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Computed tomography findings of Wilm's tumor in horseshoe kidney

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Abstract

Horseshoe kidney is the most common renal fusion anomaly with fusion across the midline. It has a prevalence of 0.25% among the general population. It may exist as an isolated condition or have associated abnormalities. The most frequently associated complications are ureteropelvic obstruction, calculus formation and infection. There is also a higher risk of renal lesions in trauma and an increased incidence of neoplasms. The role of a radiologist is important for pre- operative management and treatment of the complications.

Keywords

Horseshoe; complications; neoplasms; pre-operative.

Introduction

Fusion anomalies of the kidneys can be of the crossed fused ectopia type or the commoner, horseshoe kidney. Their clinical importance lies in the fact that they are associated with other systemic anomalies and vascular variations which can influence the surgical treatment. The prevalence of horseshoe kidney is 0.25% among the general population. Hydronephrosis, calculi, infection, increased risk of trauma and malignancy is associated with this condition. Understanding the embryology and anatomy is important and cross-sectional imaging plays an important role in the diagnosis & management.

Case presentation

A three-year-old child presented with a palpable abdominal mass and painless hematuria since two

weeks. There was no history of fever and no recent change in bowel habits. Physical examination was normal. Blood pressure was 110/65 mmHg at the time of admission. He had never had any trauma or surgery in the past. An ultrasonography done outside at another institution had revealed a horseshoe kidney with a large heterogeneously hypo echoic abdominopelvic mass lesion arising from the right side. He was subjected to a contrast enhanced Computed Tomography (CECT) of the chest and abdomen.

Horseshoe kidney was seen with a large heterogeneous mass arising from the right kidney and extending anteriorly with displacement of the surrounding bowel loops. It measured ~ 10.8x10.5x10.2 cm. There were no calcifications within the mass lesion. It showed heterogeneous enhancement with few ill-defined, non-enhancing areas, representing necrosis. IVC and right renal vein were compressed and infiltrated by the mass. Medially, it extended across the midline with infiltration of the aorta. The mass was seen to extend anteriorly & inferiorly with a lobulated, exophytic component. The right proximal ureter was not well visualised, likely compressed & infiltrated by the mass lesion. Biopsy confirmed the diagnosis of Wilm's tumor and the child was referred to a higher institution for further management.

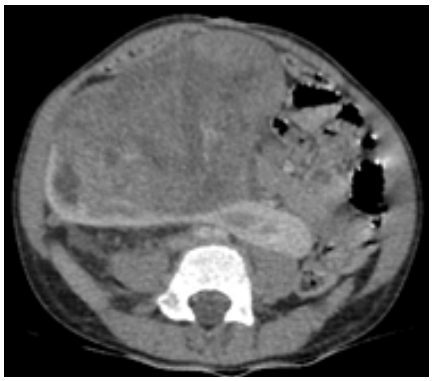

Figure 1: Contrast enhanced CT in the axial plane shows fusion of the kidneys represented by a band of renal parenchyma or isthmus across the midline. There is a large, heterogeneously enhancing mass lesion arising from the right side, extending anteriorly and across the midline.

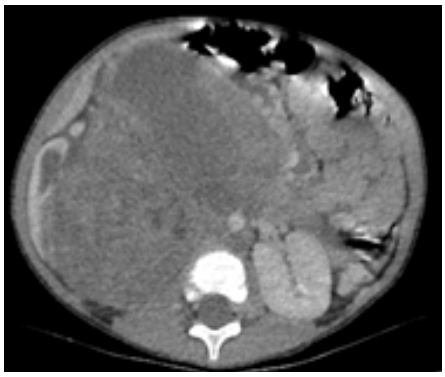

Figure 2: Right renal vein is seen to be displaced anterolaterally with encasement by the mass. The mass is seen to be surrounding the aorta (> 180 degrees), suggesting infiltration.

