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Case Report

Radiology Section

Multi-Modality Imaging of Wilms Tumour (WT) in Horseshoe Kidney (HSK)-A Rare Case

BM ASHWINI[1], ANITHA KINI[2], ANIL KUMAR SHUKLA[3]

ABSTRACT

Horseshoe kidney (HSK) is the most common renal fusion anomaly. The association of Wilms tumor (WT) and HSK is uncommon. The most common renal tumor in HSK is WT in pediatric population. Imaging plays a crucial role in evaluation of extent and aids in better management. The aim of this article is to present an uncommon case of WT associated with HSK along with a review of published literature.

Keywords: Calcification, Nephroblastoma, Renal fusion

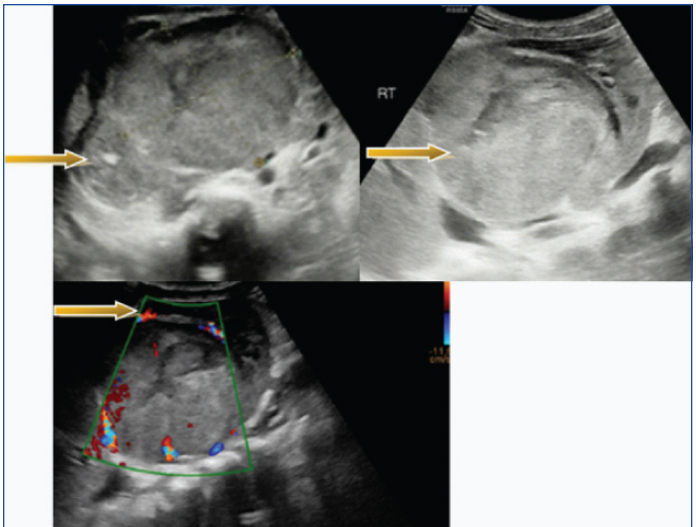
CASE REPORT

A five-year-old boy presented to the Paediatric Department of the institution with complains of fever and pain in the abdomen since four days. The patient had no history of weight loss, reduced appetite or haematuria. Vitals were stable and general physical examination was normal. Per abdomen examination revealed a soft to firm mass present in the right lumbar region which moved with respiration. Rest of the systemic examination were within normal limits. Clinical examination did not reveal any syndromic association. Laboratory investigations were within normal limits.

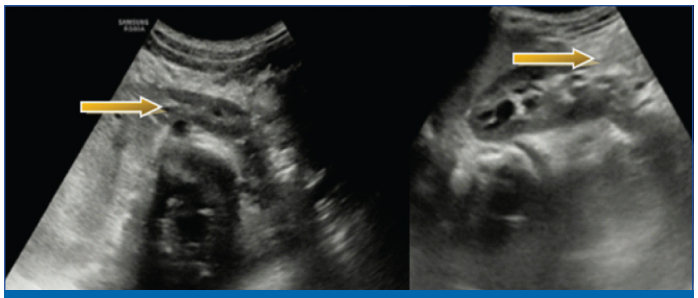
Patient was referred to Department of Radio-diagnosis for ultrasound and Contrast Enhanced Computed Tomography (CECT) examination for further evaluation.

Ultrasound examination of abdomen revealed a large hetero-echoic lesion arising from the upper and mid portion of the right kidney measuring approximately 7.3 cm×7.3 cm×6.5 cm (Volume ~173 cc). Hypo-echoic areas were noted within the lesion with multiple tiny foci of calcification and minimal internal vascularity. There was fusion of the lower poles of both kidneys with anterior mal-rotation of the left renal pelvis [Table/Fig-1,2].

