Case 15228

Eurorad

A rare case of primary renal primitive neuroectodermal tumour/Ewing's sarcoma

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ISSN: 1563-4086
Section: Uroradiology & genital male imaging
Area of Interest: Abdomen
Procedure: Diagnostic procedure
Procedure: Contrast agent-intravenous
Imaging Technique: Ultrasound
Imaging Technique: CT
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 23 years, male

Clinical History:

A 23-year-old male patient presented with right flank pain. No complaints of haematuria. No history of smoking.

Imaging Findings:

Ultrasonography of abdomen shows a well-defined heterogenous mass lesion measuring 4.3 x 5.2 cms, involving lower pole of right kidney, with no evidence of calcification or vascularity (Figs. 1a and 1b).

Computed tomography shows a well-defined lobulated isodense mass lesion involving the lower pole of right kidney with few areas of hypodensities and no evidence of calcification. Post-contrast study shows minimal heterogenous enhancement with few non-enhancing necrotic areas within, in both arterial and delayed phases. (Figs 2a, 2b and 2c) No evidence of ascites, lymphadenopathy, adjacent organ invasion or vascular complications.
Coronal and sagittal reformatted CT images demonstrate the tumour involving the lower pole of right kidney whichis minimally enhancing as compared to the normal renal cortex. (Figs 3a and 3b)

Based on these findings diagnosis of malignant renal mass was made. The patient underwent nephrectomy. Biopsy and immuno-histochemistry revealed the diagnosis of PNET.

Discussion:

Primitive neuroectodermal tumour (PNET) or Ewing's sarcoma (EWS) are highly aggressive malignancies originating from the neuroectodermal crest cells.

Histologically these tumours exhibit small round cells with "Homer-Wright rosettes". This feature can be seen in

neuroblastoma, retinoblastoma, medulloblastoma, blastemic nephroblastoma, rhabdomyosarcoma, small cell osteosarcoma and lymphoma. The surface membrane antigen MIC2 (CD99) overexpression and chromosomal translocation between chromosome 11 and 22 {t (11, 22)(q24;q12)} are diagnostic of PNET tumours and useful to distinguish it from other tumours. [1]

PNET/EWS is primarily a bone or soft tissue tumour. Peripheral PNETs are uncommon with 1% incidence of all sarcomas. Peripheral PNETs are usually seen in the chest wall, extremities, paravertebral regions, pelvis, and retroperitoneum.[2]

Primary PNET/EWS of the kidney is very rare. The adrenergic fibers accompanying efferent arterioles and descending vasa recta that innervate the kidney have been proposed as the possible origin of this tumour.[3]

Renal PNET affects children and young adults, with a male predominance. It presents as a flank pain, palpable abdominal mass, haematuria or weight loss.

On imaging, these tumours show heterogeneous enhancement with areas of necrosis or haemorrhage reflectingthe aggressive nature. Extensive vascular invasion of surrounding vessels and thrombosis of IVC or renal vein can be seen. Direct invasion of the adjacent structures like pancreas, spleen, stomach, psoas muscle and ureter is also reported. Punctate calcifications are evidenced in 10 % of tumours. It can metastasise to the lung, lymph nodes, liver, and bones [4].

Advanced RCC is a close differential for PNET as both can show necrotic and haemorrhagic mass with vascular thrombosis and adjacent structural invasion; however, we can differentiate them based on following characteristics.
1. Multiple septa like structures are detected in both necrotic and solid portions of PNET.
2. PNET shows diffuse multifocal haemorrhage and necrosis whereas RCC shows central necrosis or haemorrhage.
3 PNET shows very weak enhancement both on the arterial and delayed phases, unlike conventional RCCs. The degree of enhancement of PNET is negligible compared with that of the normal renal cortex. [5]

Surgery and chemotherapy are the treatment modalities with a five-year survival rate of 45% - 55% [6].

Renal PNET/EWS should be considered as a differential in a case of large renal mass with aggressive features in younger symptomatic patients because this has a significant impact on the treatment and prognosis.

