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Primitive Neurectodermal Tumour of Kidney – An Uncommon Tumour At a Rare Site

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ABSTRACT

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A case of Primitive Neuroectodermal Tumour (PNET) of the kidney in a 31-year-old gentleman is presented. Few cases are reported in the literature with a variable, nonspecific presentation and an aggressive behaviour. In our case, a radical nephrectomy, lymphadenectomy and IVC repair was performed. The surgical specimens were formalin-fixed and paraffin embedded. The sections were stained with routine H&E. Immunohistochemistry was performed. The immunohistochemical evaluation revealed CD-99positive, Membrane positivity, NSE positivity, Synaptophysin-focal positivity and EMA-negativity. The clinical presentation and the macroscopic aspect, together with the histological pattern, the cytological characteristic and the cellular immunophenotype addressed the diagnosis towards primary PNET of kidney Since sometimes it is difficult to discriminate between PNET and Ewing's tumour, we reviewed the difficulties in differential diagnosis. These tumours have a common precursor but the stage of differentiation in which it is blocked is probably different. This could also explain their different biological behaviour and prognosis.

Keywords: Renal PNET, Ewing's Sarcoma

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CASE REPORT

A 31 year old gentleman presented with abdominal lump of one month duration , haematuria on and off 2 months duration which is gross, intermittent, painless with passage of clots and associated abdominal pain. On general examination patient is hypertensive. On physical examination revealed an abdominal lump of size 20*15cm which is hard and immobile. MR Urogram revealed a large mass in right kidney having highly heterogeneous signals most of the mass shows intermediate low T2 signals and the mass involved renal artery and vein and extends over the IVC and psoas major. Metastatic workup done came as negative. Patient underwent radical nephrectomy and IVC repair on 02-08-2017 and the specimen of size 25*20cm, histopathology report came as primitive neurectodermal tumour

DISCUSSION

PNET of the kidney was first reported by Seemayer and colleagues in 1975[1] and is exceedingly rare. It usually affects young adults at a median age of 28 years and has a male predominance of 3:1.[2,3] The common presenting symptoms are flank or abdominal pain, mass in the abdomen, and haematuria[4] Patients are usually asymptomatic until the tumour reaches a large size; the maximum diameter of such tumours is often 10 cm.[5,6] Systemic symptoms such as weight loss and fever may also occur.[7] Most of the patients present at an advanced stage with distant metastasis, which is in concordance with the aggressive behaviour of this tumour.[2,8] Common sites of metastasis include the lung, liver, and bone[4]

The imaging characteristics of renal PNET are often nonspecific and overlap with those of other renal tumours, such as renal cell carcinoma, Wilms tumor, neuroblastoma, lymphoma, and desmoplastic small round cell tumor.[9] Renal PNETs appear hypoechoic, isoechoic, and /or hyperechoic to the adjacent renal parenchyma on ultrasound and show increased vascularity on Doppler imaging. CT scan shows a large heterogenous mass with areas of hemorrhage or necrosis.[10] On magnetic resonance imaging (MRI), the tumor appears as a lobulated isointense and /or hypointense mass on T1-weighted images and as a heterogeneous to hyperintense mass on T2-weighted images, with het-

