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Primitive neuroectodermal tumor of kidney – A rare case with review of literature

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Abstract

Primitive neuroectodermal tumor (PNET) is a member of Ewing's sarcoma family of tumors which commonly occurs in bones. It rarely presents as primary renal tumor. This tumor is aggressive and has poor prognosis, hence should be differentiated from other small blue round cell tumors. We present a case of PNET of kidney in a 37-year-old female patient who presented with vague pain and lump in left loin since 4 months.

Keywords: Renal mass, Primitive neuroectodermal tumor (PNET), Immunohistochemistry.

Introduction

Extraosseous primitive neuroectodermal tumor (PNET) represents 6% of Ewing's sarcoma family of tumor. It was introduced by Tefft in 1969 [(1)]. PNET of kidney is a rare tumor with poor prognosis [(2)]. We report a case of PNET arising from left kidney in a 37-year-old female patient to highlight its rarity and aggressive nature.

Case Report

37-year-old, female patient presented with vague pain and lump in the left loin since 4 months. There was history of irregular fever, weight loss and hematuria.

CECT (Abdomen + Pelvis) revealed a well-defined large exophytic, complex, solid cystic lesion, involving lower pole of left kidney suggestive of neoplastic etiology.

Mass was resected and sent for histopathological examination.

Histopathological Examination

Received left kidney with a solid cystic mass at lower pole of kidney, measuring 14 x 12.5 x 7cm. Cut section of mass showed solid and cystic areas with variegated appearance (fig 1. a, b, c, d). Microscopy revealed a tumor tissue arranged in sheets showing monotonous, small, round nuclei with finely dispersed chromatin and scanty cytoplasm. At places tumor cells were arranged in rosettes (fig 2.a, b, c).

Immunohistochemistry revealed membranous diffuse positivity for CD99, nuclear diffuse positivity for FLI 1

and high index of Ki-67. Negative markers were WTI negative, chromogranin negative, LCA negative and CD 56 negative.

Considering the microscopic findings and IHC profile the tumor was diagnosed as primitive neuroectodermal tumor of kidney.

Discussion

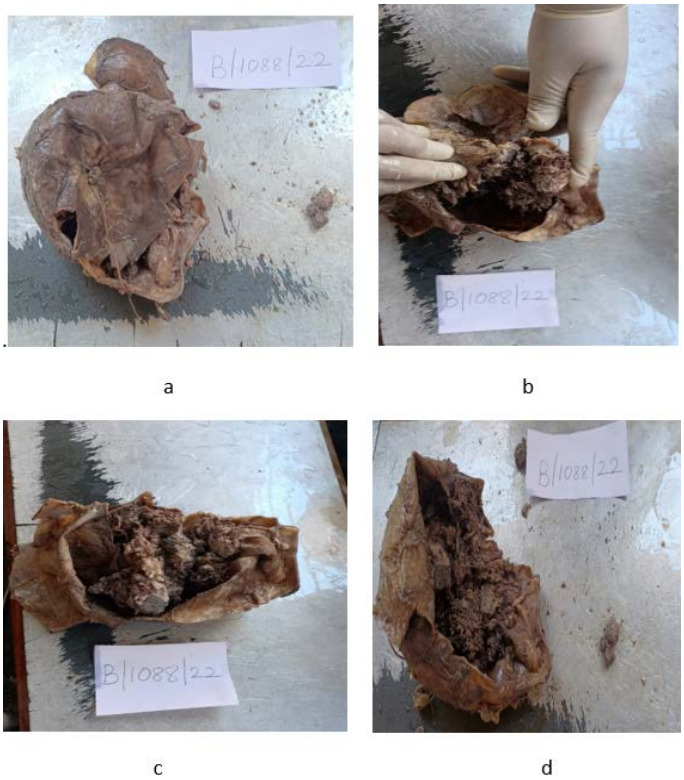
PNET forms a group of bone and soft tissue tumors derived from primitive neural crest cells which are found in bone or soft tissues of extremities and less commonly in the viscera or kidneys [3,4,5]. Primary PNET rarely presents as primary renal mass [6]. Most patients are young adults with no sex predilection, having aggressive clinical course [3]. Similar picture was seen in our case. The clinical presentation of PNET of kidney may be nonspecific like vague abdominal pain, low grade fever, cachexia and hematuria as observed in our case.

No specific signs of PNET have described on ultrasonography, CT or magnetic resonance imaging [5]. Definitive diagnosis of this tumor is done with histo pathological examination and immune histochemistry. Histopathological features of this tumor are monotonous proliferation of immature small round cells with formation of rosettes. Sometimes it is difficult to differentiate PNET from other small round cell tumors such as neuroblastoma, adult Wilm's tumor synovial sarcoma, malignant lymphoma and desmoplastic small round cell tumors [7]. PNET have chromosomal transaction t (11;22) (q 24; q 12) which lead to formation of EWL/ FLI-1 fusion protein diagnosed with RTPCR or FISH [3]. Our case also revealed FLI 1 positivity. PNET shows strong positivity for CD99. The Negative markers are cytokeratin (for nephroblastoma and synovial sarcoma), LCA (for lymphoma), NSE (for neuroblastoma). In our case positive tumor markers were CD99 and FLI 1. This tumor is highly aggressive and is often diagnosed in advanced stage already involving perinephric fat, hilar lymph nodes and renal veins or with distant metastasis. The 5-year disease free survival rate is 45 to 55% [8]. The recommended treatment for extraosseous PNET is with similar regimens as with PNET of bone as standard treatment for renal PNET has not been established because of rarity of this tumor [9].

