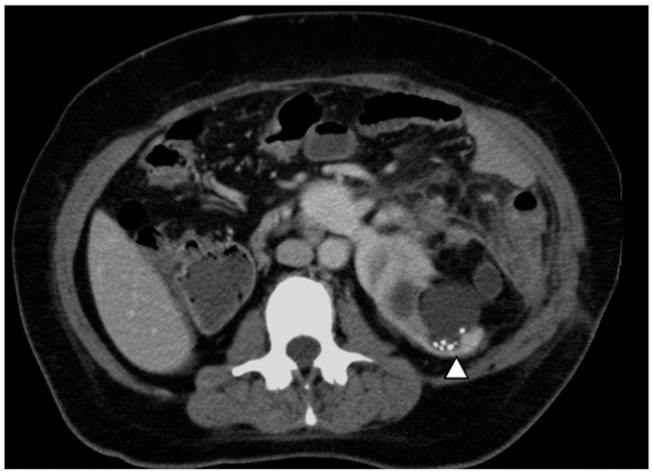


(b)

Figure 5. (a, b) Contrast-enhanced CT in corticomedullary phase, axial views at different levels show left ureteropelvic junction obstruction, stones in the upper calyceal group (arrow head) and thinning of renal parenchyma, associated with inflammatory changes of the anterior and left perinephric fat (straight arrow), compatible with perinephric phlegmonous process.



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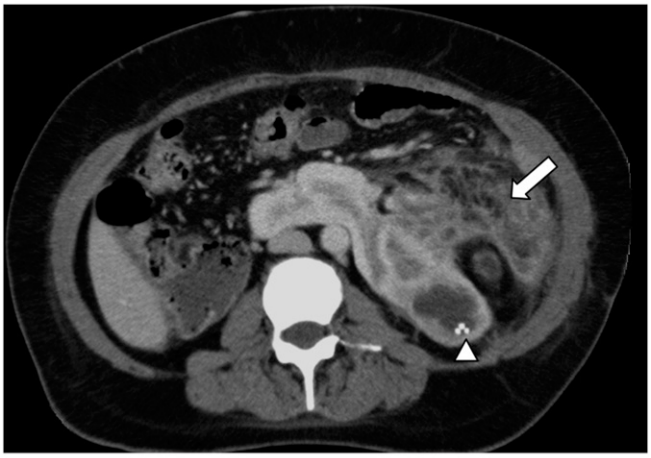


(b)

of a long-standing urinary obstruction associated with chronic renal infection, where the renal parenchyma is destroyed and replaced by lipid-laden macrophages.[8]

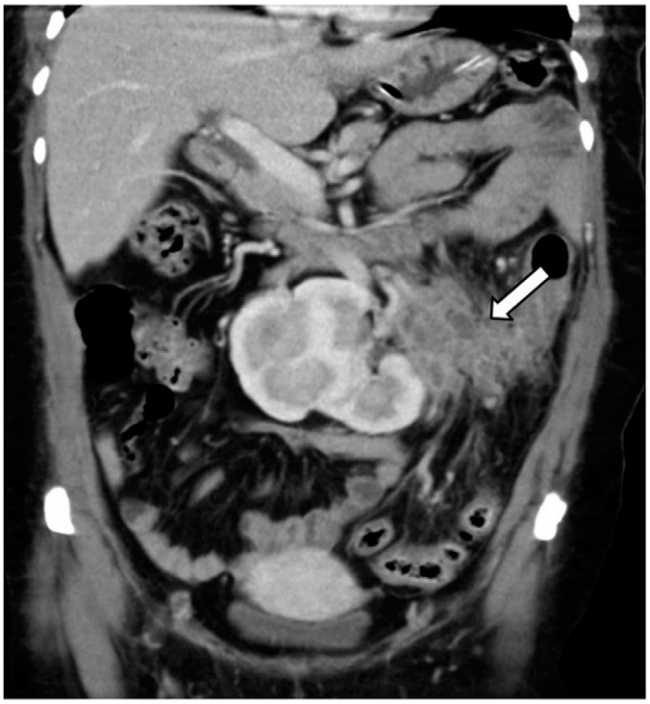
Renal trauma

In closed abdominal trauma, even of low intensity, HSKs are more prone to lesions.[4,5] The isthmus is the most affected area, mainly owing to compression or fracture against the lumbar spine[4,5] caused by trauma in an anteroposterior direction.[4,5] This is due to the position of the renal pelvis, anterior to the lumbar vertebrae and lack of rib protection.[4,5] It can present clinically as gross haematuria in a low-intensity trauma event[5] (Figure 8).

