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Original Article

MelanoticNeuroectodermalTumor of Infancy: A Case Report

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ABSTRACT

Melanotic Neuroectodermal Tumor of Infancy (Melanotic Progonoma) is relatively a rare tumor of neural crest origin with maxilla as the commonest site. Histopathologically confirmed case of Melanotic Neuroectodermal Tumor of Infancy in a nine months old female is reported herewith.

Introduction

MelanoticNeuroectodermalTumor of Infancy (MNTI) is an extremely uncommon osteolytic pigmented neoplasm of neural crest origin, primarily affecting the jaw of newborns and infants. (1, 2) A diligent search was done only to find 365 cases of this tumor in the world literature till October 2008. (3)

Majority of these tumors are slow growing and asymptomatic. They are locally aggressive, intraosseous and osteolytic. The rate of recurrence is 15% and the rate of malignant transformation is 6.6%.4 Histopathology of the tumor is characteristic and therefore conclusive.

Clinical Summary:
(History narrated by mother)

Present case was nine months female child, brought with swelling of left upper alveolus of four months duration, extending into upper lip and causing difficulty in chewing, swallowing as well as sucking. Baby underwent age matched vaccination schedule.

Except pallor, general examination was normal.

On local examination, swelling was firm to hard about 8 x 6 cm in dimensions, appeared to arise from left upper alveolus with extension in upper lip and base of nose. It caused nasal obstruction and difficulty in opening the jaw. The swelling had distinct black discoloration. Systemic examination was within normal limits.

CT scan showed a heterogeneously enhancing mass into left maxilla. Necessary hematological investigations were carried out which were within reference range. With these findings and investigations provisional diagnosis of alveolar cyst or benign tumor was entertained and patient was posted for surgery.

Pathological findings:

Tumor was almost inoperable due to its adherence to underlying bone and hence removed in pieces. It appeared to occupy left upper alveolar margin, upper lip and anterior half of ipsilateral hard palate. However septal cartilages appeared to be spared.

Grossly, it was irregular lobulated multiple pieces measuring 4 x 3 x 2 cm with variable consistency and distinct black discoloration.

Microscopically, multiple sections studied showed a tumor lined by stratified squamous epithelium composed of two types of cells. (Figure1). One type of cells were large, polygonal with hyperchromatic nucleus and abundant intracytoplasmic melanin pigment, arranged in pseudoglandular and alveolar pattern. Centre of alveoli showed nests of small round cells with round nuclei having stippled chromatin(Figure 2). Surrounding bone showed focal infiltration by tumor cells.(Figure 3)

Figure1: Glandular and alveolar arrangement with peripheral polygonal cells containing abundant melanin (10 x 10, H & E)

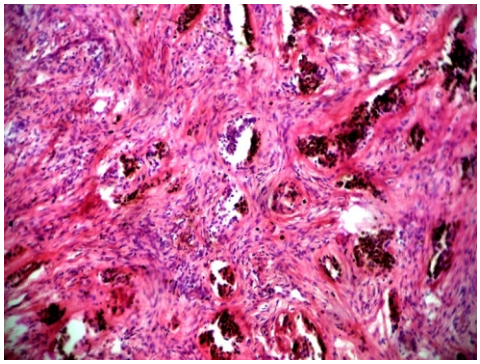

Figure 2: Alveolar and glandular arrangement showing two types of cells. (40 x 10, H & E)