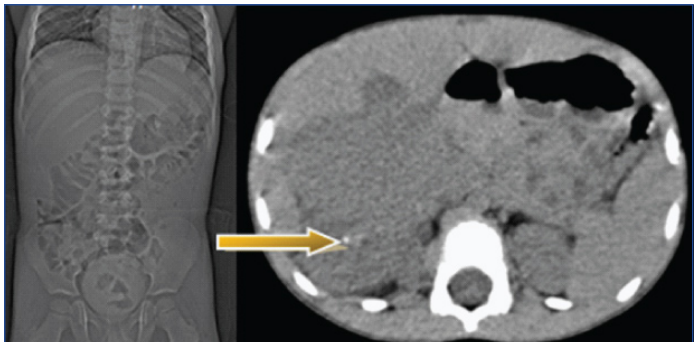
CECT of abdomen showed a large, ill-defined heterogeneously enhancing mass (pre-contrast HU=35-45, post contrast HU=60-70) arising from the upper and mid portion of the right kidney. The mass was causing compression of the right renal vein and inferior vena cava without any encasement of the vessels. The abdominal aorta was normal in calibre. Few calcific foci (HU=150-164) were seen within the lesion. The fat planes between the lesion and liver and adjacent bowel loops were maintained. There was delayed excretion of contrast in the right kidney with infero-lateral displacement of the pelvi-calyceal system. There was fusion of the lower poles of both the kidneys with isthmus comprising of renal parenchymal tissue- Median type (M type) HSK. Bilateral adrenal glands were normal. No lymph adenopathy or fluid around the mass. Bilateral accessory renal arteries were seen. No lytic or sclerotic bony lesions. No ascites or metastases. In correlation with age, clinical presentation and imaging findings, a diagnosis of WT in right kidney with HSK was made [Table/Fig-3,4]. Informed consent was obtained from the parents of the child to publish clinical data.



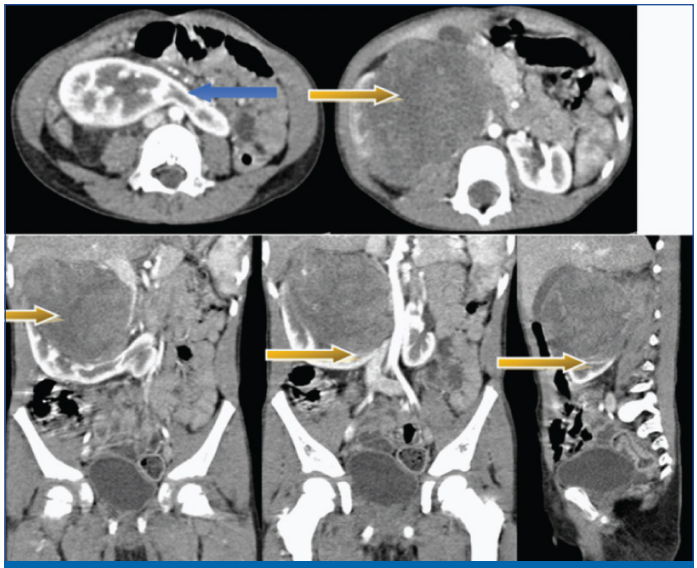
[Table/Fig-1]: Ultrasound of abdomen (Transverse view)– A well-defined large hetero-echoic lesion arising from the upper and mid portion of the right kidney with hypo-echoic areas and multiple tiny foci of calcification. On colour Doppler shows peripheral vascularity (arrows).



[Table/Fig-2]: Ultrasound of abdomen (Transverse view)- shows fusion of the lower poles of both kidneys with anterior mal-rotation of the left renal pelvis (arrows).



[Table/Fig-3]: Topogram and Non contrast CT of abdomen (Transverse view)- shows a soft tissue mass in the right hypochondriac and right lumbar region. The mass shows a foci of calcification (arrow).



[Table/Fig-4]: Contrast CT of abdomen (Transverse, coronal and sagittal views)- shows a large, ill-defined heterogeneously enhancing mass arising from the upper and mid portion of the right kidney. The abdominal aorta was normal in calibre. The fat planes between the lesion and liver and adjacent bowel loops were maintained (yellow arrows). There was fusion of the lower poles of both the kidneys with isthmus compromising of renal parenchymal tissue (blue arrows).

DISCUSSION

HSK is the most common renal fusion anomaly [1]. It results from the abnormal fusion of metanephric blastemas of the two kidneys during the 6th-7th week of gestation, which results in failure of normal ascent and rotation of kidneys [2]. Increased incidence in males (M:F=2:1) is noted [1]. Usually HSK are discovered incidentally in one-third of the patients [3]. Clinical presentation will be asymptomatic or present with symptoms secondary to Pelvic-Ureteric Junction (PUJ) obstruction, infection, stones, malignancy and trauma [4].

HSK is prone to develop in PUJ obstruction, infection and stone formation due to the abnormal location and orientation of the kidneys and calyces, abnormal course and insertion of the ureters and associated vascular anomalies. It is also prone to trauma due to superficial midline location of isthmus combined with absence of ribcage protection [4].