Differential Diagnosis List: Primary renal primitive neuroectodermal tumour/Ewing's sarcoma, Renal cell carcinoma, Renal lymphoma

Final Diagnosis: Primary renal primitive neuroectodermal tumour/Ewing's sarcoma

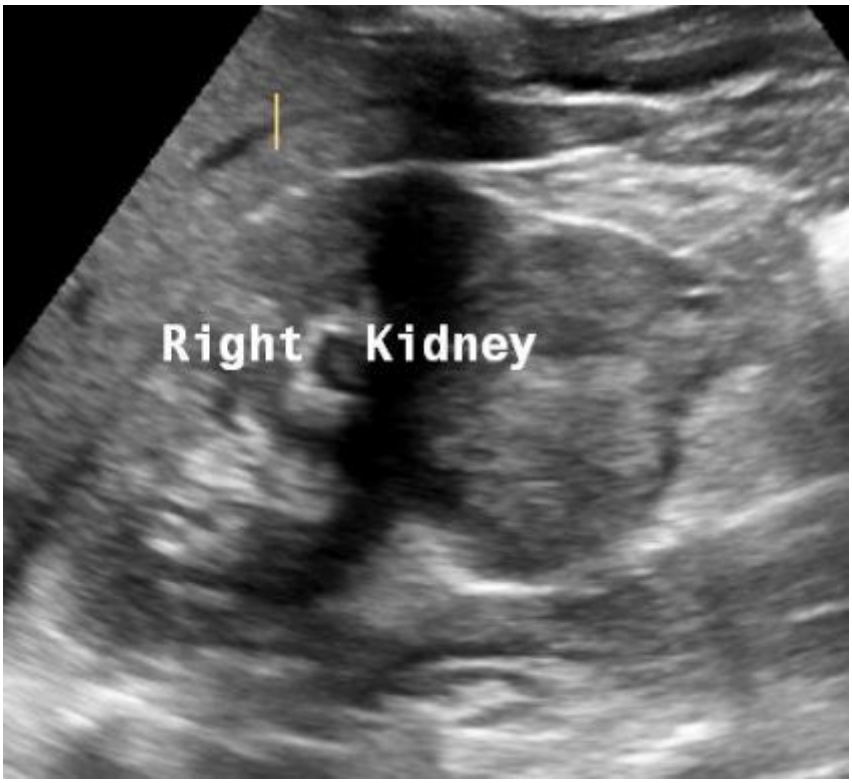
References:

Mukkunda R., Venkitaraman R., Thway K., Min T., Fisher C., Horwich A., Judson I (2009) Primary Adult Renal Ewing's Sarcoma: A Rare Entity. (PMID: 19478963)
Maccioni F, Della Rocca C, Salvi PF, et al (2000) Malignant peripheral neuroectodermal tumor (MPNET) of the kidney. Abdom Imaging (PMID: 10652933)
Parham DM, Roloson GJ, Feely M, Green DM, Bridge JA, Beckwith JB. (2001) Primary malignant neuroepithelial tumors of the kidney: a clinicopathologic analysis of 146 adult and pediatric cases from the National Wilms' Tumor Study Group Pathology Center. Am J Surg Pathol (PMID: 11176062)
Kim MS, Kim B, Park CS, Song SY, Lee EJ, Park NH, Kim HS, Kim SH, Cho KS (2006) Radiologic findings of peripheral primitive neuroectodermal tumor arising in the retroperitoneum. American Journal of Roentgenology

(PMID: 16554591)
Imaging Findings of Primitive Neuroectodermal Tumors of the Kidney (2009) (PMID: 19940655)
Hakky TS, Gonzalvo AA, Lockhart JL, Rodriguez AR. (2013) Primary Ewing sarcoma of the kidney: a symptomatic presentation and review of the literature. Ther Adv Urol (PMID: 23730330)

