IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 15, Issue 5 Ver. VIII (May. 2016), PP 45-48
www.iosrjournals.org

Primitive NeuroectodermaltumorOf The Maxillary Sinus In A Young Female: A Case Report And Literature Review

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Abstract:*Primitive neuroectodermaltumor (PNET), which belongs to the Ewing's sarcoma (ES) family of tumors, is mainly seen in children and young adults. PNETs are extremely rare in the maxilla. Here, we report a case of a 15y old female diagnosed with PNET of maxilla following detailed radiologic,histopathologic, immunohistochemicalstudies. Though the imaging features of PNET are non-specific and definitive diagnosis is only by immunohistochemistry, PNET should be included in differential diagnosis of fast growing soft tissue tumours of children and young adolescents.*

Keywords: *NeuroectodermalTumors, Primitive; Maxillary Sinus*

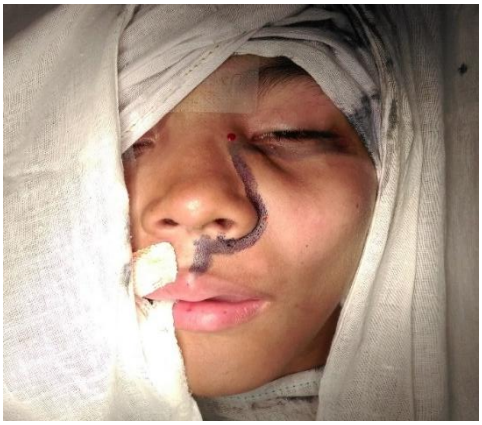
I. Introduction

Primitive neuroectodermaltumors (PNETs) are a type of small round cell tumors developing from migrating embryonal cells of the neural crest. It is classified into two types, based on location in the body: peripheral PNET and CNS PNET.Peripheral primitive neuroectodermaltumors (pPNETs) are rarewith varying incidence of occurrence in head and neck region. Primitive neuroectodermaltumors (PNETs) and Ewing's sarcoma belong to the family of soft tissue sarcomas. PNETs and Ewing's sarcoma share unique chromosomal translocation t(11;22)(q 24;q 12) leading to the formation of the EWS-FLI-1 fusion protein. They present clinically with localized pain and a mass, with or without generalized symptoms of fever, anaemia, and malaise; and on histopathology, as small round blue cell tumor. It is a rare tumor, usually occurring in children and young adults under 25 years of age. PNET is a very aggressive malignancy with a poor survival rate.

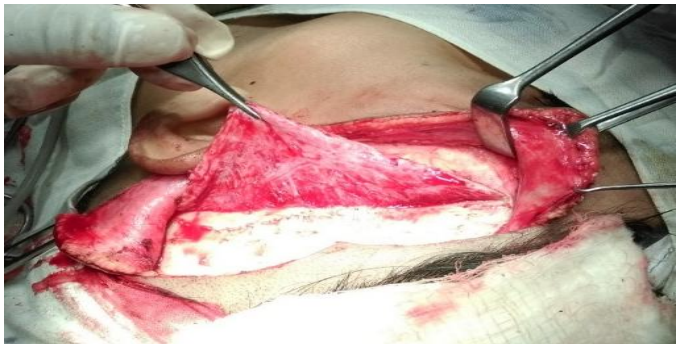
II. Case Report

A 15year old female presented to our department with swelling of the left cheek and near the left third molar tooth since 2 years. On further evaluation the patient provided with further information that there was proptosis of the left eye one year back. On examination a diffuse swelling was seen on the left cheek of about 3*3 cm with an apparently normal overlying skin. Intraorally a firm, immobile swelling of 1*1 cm was seen near the left 3rd molar. No cervical lymph nodes were palpable. Contrast enhanced computed tomography revealed a heterogeneously enhancing soft tissue attenuated growth involving the left maxillary sinus and the left nasal cavity with destruction of the medial walls and the floor of maxillary sinus. Punch biopsy revealed malignant round cell tumour. On immunohistochemistry, the tumor cells express mic-2(CD 99) and are immunonegative for desmin(marker for rhabdomyosarcoma), myogenin, synaptophysin, chromoganin A, cytokeratin(marker for carcinoma), EMA, CCA and TdT.

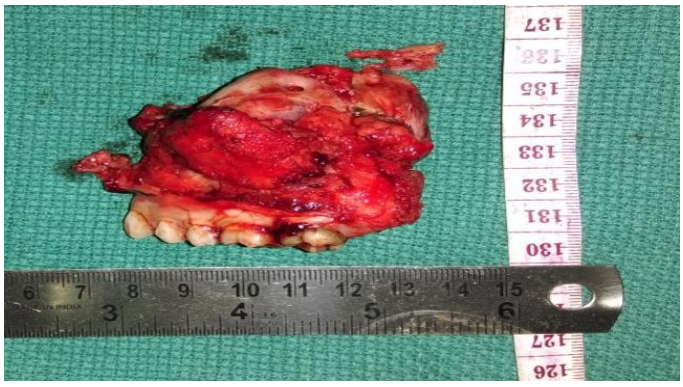
The diagnosis of PNET was made. The patient was given 31 cycles of chemotherapy with Ifosphamide, vincristine, etoposide, actinomycin and doxorubicin in combinations followed by 32 cycles of radiotherapy. Following chemoradiation a computed tomography scan was done which revealed a non-enhancing lesion in the left maxillary sinus buldging into the nasal cavity with sclerosis and irregular destruction of medial wall, focal erosion in the adjacent hard palate on left side and erosive changes in the anterior and inferior walls of the left maxillary sinus including the alveolar margin. The patient was planned for surgery under general anaesthesia. The patient underwent a left maxillectomy and reconstruction with temporalis muscle rotation flap in our hospital. Specimen was sent for HPE which revealed tissue composed of dense fibrcollagenised tissue with areas of calcification and mild lymphocytic infiltration. Patient was discharged 10 days after the operation with no immediate postoperative complications. She was followed up to1 year and there were no postoperative complications or recurrence of disease during this period.