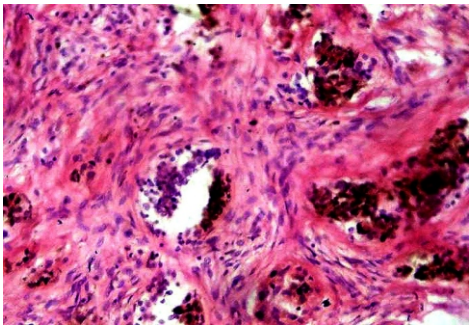

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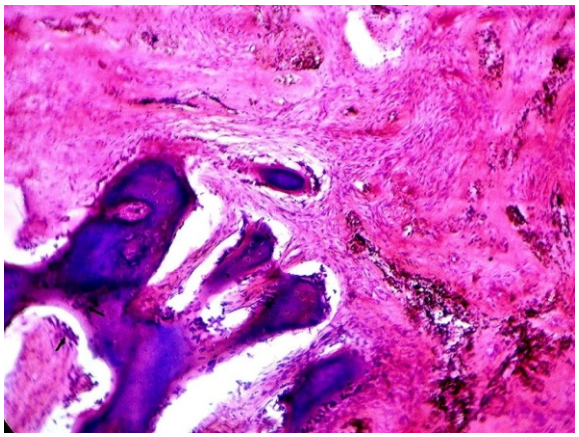
Figure 3: Bony trabeculae with invasion by tumor cells. (40 x 10, H & E)



Discussion

MelanoticNeuroectodermalTumor of Infancy (MNTI) is a rare neoplasm of neuroectodermal derivation, previously thought to arise from odontogenic epithelium. (5) Only 365 cases have been reported in the world literature till October 2008.(3) An exact number is difficult to discern because of the terms that have been applied to the lesion in the past like pigmented ameloblastoma, pigmented epulis, retinal choristoma, melanoameloblastoma. More than 90% patients present in first year of life (2,5,6). Usually the median age is 4 months (2). There is no sexual predilection. (2, 5, 7) Commonest location of the tumor is in the head and neck region, with most arising in anterior part of the maxillary ridge. (1, 2, 5, 7, 8) Other common sites include skull, mandible and brain. (2, 5, 7, 8) Present case was nine months old female with a lesion on anterior part of left maxillary alveolus extending into the surrounding tissue.

This tumor usually begins as solitary cystic or solid mass often pigmented blue or black due to presence of melanin. (1, 2, 5)

The studied case had lobulated solid, blackish mass in the maxilla.

The behavior of this tumor is generally benign but it is locally aggressive. (2, 5, 7, 8) The tumor with intraosseous location expands rapidly, destructs bone and displaces teeth. (2, 7)

This patient had characteristic history of rapid growth, with extension in the surrounding organ in four months duration.

Histology of this tumor is unique and characteristic. Tumor has typical biphasic pattern with two types of cells. Tumor showed polygonal cells arranged in sheets and, alveolar or glandular structure containing abundant melanin pigment. The central portion of alveoli contained characteristic small, round neuroblast like cells with hyperchromatic nuclei. (2, 5, 7)

This studied case had typical histomorphology as described in literature. Tumor showed infiltration of bone in this case.

This patient had all characteristic features of tumor including rapid expansible growth in anterior part of maxilla. All characteristic histomorphological features of biphasic picture, abundant melanin pigment and bony infiltration and hence diagnosis of Melanotic Neuroectodermal Tumor of Infancy (MNTI) was made.

In 1966, high urinary Vanillyl-Mandelic Acid (VMA) was found to be excreted in the urine of patients with this tumor(2, 7, 9) This suggested the neural crest origin of this tumor. It was later confirmed by immunohistochemistry and electron microscopy. However either the presence or absence of VMA in the urine neither confirms nor rules out the diagnosis of tumor. (2, 5) A significant number of cases of MNTI associated with normal levels of VMA appear in the literature.10Because the diagnosis of MNTI was not entertained clinically, VMA estimation was not done.

Immunohistochemistry is of assistance in cases that are more difficult to diagnose. The cuboidal cells express cytokeratin and HMB-45, and negative for S-100. In this case histomorphology was very characteristic. The tumor was in conformity with the typical diagnostic features and hence it did not merit the necessity for immunohistochemistry. Immunohistochemistry helps in confirming the diagnosis in doubtful cases lacking the typical histological features.

The incidence of malignancy is very rare and accounts for 6.6% of all cases.[4] Diagnosis of malignancy is based on increased growth rate, infiltration and metastases. The treatment of choice is complete surgical excision. The local recurrence rate is 15%.[4]

Conclusion

A high index of suspicion is required to diagnose this tumor and close follow up is necessary to detect recurrence. This case is reported for its extreme rarity and the classical clinical and pathological findings. The sole aim to report this case is to make clinician aware of this entity when dealing with maxillary mass in an infant. Early clinical diagnosis and treatment at the appropriate time can prevent complications and metastasis.

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