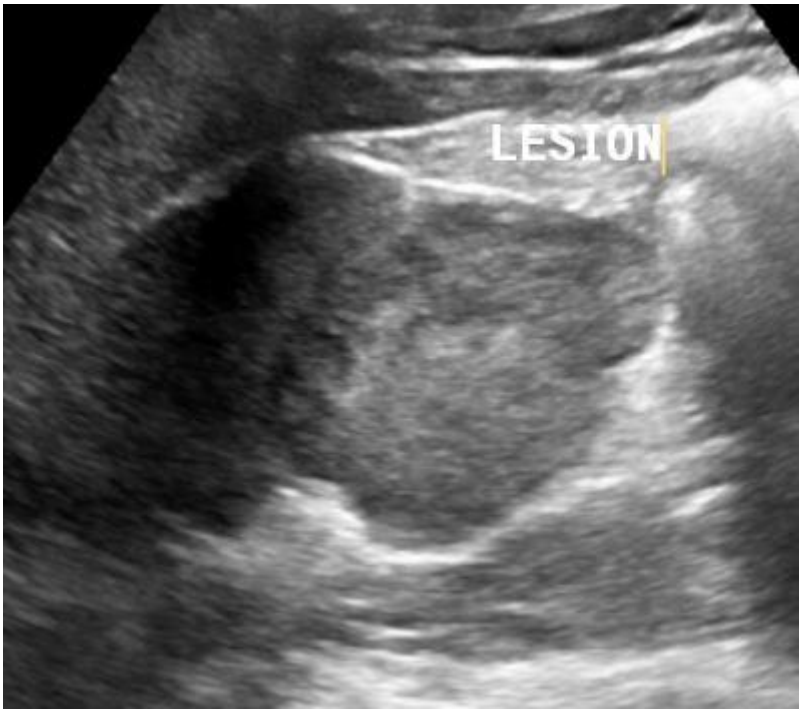
Figure 1

a



Description: Ultrasonography of abdomen shows a well-defined lobulated iso to hyperechoic mass lesion with hypoechoic areas within, involving lower pole of right kidney. **Origin:** Department of radiology,MMCRI, Mysuru

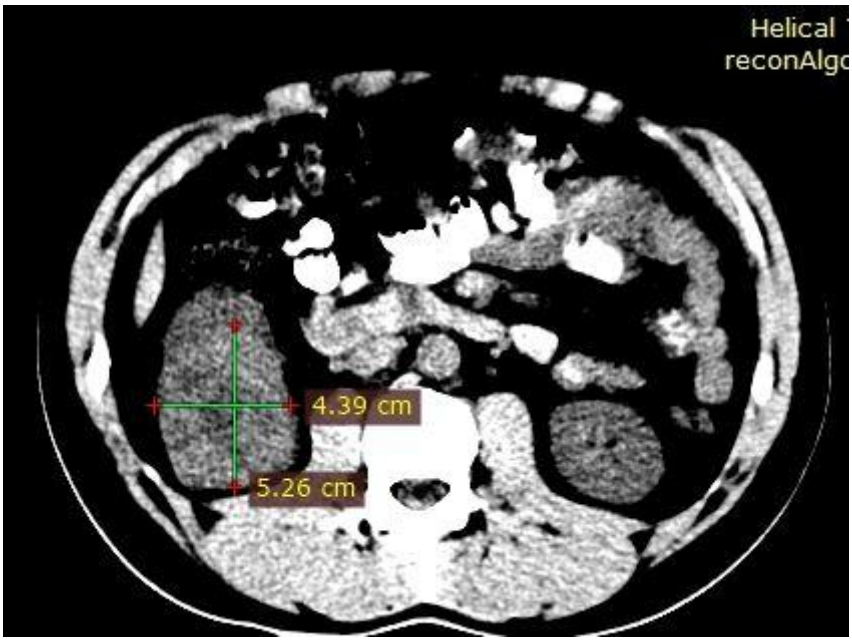
b



Description: Ultrasonography of abdomen shows a well-defined lobulated iso to hyperechoic mass lesion with hypoechoic areas within, involving lower pole of right kidney. **Origin:** Department of radiology,MMCRI, Mysuru

