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CASE REPORT

Primitive Neuroectodermal Tumor of Maxilla: A Rare Case

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ABSTRACT

Primitive neuroectodermal tumor (PNET) is an aggressive round cell malignancy of presumed neural crest origin belonging to Ewing's sarcoma family of tumors. Peripheral PNET is less common in head and neck region and only eight cases of primary PNET of maxilla have been reported. We report a case of 3-year-old boy diagnosed with PNET of maxilla based on detailed radiologic, histopathologic, and immunohistochemical studies. Though the imaging features of PNET are nonspecific and definitive diagnosis is only by immunohistochemistry, PNET should be included in the differential diagnoses of fast growing soft tissue tumors of children and young adolescents. We emphasize the need for its early diagnosis and prompt management owing to its aggressive nature and high mortality rate.

Keywords: Neuroectodermal tumor, Primitive, Maxilla.

INTRODUCTION

Tumors of head and neck region can vary from simple eruption cyst to aggressive lymphomas. Majority of these lesions are inflammatory in nature, but other etiologies include congenital, benign neoplastic and malignant lesions. When such lesions occur in oral cavity, general dentists or the pedodontists are usually consulted by parents of such children. This forms an important task at the level of primary clinician to distinguish between normal soft tissues and lesions that are manifestations of potentially life-threatening conditions, so that early and prompt referral is advised. The complex anatomic constraints of head and neck region can raise serious concerns for malignancy either because of compression or erosion of adjacent structures or due to dramatic symptomatology, like facial swelling, proptosis, etc[1]. Sound clinical judgment based on occurrence, biological behavior and prognosis can aid in better counseling regarding the need for early management.

Primitive neuroectodermal tumors (PNET) are small round cell tumors of presumed neural crest origin arising outside the central and sympathetic nervous system[2] with variable differentiation belonging to the family of Ewing's sarcoma and were first described by Hart and Earle in 1973.[3] Though primitive neuroectodermal tumors occur predominantly in central nervous system, peripheral PNETs have been reported.[4] Peripheral PNETs are commonly seen in chest wall, trunks, abdomen and extremities of children and are relatively rare in head and neck region.[5-7] Primary peripheral PNET of maxilla is a rare occurrence and only eight cases have been reported in English literature so far.[8]

CASE REPORT

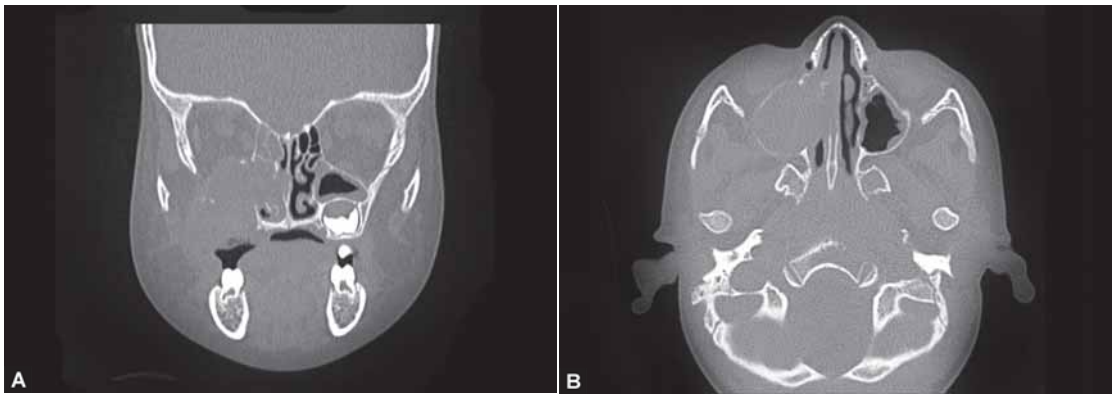
A 3-year-old-boy was brought to the department by his parents with the complaint of loose tooth and growth in the right upper jaw since 10 days. Patient had initially complained of difficulty in eating and parent observed a loose tooth which was extracted by the parent himself. Intraoral growth progressed in size after extraction of tooth. There was history of decreased diet and recurrent episodes of cold and fever since 8 to 10 months.

On examination, the child was moderately built and poorly nourished, active and playful. Diffuse swelling was noted extraorally on the right cheek region extending from infraorbital margin superiorly till lower border of mandible inferiorly and from corner of mouth to 2 cm in front of tragus of ear (Fig. 1A). Skin over the swelling was apparently normal. Intraorally, a sessile growth was noted measuring 6 × 4 cm extending anterioposteriorly from deciduous canine to the maxillary tuberosity. The lesion extended buccopalatally and appeared bilobed due to indentations of mandibular teeth over the swelling (Figs 1B and C). Mucosa over the swelling appeared hyperemic without any ulceration. The lesion was soft to firm in consistency.