Malignant potential theory hypothesizes the increased risk of development of malignancies such as Renal Cell Carcinoma (RCC), WT and carcinoids. WT is the most common primary malignant renal tumour in childhood. It has a reported prevalence of 8 cases for every 1 million children [5]. The occurrence of WT in HSK is estimated at 0.4-0.9% of all WT [6]. WT is also associated with many genitourinary and non-genitourinary congenital anomalies and syndromes. Genitourinary anomalies 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 [7]. Incidence of HSK in general population is 1:400. In a child with horseshoe kidney, there is a two-fold increased risk of having Wilms tumour compared with the general population [8].

Aetiology of WT in HSK is unknown, few hypothesis postulated are:

- WT develops as a result of sequestered metanephric blastemas in the isthmus, which harbour malignant potential [2].
- The embryologic lesion that results in a HSK may predispose the kidney to a second event resulting in WT [7].

The increased risk of WT in the isthmic location, accounting for almost half of all cases, is explained by the teratogenic event involving abnormal proliferation of metanephric blastema to form isthmus [9]. Other chromosomal anomalies have been reported in children with WT, such as trisomy 18 and 13 [10]. The WT1 gene at 11p13 has been implicated in the development of WT. Normal function of WT1 is necessary for normal genito-urinary development. No studies have identified WT1 mutations in patients with HSK and WT [11]. HSK frequently are associated with anomalous renal arteries. In the present case, there was a bilateral accessory renal artery arising from the abdominal aorta. The combination of the abnormal vasculature and the relatively large tumours seen in children increases the operative difficulty [12].

Multimodality treatment has been used to treat WT successfully. Multiple clinical trials have been conducted by the National Wilms Tumour Study Group (NWTSG) (now incorporated into the COG-Children's Oncology Group) and the International Society of Pediatric Oncology (SIOP) to determine the appropriate role of the therapeutic modalities available.

Stage and histopathology are the most important determinants of outcome in children with WT. There are currently two staging systems available reflecting treatment differences. The system used by the COG reflects staging at primary surgery. Alternatively, the staging by the SIOP is performed after preoperative chemotherapy.

One of the main controversies in the treatment of children with WT is whether to introduce preoperative chemotherapy, as suggested by the SIOP. Opponents of preoperative chemotherapy have argued that the preoperative treatment leads to either over-treatment or under-treatment owing to incorrect staging and histological evaluation. Proponents of preoperative therapy suggest that the tumour is easier to resect with a decreased incidence of tumour spillage and lower mortality and morbidity. Despite the arguments, specific patients seem to benefit from preoperative chemotherapy; these include patients with bilateral WTs, those with inferior vena cava involvement and patients with massive tumours that are un-resectable without undue risk to the patient [13].

The overall survival of patients with WT in a HSK appears to be about the same as for WT in normal kidneys [8]. The present patient was categorized as Stage I as the WT was limited to kidney with no involvement of adjacent structures or vasculature. The patient treatment plan included pre-operative chemotherapy followed by surgical excision. The histology revealed well differentiated Wilms tumour with no anaplastic changes which would be favourable prognosis and less chance of recurrence.

Compared to previous cases, the highlight of the present case is preoperative imaging which helped in assessment of bulk of the tumour for planning of preoperative chemotherapy. Documentation of bilateral accessory renal arteries by preoperative imaging is essential for proper surgical excision.

CONCLUSION

The present case was Stage I of WT. Multi detector CT (MDCT) showed proper extension of the mass which aided in better management and patient care. Horseshoe kidney is the most common renal fusion anomaly. Children with HSK and WT must be carefully examined before any surgery by use of MDCT. Preoperative chemotherapy in this condition might be a good treatment method for decreasing surgical morbidity, promoting complete excision and preserving renal function.

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PARTICULARS OF CONTRIBUTORS:
1. Postgraduate Resident, Department of Radiology, Raja Rajeswari Medical College and Hospital, Bangalore, Karnataka, India.
2. Senior Resident, Department of Radiology, Raja Rajeswari Medical College and Hospital, Bangalore, Karnataka, India.
3. Professor, Department of Radiology, Raja Rajeswari Medical College and Hospital, Bangalore, Karnataka, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Anitha Kini,
#3/1, 9th Main Road, Banashankari, 2nd Stage, Bangalore-560070, Karnataka, India.
E-mail: anusai1709@gmail.com

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