Renal malignancies

Renal cell carcinoma is the most frequent malignancy in HSK patients (45% of renal tumours) (Figure 9); yet, its incidence is similar to that in the normal kidney.[1,4,9]

Figure 6. Coronal reformatted contrast-enhanced CT in corticomedullary phase. Horseshoe kidney fusion variant with a perinephric phlegmonous process (straight arrow).



The risk of other neoplasms, such as Wilms' tumour in children, renal carcinoid and transitional cell carcinomas, is increased in patients with HSK.[1,4,5]

Wilms' tumour is present in 28% of malignant lesions, with a relative risk twice as high as that of a normal kidney;[9] 50% of which appears at the isthmus.[4,9,10] The higher relative incidence, according to some authors, would be owing to the presence of cells derived from nephrogenic primitive cells trapped within the isthmus.[4,6]

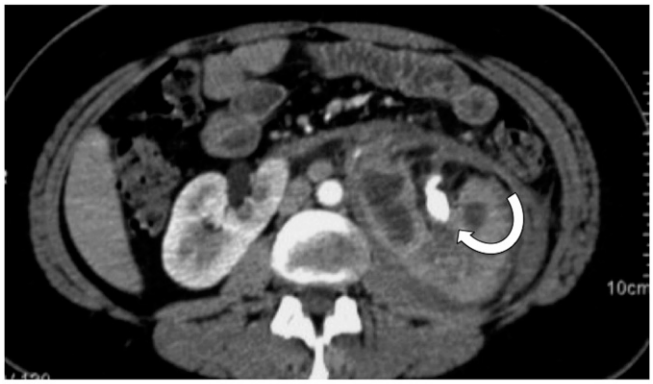
The relative risk of a carcinoid tumour is 62-times higher in a patient with HSKs.[4,5]

Transitional cell tumours account for 20% of tumours in HSKs, the relative risk being 3–4 times higher than that in normal kidneys[4] (Figure 10). The theory is that the higher incidence of infections and lithiasis favours its incidence.[4]

Other associated malformations and congenital syndromes

Among associated congenital malformations, genitourinary ones are the most common and occur in up to 66% of patients.[4] These include vesicoureteral reflux (50%), ureteropelvic junction stenosis (35%), ureteral duplication (10%), cryptorchidism and hypospadias in 4% of male patients and vaginal septum and bicornuate uterus in 7% of females.[4,5] There are other coexisting abnormalities in other organ systems, such as cardiovascular malformations (interventricular communication), gastrointestinal (intestinal malrotation, anorectal malformations and Meckel's diverticulum) and skeletal

Figure 7. (a, b) Contrast-enhanced CT in corticomedullary phase, axial views at different levels show a horseshoe kidney with left xanthogranulomatous pyelonephritis, characterized by a staghorn kidney stone (curved arrow), hypodense areas in the parenchyma (straight arrow), perinephric inflammatory changes and heterogeneous contrast uptake.



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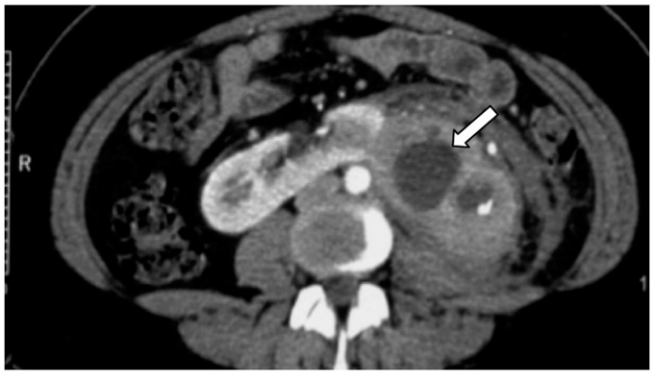


(b)

(hemivertebrae, scoliosis, rib defects, equinovarus foot deformity and hip dysplasia).[4,5]

Some genetic syndromes such as trisomy 18 (20%) or Turner syndrome (60%) can also be associated with HSKs.[4,5]