Conclusion

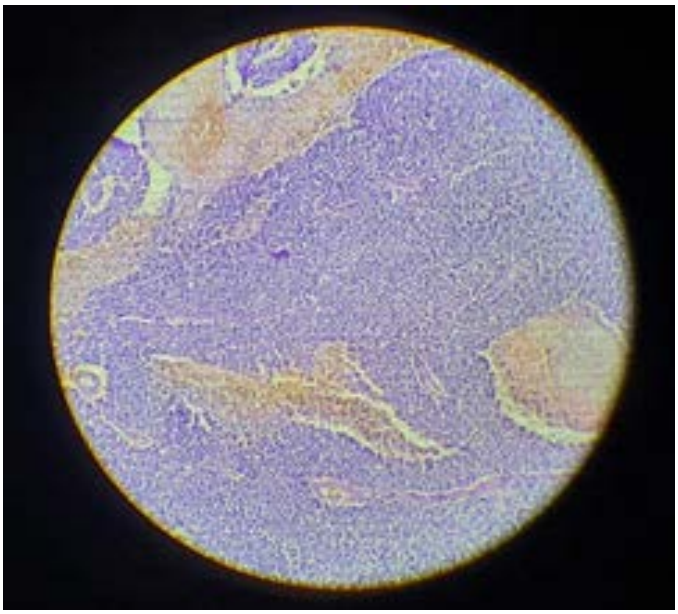
PNET of kidney is a rare tumor having aggressive nature and poor prognosis. Renal PNET should be differentiated from other small round cell tumors with the help of IHC as it has different therapeutic approach.

Figure 1.

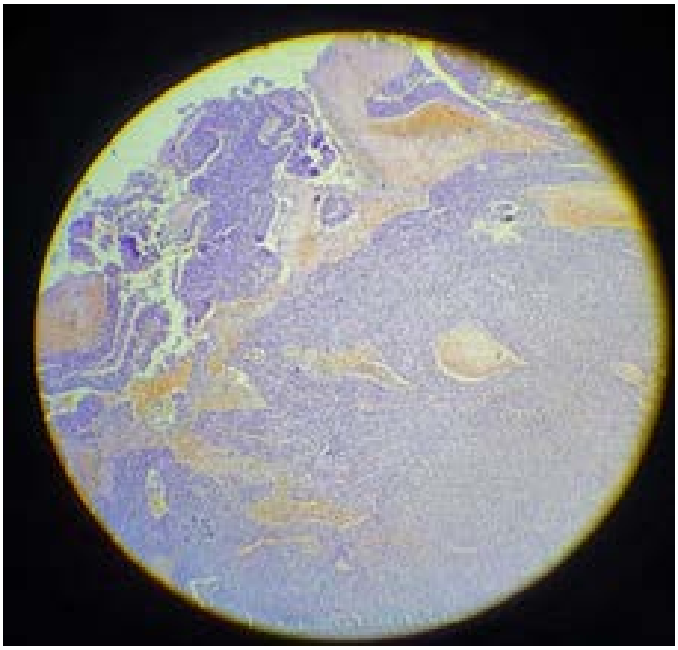


a b c d

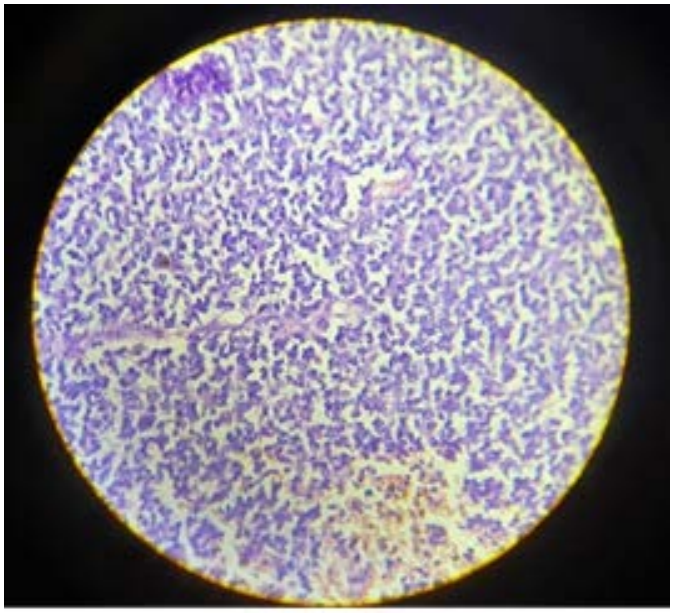
Figure 2.



(a)



(b)



(c)

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