Preoperative



Temporalis muscle graft



The tumour mass

One year follow up

III. Discussion

PNET is a raretumor in the head-neck region. Jürgenset al[2] reported 42 cases of PNETs. Of these, only 4 were head-neck PNETs. In their series of 54 cases, Kushner et al[5] found only 1 case of head-neck PNET. However, Jones and McGill[4] reported that the head-neck region was the second most common site for PNET after the thoracopulmonary region. Incidence of PNETs in the maxilla is even rarer. Of the 11 cases studied by Jones and McGill,[4]only 1 arose in the maxilla. Windfuhr[1] made an excellent review of 27 cases of head-neck PNETs. In the report, only 6 cases occurred in the maxilla. However, after an extensive search of the existing Englishlanguage literature, we could identify 10 reported cases of PNETs of maxilla.[1,3,5]PNET has been reported to occur in young people.One study[1] reported that 85% of head-neck PNET patients were younger than 20 years of age. Of the 10 reported cases of maxillary PNETs that we identified, 6 patients were under the age of 20 years. We did not find any sex predilection in the maxillary PNETs. This finding was consistent with previous reports. Most of the reported cases of PNETs of the maxilla occurred either as a soft tissue mass or swelling.

Like those of PNETs of other body parts, radiological findings of head-neck PNETs were non-specific.[1,3,4]On plain radiographs, they appear as areas of bone destruction. On CT images, the PNETs we found appeared as heterogeneously enhancing soft tissue mass. On T1weighted MRI, these tumors have isointense presentation to muscle, whereas on T2-weighted MRI, PNETs show a heterogeneous hyperintense signal. In our case the initial biopsy revealed malignant round cell tumour. In our opinion, the lesion arose from the mucosal lining of the maxillary sinus. However, we did not rule out the bony area surrounding the maxillary sinus as the possible site of origin. Once developed, the lesion infiltrated the maxillary alveolar bone, which resulted in mobility of the maxillary molar tooth. On immunohistochemical examination the tumour cells expressed CD99(mic22) which is highly expressed in all peripheral PNETs. Ibarburenet al[6]studied the CT and MRI findings of 17 patients with peripheral PNET. They reported that both CT and MRI are very useful in preoperative staging and in planning the surgical approach. CT and MRI were also helpful in the detection of recurrent and metastatic disease.

Due to their highly unpredictable behavior, the management of PNETs is often challenging. Thus far, no standard treatment protocol has been developed for PNETs.[1] The current approach for the management of these tumors includes a combination of therapeutic modalities like early surgery along with multiple chemotherapy to treat the residual disease and to prevent metastatic or recurrent disease.[1,4,5]Multi-agent chemotherapy including vincristine, doxorubicin, and cyclophosphamide improves survival without any significant morbidity.[7]Zimmermman et al[7]reported that chemotherapy is mandatory as the first-stage treatment in order to avoid a mutilating surgical procedure and intraoperative tumor cell dissemination. This treatment may be supplemented by radiotherapy, which should be given to patients who do not have a surgical option that restores physiology and to patients whose tumor has been excised with an inadequate margin.[1] Given the particularly aggressive nature of PNET, effective local control requires tumor-free surgical margins and wide safety margins.However, such margins may not always be available in the maxillary region because of its proximity to vital structures. In the present case, our patient underwent initial chemoradiation reducing the size of the tumour giving us adequate tumour free margins during surgery.

Although steadily improving, the prognosis of PNET is generally considered to be poor with a high incidence of rapid metastasis to distant sites such as the lung, liver, and bone. Jones and McGill[4]reported that 27% patients with head-neck PNET had metastatic disease at the time of presentation. Prognosis of PNET mainly depends on tumor location, tumor size, and presence of metastatic disease at the time of initial diagnosis, and response to initial chemotherapy. The reported 3-year survival rate is about 50%.[2,3]Survival in the study of Mendel et al[8]was an average of 27.3 months for asymptomatic patients and 15.2 months for symptomatic patients from the time of initial diagnosis. In the cases of maxillary PNET we reviewed, [1,3,4]elderly patients seemed to have a relatively poor prognosis compared with their younger counterparts although only 3 patients had undergone follow-up for 3 years at the time of the study.

PNET is a very aggressive malignancy with a poor survival rate. It should be included in the differential diagnosis of fast-growing soft tumors, particularly those that have imaging features of high cellularity.

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