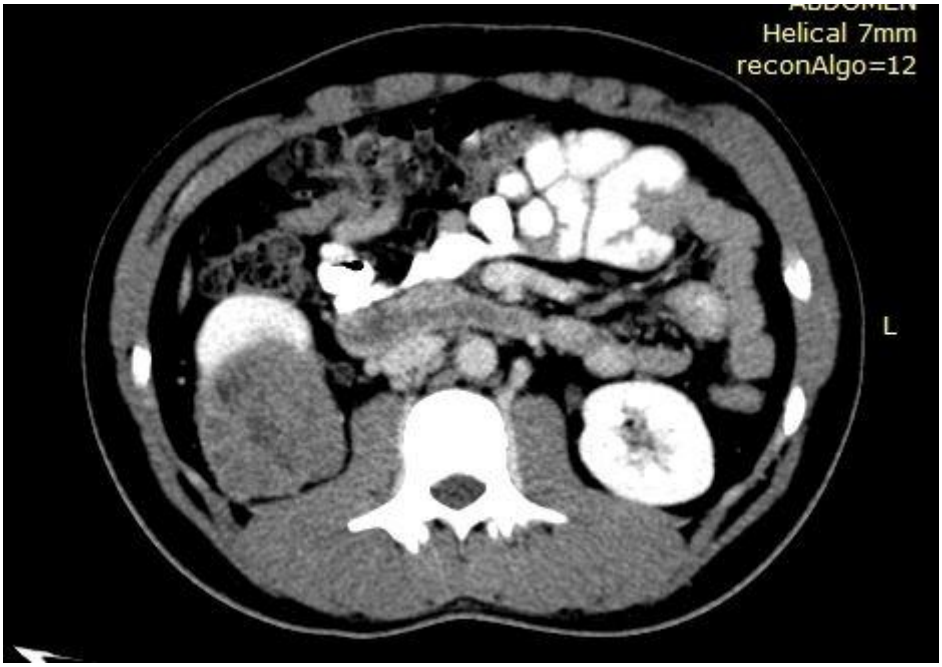
Figure 2

a



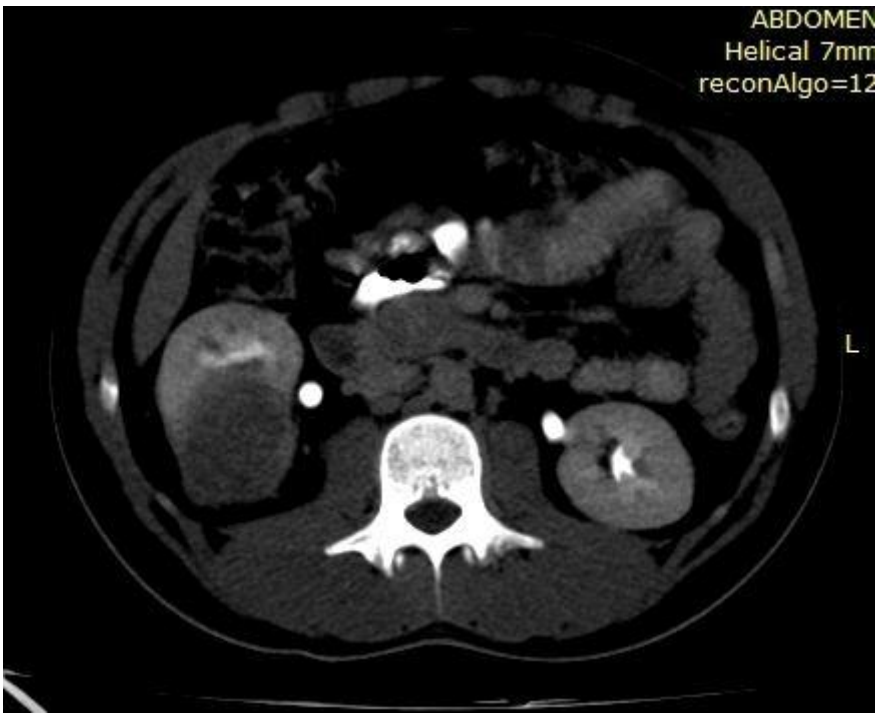
Description: Axial plain computed tomography image shows a well-defined lobulated isodense mass lesion involving the lower pole of left kidney, with few areas of hypodensities. **Origin:** Department of radiology,MMCRI, Mysuru

b



Description: Contrast-enhanced axial computed tomography image shows minimal heterogenous enhancement with few nonenhancing necrotic areas within the mass lesion of right lower pole. **Origin:** Department of radiology,MMCRI, Mysuru

