

CASE REPORT

MALIGNANT SMALL ROUND CELL TUMOUR (PNET) OF PALATE: A RARE CYTOLOGICAL DIAGNOSIS

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ABSTRACT: Primitive neuroectodermal tumour is a malignant small round cell tumour affecting mainly the older children and adults. Palate is a very rare site for this tumour. We here report a case of 3 year male child who presented with a firm nodule of 2 x 1 cm size in the palate. Fine needle aspiration of the lump was performed and a cytodiagnosis of malignant small round cell tumour of palate was suggested. Subsequent histopathological examination supported it while Immunohistochemistry i.e. CD 99 positivity confirmed the diagnosis of PNET.

KEY WORDS: Malignant small round cell tumour, Primitive neuroectodermal tumour (PNET), Fine needle aspiration cytology.

INTRODUCTION: Primitive neuroectodermal tumour is a malignant small round cell malignancy of presumably primitive, neuroectodermal tissue or pluripotential, migratory neural crest cells arising from the soft tissue or bone, predominantly in older children and adults.¹ The cell of origin of this tumour is uncertain. The overall incidence rate of PNET is approximately 0.62 per million population in the U.S.A.² Palate is a very rare site for this tumour. The vast majority (> 90%) of tumours and tumour like lesions in the oral cavity are benign.^{3,6} Cancer of the oral cavity is extremely rare in children and adolescents. According to the SEER data, the age adjusted incidence of oral cancers for this population was 0.24 per million.^{7,8}

Here we present a cytodiagnosis of malignant small round cell tumour of palate which was confirmed on IHC as PNET.

CASE REPORT: A 3 year old male child presented to us with a firm nodule in palate of about 2 x 1 cm size. Cervical lymphadenopathy was absent. Hematological findings were normal except for the slightly raised total leucocyte count. The Peripheral blood film showed mild neutrophilia and no immature cells were found.

Fine needle aspiration was performed. The cytologic findings revealed cell rich smears comprising of dyscohesive sheets of monomorphic population of small and large round cells with scant pale-blue cytoplasm. The cells showed a high nucleo-cytoplasmic ratio. A cytodiagnosis of malignant small round cell tumour was suggested (Fig.1).

On the basis of above findings excision biopsy of nodule was done and the tissue was received for histopathological examination. Grossly, the tissue was grey- white measuring 2 x 1 cm. Microscopic examination revealed ulcerated mucosa overlying diffuse sheets of round to oval monomorphic cells with high nucleo-cytoplasmic ratio and stippled chromatin. Mitotic activity was evident to the tune of 8 per 10 high power fields. The histopathological features were consistent with cytodiagnosis of malignant small round cell tumour of palate (Fig.2).

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On Immunohistochemistry cells were positive for CD-99 and non-immunoreactive for LCA (Leucocyte common antigen) (Fig 3).

DISCUSSION: Malignant small round cell tumours are characterized by small, round, relatively undifferentiated cells. They generally include Ewing's sarcoma, peripheral neuroectodermal tumour, rhabdomyosarcoma, synovial sarcoma, non-Hodgkin's lymphoma, retinoblastoma, neuroblastoma, hepatoblastoma and nephroblastoma. Differential diagnosis of small round cell tumours is particularly difficult due to their undifferentiated or primitive character.

Fine needle aspiration cytology is successful diagnostic tool when used by a skilled cytopathologist for documenting primary and recurrent MSRCT's in pediatric patients.⁹ The exact categorization is not possible at the light microscopic level in the case of poorly differentiated tumours which poses a considerable challenge to the diagnostic skill of the cytopathologist.^{10,11}

Primitive neuroectodermal tumour (PNET) is a malignant small round cell tumour arising from the soft tissue or bone, predominantly affecting the older children and adults. The term PNET includes malignant small round cell tumours of the thoracopulmonary region (Askin's tumour),

Extra skeletal Ewing's sarcoma, peripheral neuroblastoma, and peripheral epithelioma.¹² The cell of origin of this tumour is uncertain. Originally, it was thought that EWS / PNET might arise from the neuroectoderm, but recent data have suggested that this tumour is more likely to originate from primitive stem cells and that the degree of malignancy appears to depend upon the stage of stem cell arrest during differentiation.¹³ The overall incidence rate of PNET is approximately 0.62 per million population in the U.S.A. Palate is a very rare site for this tumour. Cancer of the oral cavity is extremely rare in children and adolescents with age adjusted incidence for this population was 0.24 per million.

The present case highlights the palate as a rare primary site for malignant small round cell tumour (PNET) as well as importance of Fine needle aspiration cytology as an important diagnostic tool which equals to that of cumbersome histopathological examination.

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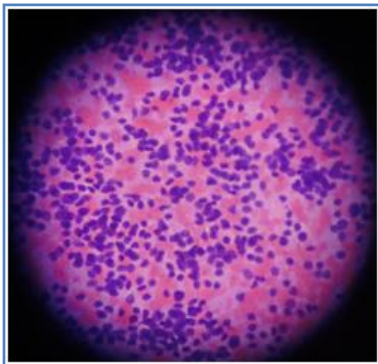


Fig. 1: Smears show heterogeneous population of small round cells (100X H& E)

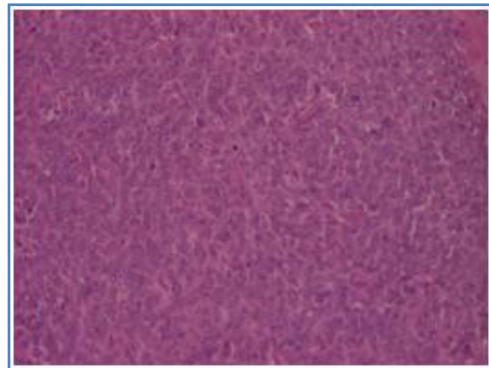


Fig. 2 Sections show small round cell tumour (100X H&E)

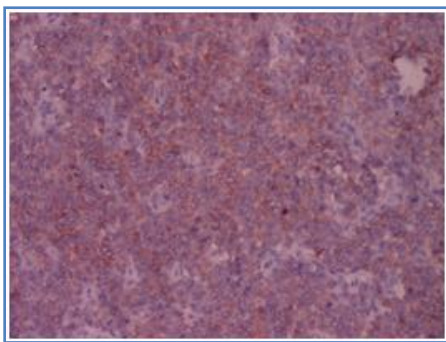


Fig. 3: On IHC CD 99 positivity

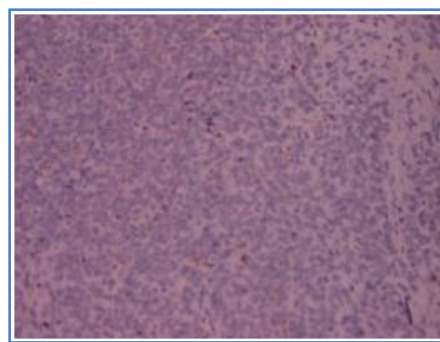


Fig.4: On IHC LCA negative

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