Melanocytic neuroectodermal tumor of infancy

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ABSTRACT
Melanocytic neuroectodermal tumor of infancy is a relatively rare pigmented neoplasm that primarily affects the maxilla of infants. Pigmentation depends upon the amount of melanocytic element present. We report a case of melanocytic neuroectodermal tumor in a three month-old child with a greater extent of neuroblastic differentiation.

KEY WORDS: Melanocytic neuroectodermal tumor of infancy, melanotic progonoma, tumor of neural crest origin

INTRODUCTION
First described by Krompecker in 1918, melanocytic neuroectodermal tumor of infancy (MNTI) is a rare tumor usually occurring below the age of one year. The majority of these tumors are slow growing, benign tumors with a 2% chance of malignant differentiation. Although computed tomography (CT) scan / magnetic resonance imaging (MRI) can help, histopathology of the tumor is characteristic and is used to confirm the diagnosis.

CASE REPORT
A three month-old male child presented with a swelling arising from the oral cavity since 1½ months, difficulty in breast-feeding and irritability. On examination, surprisingly the baby was well nourished and comfortable inspite of the huge oral swelling [Figure 1]. His breathing and other vital parameters were normal. On local examination, 8 × 6 cm-sized, pinkish-red mass was seen arising from the oral cavity. This mass was fixed, hard, nontender and mainly on the left side of the mouth. Posterior and lateral margins of the mass could not be delineated. There was no cervical lymphadenopathy. CT scan of the Head and Neck was done which showed a well-circumscribed radiolucent lesion with bony destruction arising from the maxilla [Figure 2].

The histopathology report of the knife biopsy was of melanocytic neuroectodermal tumor with a greater extent of neuroblastic differentiation [Figure 3]. Due to the large lesion, the patient was started on neoadjuvant chemotherapy consisting of vincristine, cyclophosphamide and dactinomycin.

There was about 10% regression in the mass after two weeks. Because of the poor response to chemotherapy, local therapy was planned. The treatment options of surgery and/or radiotherapy along with their related morbidity were explained and discussed with the parents. But the parents opted not to take further treatment in view of the morbidity as well as the possibility of recurrence.

DISCUSSION
MNTI was first described by Krompecker in 1918 as a congenital melanocarcinoma. It was known by many names as its cellular origin was not clear. These names included pigmented ameloblastoma, retinal anlage tumor, melanotic adamantinoma, retinal choristoma, melanotic progonoma, melanotic epithelial odontoma, pigmented teratoma, atypical melanoblastoma, pigmented epulis and retinoblastic teratoma. Some authors proved that this tumor causes a high urinary excretion of vanillylmandelic acid (VMA), suggesting a neural crest origin. Hence, they coined the term "melanocytic neuroectodermal tumor of infancy".[1]

About 200 cases of MNTI have been reported till now. Characteristically it arises from maxilla, more so from the intraoral side.[2] Bone destruction and displacement of teeth often occur because of the intraosseous location in the maxilla. Other sites are the skull, mandible and the brain.[3] The lesion is usually solitary and the mucosa over
The lesion is usually intact. It is typically bluish in color due to the presence of melanin. Although it is a benign tumor with 2% chance of malignancy, it is locally aggressive. The majority of MNTI patients (there is no sexual predilection) present in the 1st year of life. The children present with swelling in the oral cavity, which often hinders feeding. The differential diagnosis is ameloblastoma, odontoma, odontogenic myxoma, fibroma, rhabdomyosarcoma, Ewing's sarcoma, Langerhans' cell histiocytosis (LCH), non-Hodgkin's lymphoma.

The plain radiograph of MNTI shows a well-circumscribed radiolucent lesion. As the tumor advances, it destroys the bone suggesting a malignant process. In its typical premaxillary position, the tumor can displace or destroy the developing dentition. CT scan with intravenous contrast is often used to delineate the margins of osseous involvement. Additionally, MRI can be used to evaluate the bony extent of the lesion. Most MNTIs appear as typical soft tissue tumors with nonenhancing heterogeneous tissue density.

Histopathology shows biphasic pattern with the larger pigmented, melanocyte-like cells and smaller, nonpigmented neuroblast-like cells. Immunohistochemistry (IHC) is positive for cytokeratin, synaptophysin, HMB45, NSE, epithelial membrane antigen, glial fibrillary acidic protein and Leu-7.[4]

The treatment of choice in MNTI is usually complete surgical excision. This treatment can usually be accomplished with a partial maxillectomy by using a Weber-Ferguson incision and a facial degloving approach. The adjacent bone and developing teeth must be sacrificed to get an at least 5 mm margin of healthy tissue. The average local recurrence rate is 15-20%. Radiotherapy and combination chemotherapy including vinblastine, ifosfamide, etoposide, cyclophosphamide, doxorubicin and dactinomycin has been advocated for inoperable recurrence or margin-positive resection.[5]

A high index of suspicion is required to diagnose this tumor and close follow-up is necessary to detect recurrence. Permanent reconstruction can be done after growth is completed.

REFERENCES

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Figure 1: Clinical photograph

Figure 2: CT scan image

Figure 3: Histopathology showing predominant round cells and occasional pigment containing epithelium (H/E, x20)