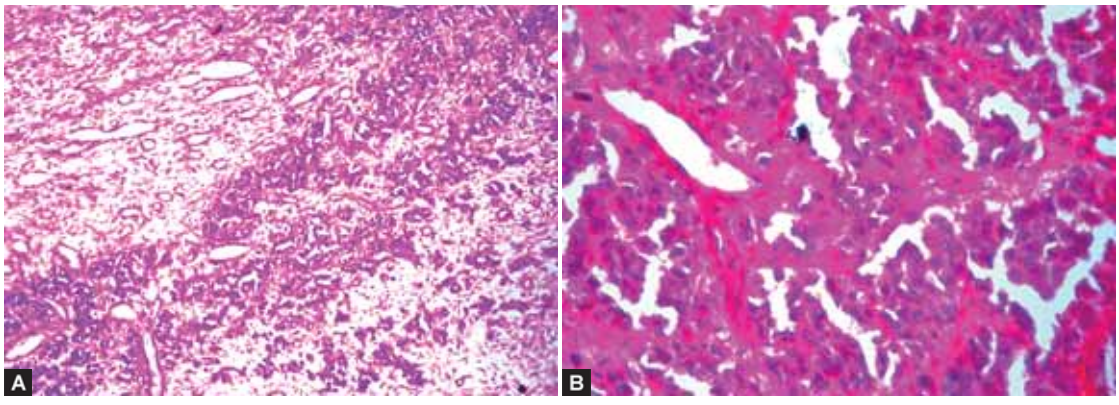
Right submandibular lymph node was palpable, firm, tender and not fixed. Cervical, axillary and femoral group of lymph nodes were not palpable.

Orthopantomograph and paranasal sinus view did not contribute any significant information and a computed tomography (CT) of maxilla was obtained which revealed a soft tissue density mass involving right maxillary sinus approximately measuring 4 × 3.2 × 2 cm (Figs 2A and B). The lesion extended superomedially into the right ethmoidal air cells, medially into the right nasal cavity and inferiorly into infratemporal fossa, and invade the body of maxilla and superior alveolar rim on the

Figs 1A to C: (A) The extent of tumor on initial presentation, (B) intraoral extent of the tumor, (C) tumor extent after 4 months



Figs 2A and B: (A) Coronal view of computed tomography scan showing tumor extent, (B) axial view



Figs 3A and B: (A) Photomicrograph (hematoxylin and eosin stain; 10×) showing malignant small round cell tumor with prominent vascularity, (B) photomicrograph showing (40×) clusters of cells separated by dense fibrous septae and round to oval hyperchromatic cells with scanty eosinophilic cytoplasm

right side. There was evidence of protrusion of mass lesion into the orbit with the erosion of underlying orbital floor. The fat plane between the mass lesion and pterygoid masses appeared to be maintained. The blood parameters were normal except mild neutrophilia. Systemic evaluation did not reveal any abnormality.

A provisional diagnosis of non-Hodgkin's extranodal lymphoma was made based on the clinical presentation of a rapidly progressing lesion. Differential diagnosis included rhabdomyosarcoma, PNET, olfactory neuroblastoma, and central giant cell granuloma.

Incisional biopsy with hematoxylin and eosin staining revealed a highly vascularized round cell tumor (Figs 3A and B). Further immunohistochemical studies favored a diagnosis of primitive neuroectodermal tumor of infancy using tumor marker of S-100. CT thorax revealed multiple pulmonary, pleural, skeletal and hepatic metastasis. A treatment plan of chemotherapy and radiotherapy extending over a period of 8 months was planned. The treatment was deferred by the parents of child and was treated symptomatically. In due course of 6 months, the patient succumbed to the disease.

DISCUSSION

Differential diagnosis for rapidly growing tumors of head and neck region includes malignant lymphoma, leukemia, rhabdomyosarcoma, neuroblastoma, undifferentiated nasopharyngeal carcinoma, malignant melanoma, PNET and Ewing's sarcoma family of tumors.[2]

Peripheral PNET are less common and constitute 1 to 4% of all the soft tissue neoplasm.[9] PNET have been reported to occur in mediastinum, retroperitonium, pelvis, trunks or extremities, head and neck region and small bowel. PNETs predominantly occur in children and young adults but can develop at any age. The lesions have an insidious onset, leading to large sizes at presentation. Distant metastases are identified in 20 to 25% of newly diagnosed patients and are the most important adverse prognostic factors, especially bone and bone marrow metastases.[10,11] The incidence of PNET is likely to be underreported in the past due to lack of immunohistochemistry that distinguish it from other round cell tumors.[12] The treatment protocol is similar to that of Ewing's sarcoma family of tumors consisting of chemotherapy and surgery for locoregional control of disease. Radiotherapy may be particularly useful for cases which are inoperable or not well responding to neoadjuvant chemotherapy.[13] 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.[5] Though the imaging features of PNET are nonspecific and definitive diagnosis is only by immunohistochemistry, PNETs should be included in the differential diagnoses of fast growing soft tissue tumors. Early diagnosis is a key in the management of PNET as it is a very aggressive tumor with poor survival rate.

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