Case Report

PRIMITIVE NEUROECTODERMAL TUMOR OF TONGUE

Ghulam Ali Memon, Abdul Razzak Memon and Raisuddin Siddiqui

ABSTRACT

We report a highly rare case of primitive neuroectodermal tumor of tongue. A 13-years old male presented with progressive lesion at the tip of tongue and the left lateral border since six months. Excision biopsy of the patient was done with local flap reconstruction of the tongue. Histopathology favoured the diagnosis of primitive neuroectodermal tumor.

KEY WORDS: *Tumor. Neuroectodermal. Tongue. Biopsy.*

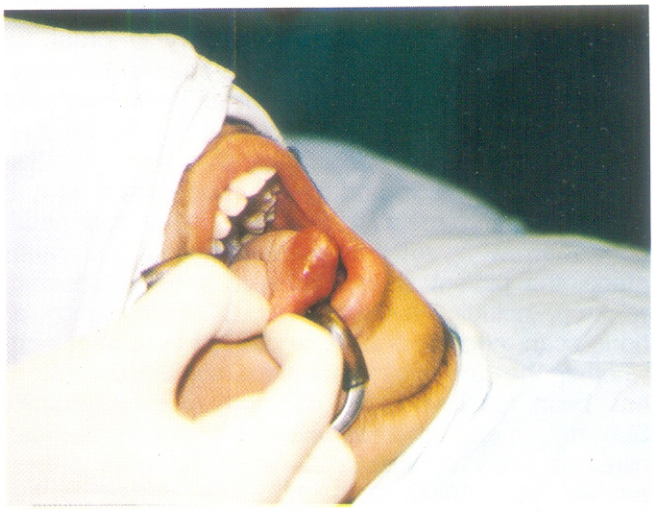
INTRODUCTION

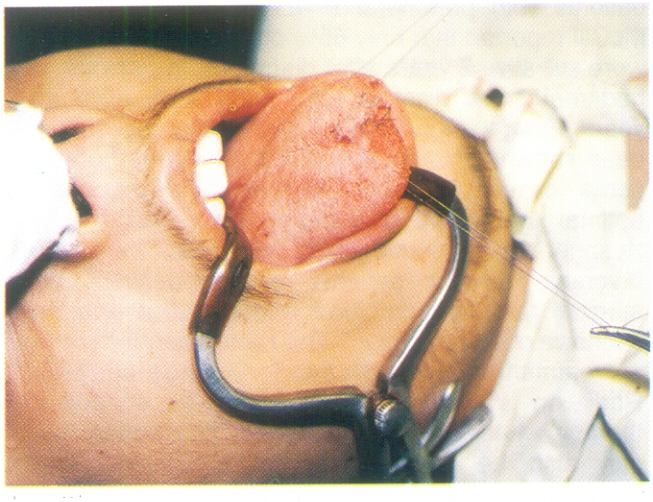
Primitive Neuroectodermal Tumor (PNET) is a soft tissue sarcoma classically described under small round blue cell tumors (SRBCT). Their incidence has been reported as 1% of all sarcomas and majority (75%) of these cases occurs in <35 years age range[1]. Some PNETs occur in the brain while the others (the peripheral PNETs) occur in sites outside the brain. Stout initially described PNETs in 1918[2]. At first, these tumors were found arising from major nerves[3,4]. Later, these tumors were reported in the chest wall, retroperitoneum, extremities, bone and paraspinal sites[5,6]. Today, this tumor is believed to be a neoplasm of nonneural soft tissue that primarily affects children and adolescents[7]. In this paper, we present a highly rare case of PNET of tongue.

