Case Report

Primitive Neuroectodermal Tumour of Kidney: An Unusual Entity

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ABSTRACT

Primitive neuroectodermal tumor (rPNET) of kidney is an extremely rare tumour which occurs in children and young adults and has only a few published reports. The present case is of a 24-year-old female presenting with complaining of pain abdomen since 2-3 months duration. USG guided FNAC showed a cellular aspirate composed of small round to oval cells with scanty or small amount of cytoplasm was seen. Cells were arranged in clusters with occasional micro-acinar or papillary arrangement was also noted. Two possibilities were suggested- 1.Carcinoid tumour .2. small cell anaplastic carcinoma. Histopathological and immunohistochemical (IHC) correlation was done. A highly malignant small cell neoplasm composed of small round cells having scanty cytoplasm and dense nuclei against hemorrhagic background was seen. IHC panel was performed and was found to be strongly positive for MIC-2 (CD99) and showed weak positivity for NSE and ki -67. A final diagnosis of Primitive Neuro-ectodermal Tumour of kidney was made.

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Introduction

Renal primitive neuroectodermal tumor (rPNET) as a member of Ewing's sarcoma family is extremely rare and usually occurs in children and young adults.[1] PNET of the kidney is a rare tumor with only a few published reports. It was first described by Arthur Purdy Stout in 1918 and was included in the family of small round cell tumors. [2] In view of poorer prognosis and different therapeutic approach, renal PNET should therefore be differentiated from other primary renal neoplasms such as Wilms tumor, renal neuroblastoma and malignant rhabdoid tumor which on histology resemble renal PNET(r PNET).[3]

Case Report

A 24-year-old female presented with the complaints of pain abdomen since 2-3 months duration. CT scan of the abdomen was performed which revealed a large expansile solid cystic exophytic mass in the right kidney extending upto gall bladder fossa & causing tumour thrombus in right renal vein and adjoining IVC (inferior vena cava) lumen .

USG guided FNAC was performed and a cellular aspirate composed of small round to oval cells with scanty cytoplasm was seen. Cells were arranged in clusters with occasional micro-acinar or papillary arrangement. Two possibilities were suggested; 1. carcinoid tumour and 2. small cell anaplastic carcinoma. Radical nephrectomy was performed. On gross examination, a circumscribed grayish white mass measuring 5x4 cm was seen occupying the lower pole of right kidney (ure 1). Focal areas of hemorrhage and necrosis were also noted. Histopathological and immunohistochemical (IHC) analysis was performed. On microscopic examination, a highly malignant neoplasm composed of small round cells having scanty cytoplasm and dense nuclei set against hemorrhagic background were present (ure 2). An IHC panel was performed and found to be strongly positive for MIC -2 (CD99) (ure 3) and weakly positive for NSE (ure 4) and Ki-67. A final diagnosis of Primitive neuro-ectodermal tumour of kidney was made.

Discussion

rPNET first described by Mor et al. in 1975 is a very rare and aggressive malignant tumor.[4] rPNET usually occurs in children and young adults. Boys and men are more likely to suffer from rPNET and the sex ratio (male:female) is about 3:1. The tumors tend to be very large and the maximum diameter of rPNET is always>10 cm.[5,6,7] So far, most literature about rPNET was isolated case reports and the largest case series including 79 patients with rPNET was described by Parham et al. in 2001.[5] The age of these patients ranged from 2 months to 73 years old with a median age of 20 years.

It is characterized by small, round, monomorphic cells with dark nuclei and ill defined cytoplasmic borders. IHC staining of rPNET is always positive for different neural biomarkers such as S-100, Leu 7 (HNK-1), and particularly NSE. Additionally, CD99 also named MIC-2 antigen is crucial in the diagnosis of rPNET and the positive expression ofCD99 has been demonstrated in more than 90% of rPNET [6] But, CD99 is not specific and cannot be used as an absolute marker.

PNET is a highly aggressive neoplasm and should be differentiated from other round cell tumours of kidney like neuroblastoma, adult nephroblastoma, malignant lymphoma, small cell carcinoma and monophasic poorly round cell variant of synovial sarcoma. IHC markers should be used to confirm diagnosis are cytokeratin (nephroblastoma, small cell carcinoma, and synovial