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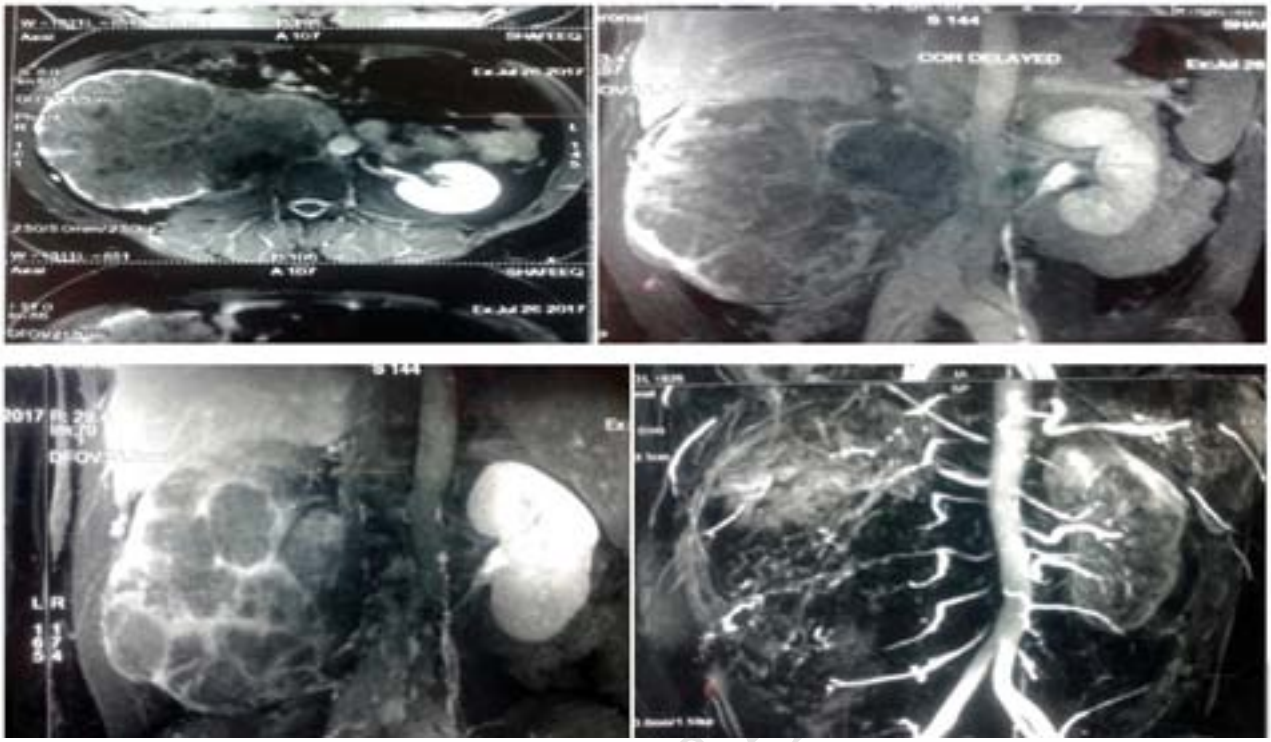
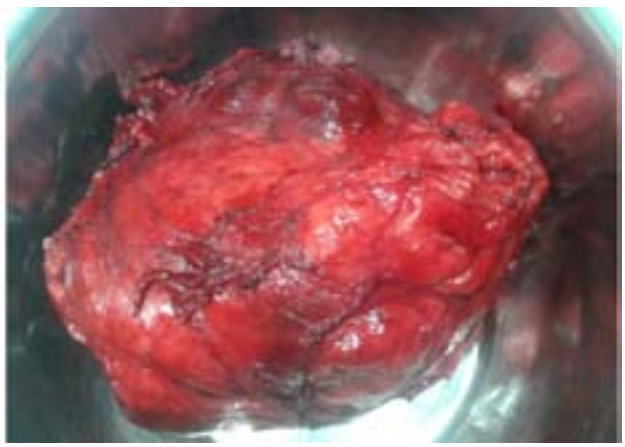


AQ2 Figure 1. **MR Urogram - A large mass in right kidney having highly heterogeneous signals most of the mass shows intermediate low T2 signals and the mass involved renal artery and vein and extends over the IVC and psoas major**

erogenous contrast enhancement.[4,10] MRI and CT also help to evaluate renal vein and inferior vena caval involvement. Technetium-99m scintigraphy is useful for the detection of bone metastases.[4]

