

PRIMITIVE NEUROECTODERMAL TUMOR OF TONGUE – A CASE REPORT

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ABSTRACT

A very rare case of Primitive Neuroectodermal tumor (PNET) occurring at the tip of the tongue is reported.

The patient presented at a private dental clinic at Hyderabad, Sindh, (Pakistan) with a tiny nodule at the tip of the tongue. Fine needle aspiration was carried out followed by excision biopsy and marker studies which confirmed the diagnosis.

It was concluded that the possibility of PNET tumor should be kept in mind while evaluating lesions occurring on tongue.

Key Words: Primitive Neuroectodermal tumor, tip of tongue.

INTRODUCTION

Oral cavity is a site for numerous diseases in children. Majority (80.3%) are inflammatory lesions. Others include tumor like lesions (40.5 %) and tumors. Benign tumors constitute 40.5% and Malignant tumors 9.5% in children.¹ The malignant tumors in children include round cell sarcomas (Primitive Neuroectodermal Tumors and Rhabdomyosarcomas), Hodgkin / Non-Hodgkin Lymphomas, Alveolar soft part sarcoma and fibrosarcomas.^{2,3} The Primitive Neuroectodermal Tumors (PNET) are of two types Central and Peripheral. Those arising in central nervous system are rare, highly malignant, cystic and sometimes hemorrhagic tumors.^{4,5} Peripheral PNET tumors occur in bone and soft tissues. They belong to Ewing's family of tumors which are associated with chromosomal translocation t (11; 22) and functional fusion of EWS gene to any of the several transcriptional factor genes.^{6,7 and 8} The diagnosis includes fine needle aspiration cytology followed by excisional biopsy and immunocytochemistry.⁹ Treatment includes maximal local surgical resection with chemotherapy and Radiotherapy.¹⁰ The prognosis is poor and few patients survive upto 2 years.¹¹

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Received for Publication: March 7, 2013

Accepted: March 30, 2013

CASE REPORT

A 13 years old school boy presented to a private clinic in June 2004 complaining of painless swelling on tongue. He had slight difficulty in speaking and eating. Clinical examination showed 1x1 cm firm, mobile, non-tender nodule at the tip of the tongue. The overlying mucosa was normal. A clinical diagnosis of retention cyst was made and Fine needle aspiration cytology was carried out. The smears showed a monotonous population of small, uniform, round cells with hyperchromatic to vesicular nuclei with little or no cytoplasm. The cells were slightly larger than lymphocytes. A provisional diagnosis of Non-Hodgkin Lymphoma, intermediate cell type was made. The patient was referred to Liaquat University of Medical and Health Sciences, Jamshoro for surgery. Excision biopsy was done with local flap reconstruction of the tongue and the specimen was submitted to Aga Khan University Hospital for histopathology and marker studies. The tumor cells were positive for glycogen. The sections were stained with a panel of monoclonal antibodies using Envision system. The tumor was found positive for Vimentin, EMA, MIC-2, BCL-2 (focal) and S-100 protein. Hence a diagnosis of malignant round cell sarcoma was made.¹² The differential diagnosis included PNET, Lymphoma and Rhabdomyosarcoma. Immunohistochemical studies were in favor of PNET. The child was followed up. There was recurrence of the tumor and the child developed widespread metastasis in lungs, liver and bones. He expired in 2 years.



Fig. 1: Clinical presentation of the PNET tumor at the tip of tongue.
Male: 12 years.

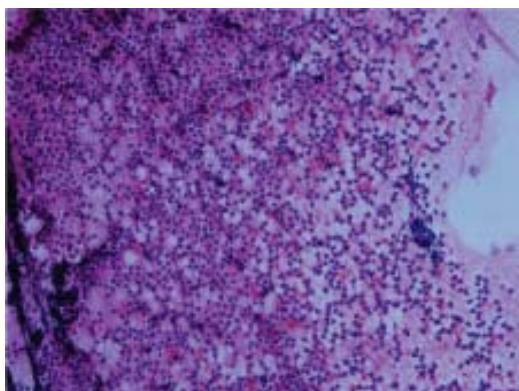


Fig 2: Microscopic picture of PNET tumor (Fine Needle Aspiration Cytology Smear) x 450

DISCUSSION

A solitary nodule occurring at the tip of the tongue in a child may be a retention cyst, a benign tumor like rhabdomyoma or granular cell tumor or a malignant tumor like lymphoma, PNET or Rhabdomyosarcoma. Although the most common malignant tumor of tongue is squamous cell carcinoma but it is extremely rare in children.