Differential diagnosis

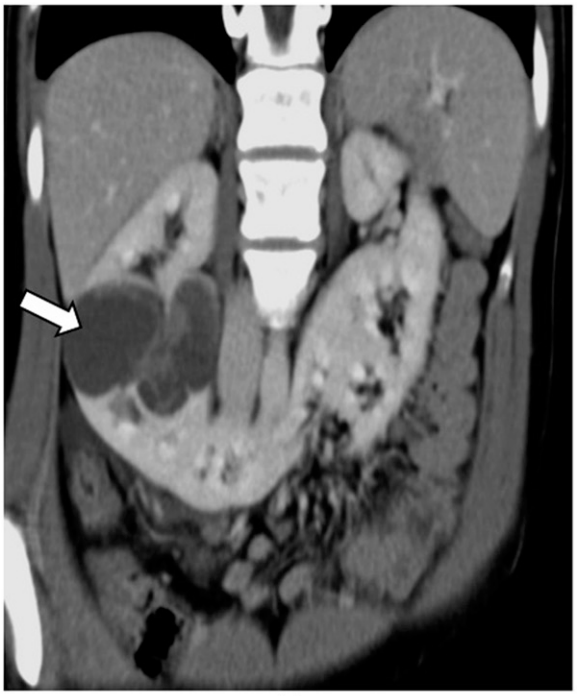
The main differential diagnosis of HSK is another variety of renal ectopia known as pancake kidney, disc kidney or shield kidney. There is a complete midline fusion of the kidneys at the pelvis.[6] It is always found below the L2 vertebral body and is frequently associated with vascular abnormalities of the aortic branches[6] (Figure 11).

CONCLUSION

HSK is the most common renal fusion abnormality, usually an incidental finding, although it is prone to develop traumatic, infectious and tumour pathologies.[1,4] This may be explained by multiple factors such as location and anatomy, embryogenesis, presence of an isthmus and vasculature variability.[1,4]

These pathological conditions must be known by radiologists and clinicians. CT provides an adequate characterization of this condition.

Figure 8. CT of an 8-year-old female who fell from a bicycle and hit the lumbar region. (a) Coronal reformatted contrast-enhanced CT in nephrographic phase, (b) contrast-enhanced CT in nephrographic phase, axial view and (c) CT with contrast in excretory phase, axial view. Multiple pre-existing dilated renal calyces (straight arrows), complicated with haemorrhage, show internal fluid–fluid levels (arrow head).



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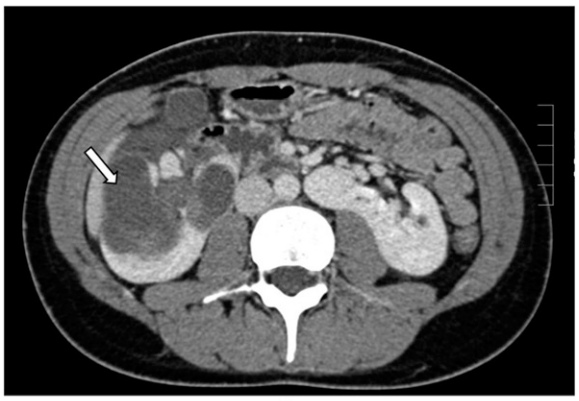

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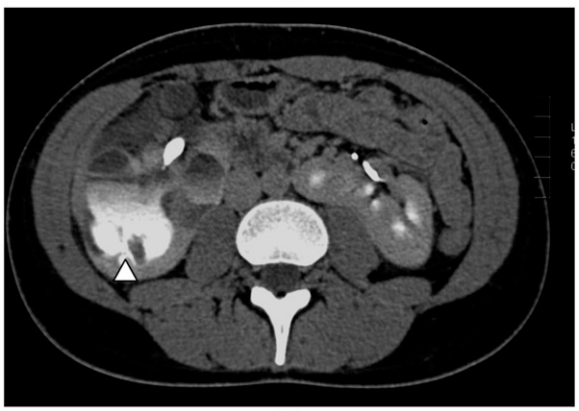


(c)

Figure 9. Coronal reformatted contrast-enhanced CT in corticomedullary phase. Horseshoe kidney with renal cell carcinoma at the right kidney upper pole (straight arrow). There is a right iliac bone lesion associated with a soft-tissue mass, consistent with metastasis (curved arrow).