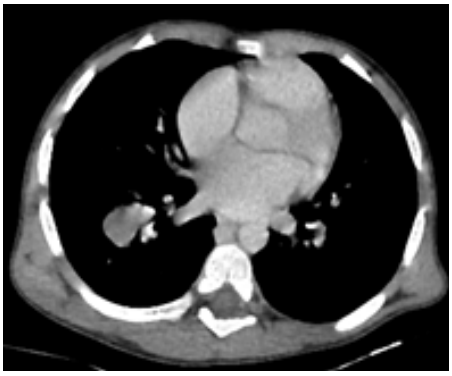

Figure 3: CECT axial image (mediastinal window) shows an enhancing nodule in the right lung, suggestive of metastasis.

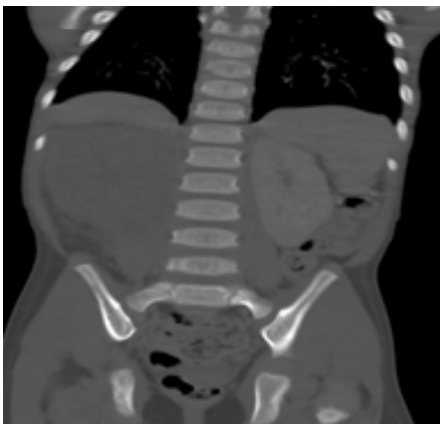

Figure 4: Coronal CT image (bone window) shows no lytic or sclerotic lesions in the visualised bones.

Discussion

Renal fusion anomalies may be discovered incidentally or as a part of a syndrome (Mayer von Rokitansky and Ullrich–Turner syndromes), or may be associated with complications [1]. The incidence of

horseshoe kidney is 0.25% among the general population, being twice as frequent in males [2]. There is fusion of the kidneys, most commonly at the lower poles, with an isthmus of renal parenchyma crossing the midline. Ultrasonography and multiphasic CT are the commonly performed investigations. Position and renal fusion abnormalities are as a result of an interruption in the normal embryological migration of the kidneys. The lower poles face antero medially with the ureters usually crossing in front of the isthmus. The renal arteries may originate from the abdominal aorta, iliac arteries or inferior mesenteric artery. Venous drainage may occur through supernumerary veins, which directly or indirectly drain into the inferior vena cava [3]. The associated complications may be ureteropelvic junction obstruction, calculus formation, renal infections, and higher risk of abdominal trauma and increased incidence of certain renal neoplasms.

0.4 to 0.9 % of all Wilms' tumors occur in horseshoe kidneys with a two times higher risk in children with horseshoe kidney when compared to the general population. The increased risk of the above in the isthmus is explained by the teratogenic event involving abnormal proliferation of metanephric blastema to form isthmus [4]. Genitourinary anomalies associated with Wilms tumor are cryptorchidism, male pseudo-hermaphroditism, hypospadias and renal anomalies such as hypoplasia, ectopic, duplication anomalies and horseshoe kidneys. Non-genitourinary anomalies include sporadic aniridia and hemi-hypertrophy. They present as a large, painless abdominal mass. Microscopic hematuria and hypertension may be found during investigations. The points which aid differentiation from neuroblastoma, another common abdominal mass in the pediatric age group include a younger age of presentation in case of neuroblastoma (below two years of age). Imaging features which favor Wilms are a claw sign, demonstrating renal origin and displacement of adjacent structures rather than encasement. Calcifications are more common in neuroblastoma (80-90 %) [5]. Neuroblastoma most commonly metastasizes to bones and liver while Wilms tumor most commonly demonstrates lung metastasis (hematogenous metastasis indicates stage IV). Treatment is mainly surgical with few cases requiring pre-operative chemotherapy.

Conclusion

Children presenting with horseshoe kidney and renal masses must be evaluated with CT prior to surgical planning. Few characteristic imaging features can help point to a specific diagnosis. Moreover, imaging is required for the pre- operative staging of the disease.

Final diagnosis

Horseshoe kidney with Wilms tumor (stage IV)

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