CASE REPORT

A 13-years old male belonging to Hyderabad presented at Liaquat University Hospital Jamshoro, Pakistan in July 2004 with progressive lesion at the tip of tongue and the left lateral border since last six months. He had no history of bleeding, ulceration or pain but had little difficulty in eating and speaking **(Figure I)**. His previous fine needle aspiration report had showed non-Hodgkin's lymphoma. His excision biopsy was done with local flap reconstruction of the tongue **(Figure II)**. Pathology report showed that lesion is completely excised and tumour cells show positivity for glycogen. The sections were stained with a panel of monoclonal antibodies using envision system and Vimentin, EMA, MIC-2, BCL-2 (focal) and S-100 protein were found positive. Hence, a diagnosis of malignant round cell tumor was made. The differential diagnosis in this case included primitive neuroectodermal tumor, synovial sarcoma and rhabdomyosarcoma. After exhaustive immunohisto-chemical working, features were in more favor of PNET. Patient is followed for last four months with no local or regional recurrence of the disease.

FIGURES I AND II:
PRE AND POSTOPERATIVE PRESENTATION OF TONGUE OF THE PATIENT





DISCUSSION

The actual incidence of PNETs is difficult to ascertain because diagnostic criteria have recently been delineated[8,9]. These tumors can occur in any age group, but mostly occur in adolescents. PNETs are typically grouped with other round, small-cell tumors including Ewing's sarcoma, neuroblastoma, lymphoma and rhabdomyosarcoma. There is no apparent predilection for either sex. Among the cell lines that might be responsible for the development of PNETs, there are three likely candidates: the neural crest, primordial germ cells and uncommitted mesenchymal cells. The exact classification of these rare neoplasms has been a source of continued controversy[3,9-13]. Dehner had reviewed the contemporary medical literature and discovered a distinction between a central PNET and a peripheral PNET[10]. These tumors are most often found in the chest, pelvis, abdomen, and extremities[6,14,15]. Light microscopic examination generally reveals that PNETs contain sheets of small, round to oval cells with a scant amount of cytoplasm and coarse chromatin material. The presence of .. rosette formation is believed to be necessary in order to make the diagnosis[12]. Immunohisto-chemical studies have improved ability to differentiate these tumors. These tumors cannot be diagnosed solely on the basis of radiographic techniques. Peripheral PNETs are highly aggressive and have a propensity for local recurrence and metastasis to the lung, bone and bone marrow[8,15]. After a tissue diagnosis has been made, the patient should undergo a full metastatic work-up, including a chest x-ray, CT of the lungs, a bone scan, and bone marrow aspiration to ascertain whether the tumor has metastasized. Bone erosion has been reported to be a common initial finding. Similarly, many affected patients have metastatic disease; rates of metastasis in three large series ranged from 20 to 31%[8,14,15]. The prognosis for patients with peripheral PNET is poor. Jones and McGill reported that only 65% of patients they studied were still alive 2 years after diagnosis. In patients who had metastatic disease, only 38% were alive at 2 years. Even though PNET is highly aggressive, it has been shown to be curable with multimodal therapy in some cases. Because of their rare occurrence, optimal therapy is challanging. In a large series, Kushner et al found that outcomes were more favorable among patients who underwent early surgical removal combined with radiation and dose intensive chemotherapy[14]. The recommended manner of treatment is similar to that for Ewing's sarcoma[15]. Radiation is generally administered as an adjuvant therapy when surgical excision is incomplete, but it can also be used as a primary treatment for unresected lesions. Although PNETs are believed to be radiosensitive, radiation is not adequate to cure macroscopic disease.

In conclusion, PNET is an aggressive tumor that is rarely encountered. Diagnosis depends on extensive histologic evaluation, including immunohistochemistry and electron microscopy. Treatment should include aggressive surgical resection combined with adjuvant radiation and chemotherapy. Because the rate of distant metastasis at the initial diagnosis approaches 30%, preoperative evaluation of the chest, bone, and bone marrow is warranted[8,14,15]. Radiation and chemotherapy have palliative value in unresectable cases. Heightened awareness and early intervention might result in a better chance for a good outcome in patients with this aggressive tumor. In these tumors, follow up of longer duration is also usually required.

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