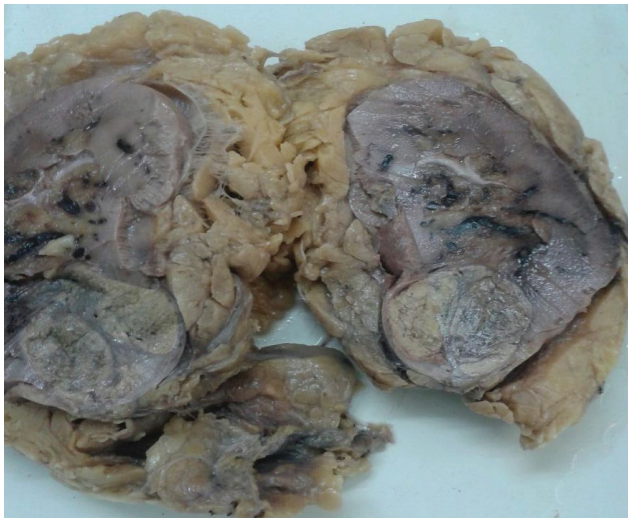

Fig. 1: Gross photograph of the tumour seen at the lower pole of kidney

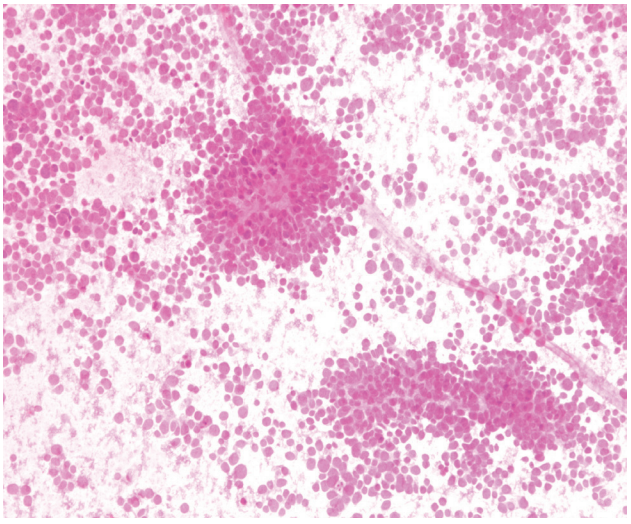

Fig. 2: Small round cells with scant cytoplasm arranged in micro-acinar arrangement (H &E, 40X)

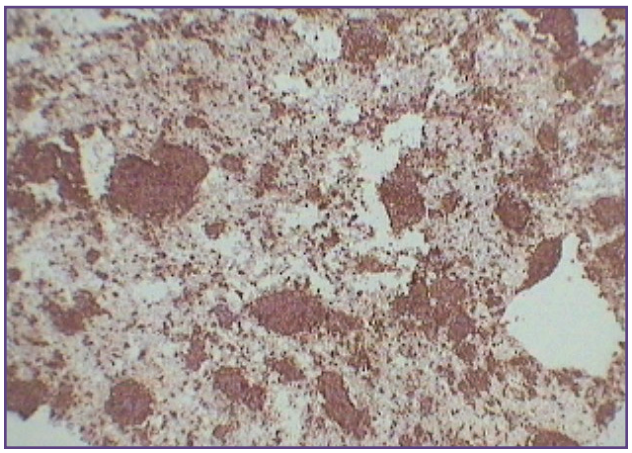

Fig. 3: Tumour cells showing strong positivity for MIC-1 (CD99, 40x)

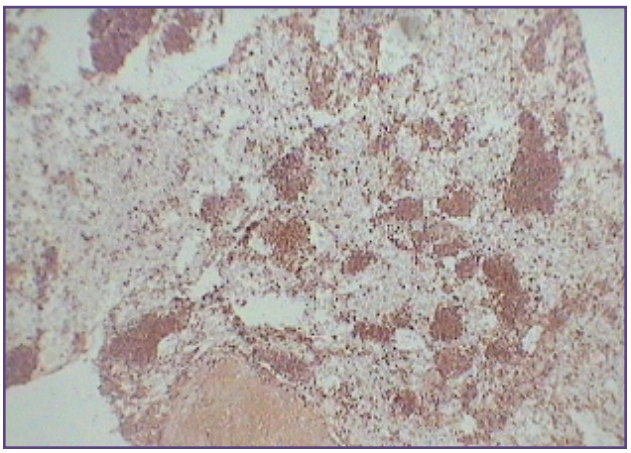

Fig. 4: Tumour cells showing weak positivity for NSE (IHC, 40x)

sarcoma), LCA (Lymphoma), NSE/chromogranin A (neuroblastoma) and CD-99 for PNET.[3,6] Though in some cases of PNET, the tumor cells may show NSE positivity as seen in neuroblastoma, the confirmation of PNET should be done by demonstrating CD99 positivity where the neuroblastoma cells are negative. CD99 positivity often are seen in synovial sarcoma which may be seen in kidney also. However, synovial sarcoma has characteristic morphology, bcl-2 marker and cytokeratin positivity. In our case, out of the custom IHC panel, Pan cytokeratin, Synaptophysin, Desmin and CD45 were negative. Moreover, cytogenetic studies (not performed pertaining to high cost of FISH to demonstrate different translocations related to PNET, synovial sarcoma, non-Hodgkin lymphoma, small cell carcinoma are corroborative .[3]

Renal PNET should be considered as a differential diagnosis of any rapidly enlarging mass presenting with local infiltration and clinical aggressive behaviour. Demonstration of reciprocal translocations (t11,22), (q24;12) is a very useful tool in the diagnosis.[7]

Conclusion

Exact diagnosis of PNET of kidney should be done using IHC and genetic markers to adopt appropriate management protocol with the hope of good clinical consequences. As the specific genetic defect can be identified, genetic therapy creating antisense oligonucleotides against the EWS-FLI-1 fusion gene will have good results to prolong the survival of patients. From our study, it can be suggested that in the setting of unavailability of molecular genetic analysis, clinicopathological features with IHC markers including suitable panel like CD99, cytokeratin, NSE, Desmin, LCA, bcl-2, can substantiate diagnosis of PNET of kidney.

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Competing Interests

Competing interests: None declared

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