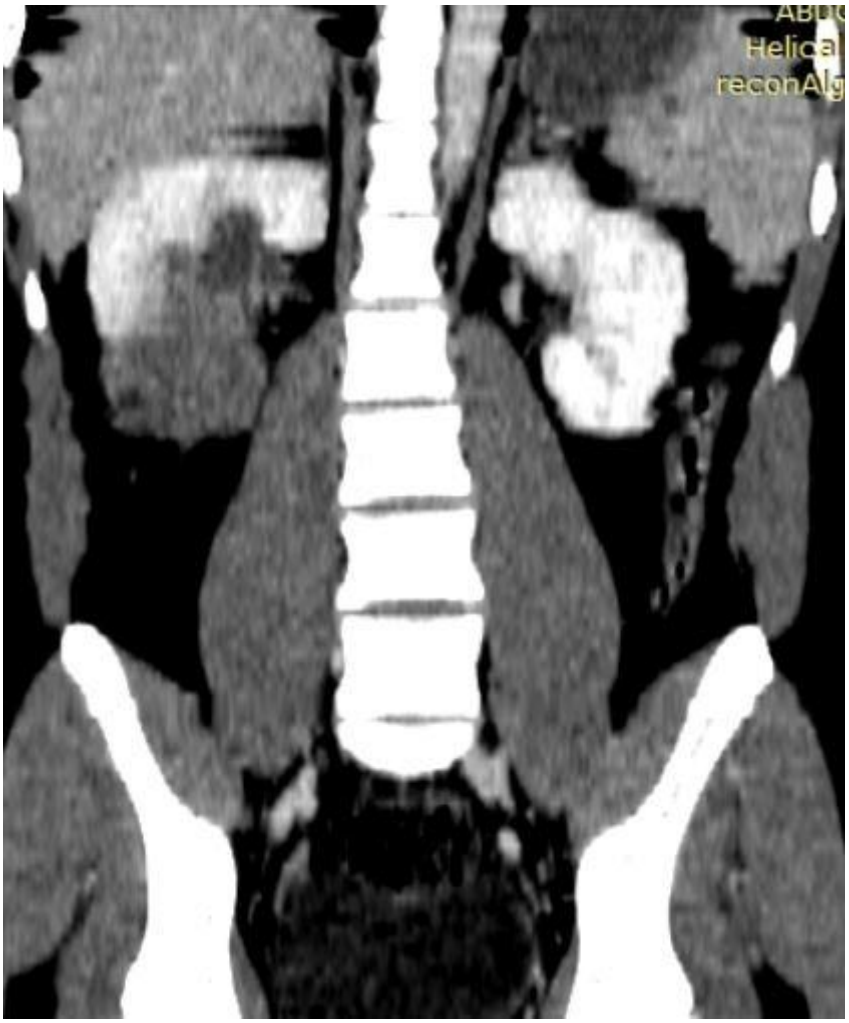
c



Description: Post-contrast delayed axial CT image showing sharply demarcated hypoenhancing mass lesion in the lower pole of right kidney. **Origin:** Department of radiology,MMCRI, Mysuru

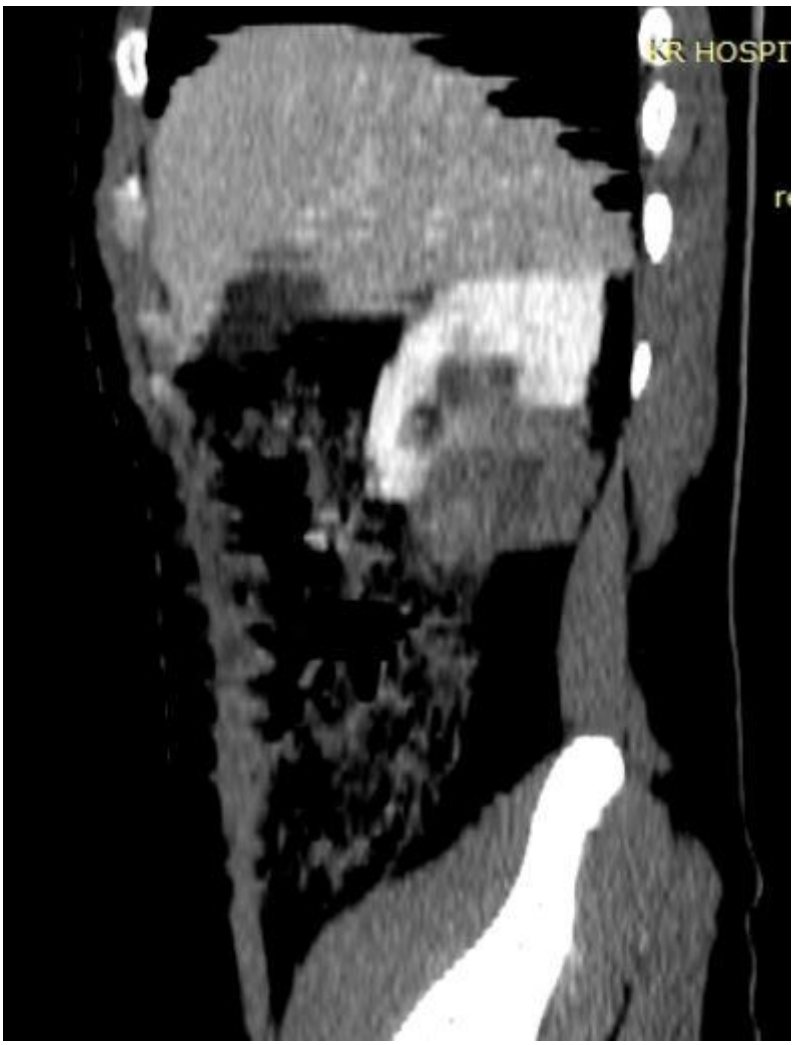
Figure 3

a



Description: Coronal reformatted CECT image demonstrates the tumour involving the lower pole of right kidney which is minimally enhancing as compared to normal renal cortex. **Origin:** Department of radiology,MMCRI, Mysuru

b



Description: Sagittal reformatted CECT image demonstrates the tumour involving the lower pole of right kidney which is minimally enhancing as compared to normal renal cortex **Origin:** Department of radiology,MMCRI, Mysuru