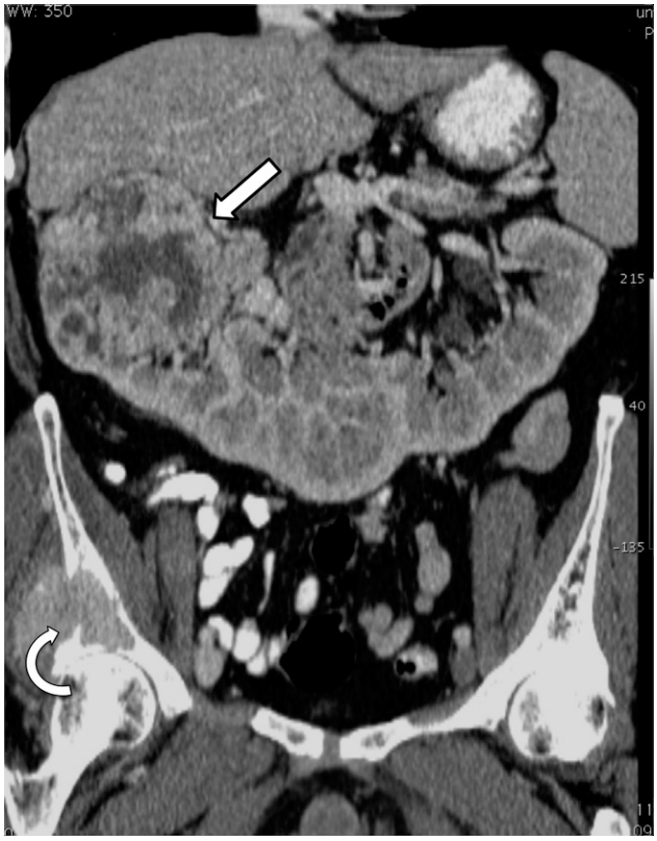
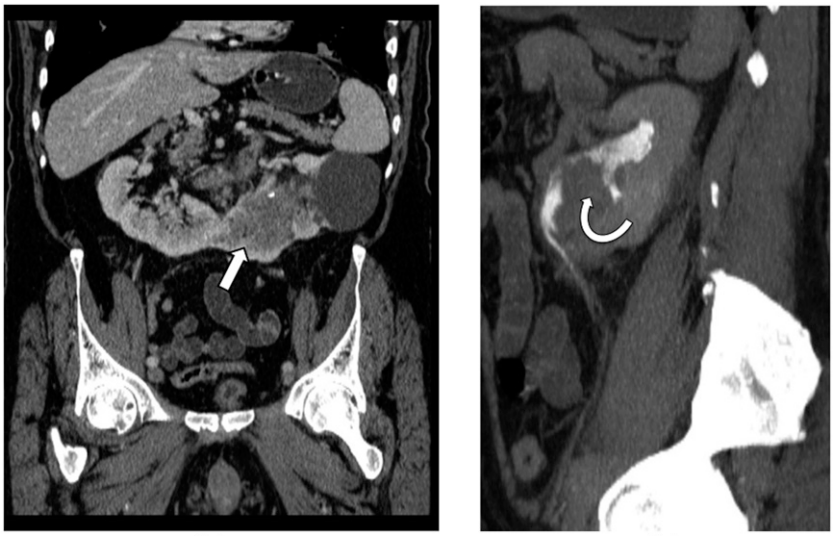
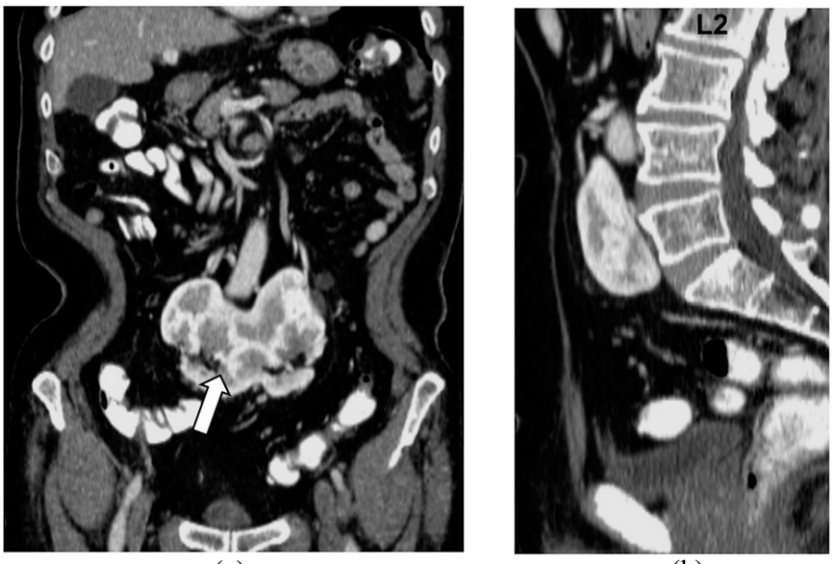


Figure 10. (a) Coronal reformatted contrast-enhanced CT in corticomedullary phase and (b) sagittal reformatted CT with contrast in corticomedullary phase. Horseshoe kidney with a hypervascular tumour (straight arrow) invading the left renal medulla and calyces (curved arrow), consistent with transitional cell tumour.



(a) (b)

Figure 11. (a) Coronal reformatted contrast-enhanced CT in corticomedullary phase and (b) sagittal reformatted CT with contrast in corticomedullary phase. Pelvic kidney, also known as pancake, disc or shield kidney (straight arrow), showing fusion of the kidneys across the midline at the pelvis. It is always found below the L2 vertebral body and is frequently associated with vascular abnormalities of the aortic branches.



(a) (b)

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