The diagnostic tools include FNAC followed by excision biopsy. Immunocytochemistry is required to differentiate between the various sarcomas.

Peripheral PNET are rare highly aggressive tumors. More common in males and the age group is 10-20 years. Annual incidence in USA is 2.1 cases per million children.⁸ They clinically appear as solitary well defined nodules and rapidly increase in size. Histopathology shows small anaplastic cells with mitotic activity and rosette formation. Although most are completely undifferentiated, they may show neuronal or glial differentiation. The Primitive neuroendocrine tumors are positive for Vimentin, EMA, MIC-2, BCL-2 and S-100 protein. The patient needs CT scan, MRI and Bone scan to exclude metastasis at the time of diagnosis. Treatment includes surgery followed by Radiotherapy and Chemotherapy. The patient needs regular check ups and clinical follow up. The prognosis

of PNET is poor and few patients live beyond two years.

In the literature reported rare benign tumors occurring at the tip of the tongue include Schwannoma and Neurilemmoma.^{13,14} Few malignant PNET tumor occurring at the tip of the tongue have been reported in the literature.¹⁵ The patient was treated by giving Radiotherapy and Chemotherapy after surgical removal. Multiple metastases occurred in liver within 10 months. The child survived for 2 years.

CONCLUSION

Peripheral primitive neuroectodermal tumors are rare but they do occur at unusual locations like tip of the tongue. The possibility of PNET should be kept in mind while evaluating lesions occurring on tongue.

REFERENCES

- Ulmansky M, Lustman J and Balkin N. Tumors and Tumor like lesions of the Oral Cavity and related structures in Israeli Children. *Int. J Oral and Maxillofacial Surg* 1999; 28(4): 291-94.
- Trobs RB, Mader E, Friedrich T, Bennek J. Oral tumors and tumor-like lesions in infants and children. *Pediatr Surg Int*. 2003; 19(9-10): 639-45.
- Lingual alveolar soft part sarcoma in a child managed successfully with surgery and chemotherapy. *Indian J Cancer* 2010; 47(2): 234-35.
- Ashwal S, Hinshaw DB, Bedros A. CNS Primitive Neuroectodermal Tumors of Childhood. *Medical and Pediatric Oncology* 1984; 12 (3): 180-88.
- Hart MN, Earle KM. Primitive Neuroectodermal Tumors of the Brain in children. *Cancer* 1973; 32(4): 890-97.
- Dehner LP. Primitive Neuroectodermal Tumor and Ewing Sarcoma. *Am J Surg Pathol* 1993; 17(1): 1-13.
- Alva E, Gerald WL. Molecular Biology of Ewing's sarcoma/ Neuroectodermal tumor family. *J Clin Oncol* 2000; 18 (1): 204.
- Grier H. The Ewing Family of Tumors Ewing's Sarcoma and Primitive Neuroectodermal tumors. *Pediatric Clinics of North America* 2009; 449 (4): 991-1004.
- Daskalopoulou D, Rapidis AD, Maounis N, Markidou S. Fine needle aspiration cytology in tumors and tumor like conditions of Oral Cavity and Maxillofacial regions. *Cancer Cytopathology* 1997; 81(4): 238-52.
- Zimmerman MA, Goumnerova LC, Proctor M, Scott RM, Marcus K, Pomeroy SL et al. Continuous Remission of newly diagnosed and relapsed central nervous system atypical Teratoid/Rhabdoid tumor. *Journal of Neuro-Oncology* 2005; 72(1): 77-84.
- Ultin C, Cetinayak O, Aksu G, Fayada M, Ataergin S, Beyzaoglu M. Auris Malignant Peripheral Neuroectodermal (p PNET) of Tongue. *Nasus Larynx* 2007; 34(1): 115.
- Memon GA, Memon AR, Siddiqui R. Primitive Neuroectodermal tumor of Tongue. *JLUMHS* 2004; 3(2): 297-98.
- Craig RDP. Neurilemmoma of the Tongue. *Arch. Dis. Childh* 1964; 39: 297-98.
- Verma RK, Dhingra S, Gupta K, Panda NK. Lingual Schwannoma – a case report. *Oral Surg* 2011; 4(2): 82-85.
- Sethi B, Smith GT. Primary Primitive Neuroectodermal tumor arising in small Bowel. *Histopathology* 2007; 50(5): 56-60.