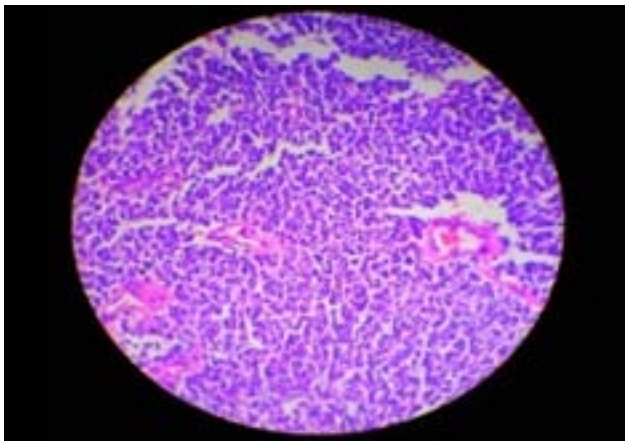
Diagnosis of renal PNET is challenging. Although radiological features may be suggestive, biopsy with immunohistochemistry is required to confirm the diagnosis. Renal PNET is characterized by small uniform round cells with dark nuclei, ill-defined cytoplasmic borders, and poorly formed rosette-like structures.[7] The histopathologic features overlap with other small round blue cell tumors like neuroblastoma, desmoplastic small round cell tumor, and lymphoma. Immunohistochemistry and molecular studies play a crucial role in differentiating these tumors. PNET shows strong positivity for MIC-2 gene product and CD-99 over the membrane of tumor cells, which is seen in more than 90% of renal PNET cases.[5] It is also positive for different neural biomarkers such as S-100, Leu 7, and NSE.[11] The Immunohistochemistry findings are further supported by the identification of a characteristic EWSR1/FLI1 fusion product that results from a t(11;22) (q24/q22;q12) translocation. This translocation is identified in 90% of cases and unequivocally confirms the diagnosis.[5,12] Among our case, IHC details were positive for CD99, NSE, Synaptophysin

Renal PNET appears to be a unique clinical entity that behaves more aggressively than PNET arising at other sites. As the tumor is highly aggressive, it is often diagnosed in an advanced stage when it has already



AQ3 Figure 2. **Gross specimen**

Figure 3. **Pathologic specimen** AQ4



AQ5 Figure 4. **Microscopy**

involved perinephric fat, hilar lymph nodes, renal veins, and the inferior vena cava. In more advanced stages, the tumor involves the liver, spleen, peritoneum, and lungs. The prognosis for renal ES/PNET is generally poor, with a 5-year disease-free survival of 45% to 55% in localized cases, whereas cases with an advanced stage at presentation have a median relapse-free survival of only 2 years.[7,11,13]

The treatment for renal ES/PNET is similar to that for ES/PNET elsewhere and includes surgery, chemotherapy, and radiation.[4,8] Surgical options include partial or total nephrectomy with cavotomy in cases of renal vein involvement.[4,12] The diagnosis of renal ES/PNET is often made postoperatively and hence chemotherapy is generally given as an adjuvant. The recommended chemotherapy regimen is vincristine, doxorubicin, and cyclophosphamide alternating with ifosfamide and etoposide given for a period of 1 year.[14] The role of radiotherapy is not clear, but it may be given in locally advanced disease and in those with positive margins. Despite aggressive therapy, the overall cure rate of renal PNET is only 20%.[15] Since the overall prognosis of this tumor is poor, an early and accurate diagnosis is crucial for the proper management of these aggressive tumors

AQ7 ## END NOTE ???

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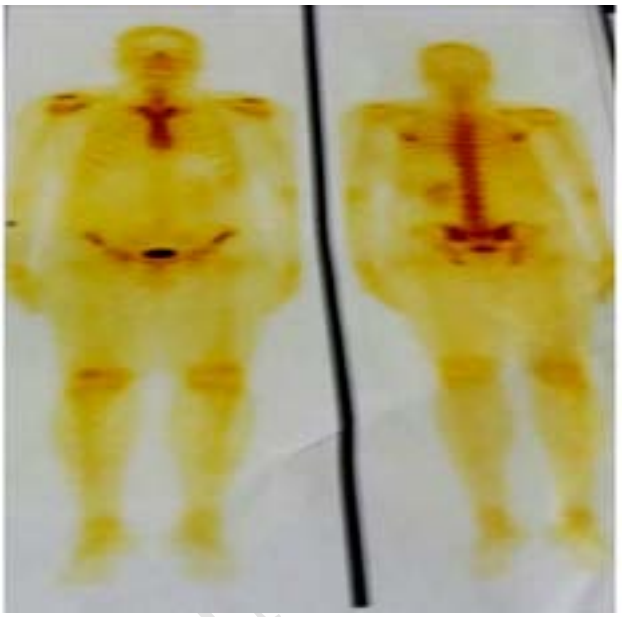

Figure 5. **Technetium-99m scintigraphy bone scan normal study** AQ6

Conflict of Interest: None declared

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