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Case Report
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Melanotic Neuroectodermal Tumor of Infancy (MNETI) of Maxilla – A Diagnostic Dilemma

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Abstract

Melanotic neuroectodermal tumor of infancy is a rare neoplasm that affects children under the age of one year. The tumor generally occurs in the head and neck area, with the maxillary alveolar crest being the most frequent site of involvement. MNETI manifests as a pigmented expansile mass with an aggressive growth pattern and a high recurrence rate. The rarity of this condition contributes to the absence of robust evidence-based therapeutic techniques. We describe MNETI in a 2.5-month-old baby who had a maxillary lesion that was treated with debulking.

Keywords: Melanotic neuroectodermal; Tumor of infancy Maxilla; Benign neoplasm

Introduction

Melanotic Neuroectodermal Tumor of Infancy (MNETI) is an uncommon pigmented tumor originating from neural crest cells frequently seen in the head and neck region in the first year of life. Most cases are observed in the craniofacial region, with the alveolar crest of the maxilla being the most involved site [1]. The behavior of MNETIs is benign but locally aggressive with a recurrence of 10% to 15% and eventually malignant in 6.97%. Although it is a benign tumor, it is important to recognize because of its rapid growth and propensity for local recurrence, as well as its small round blue cell shape in histology which can lead to a misinterpretation of malignant neoplasm [2].

A firm, painless, rapidly developing mass emerging from the anterior maxillary alveolus, which elevates the upper lip and can impair with feeding is the typical clinical appearance. Surgery is the treatment of choice with a local recurrence rate of 10% to 15% [3,4].

Local surgical excision or debulking is considered the first-line treatment for MNETI. This is particularly challenging in growing children. We report on the treatment outcome of a maxillary MNETI in a 2.5-month-old infant treated by debulking *via* intraoral approach.

Case Presentation

A 2.5-month-old female child had reported to the outpatient department of our institute with complaint of a swelling on the right side of the anterior alveolus in the pre-maxillary region progressively increasing after birth. The swelling was painless slow growing, initially small in size gradually increasing in size causing obliteration of right nasolabial fold, facial asymmetry and feeding difficulties (Figure 1). History of previous trauma, pus discharge, foul smell or bleeding was negative.

On clinical examination, a single diffuse swelling, slightly bluish in color, was found primarily on the right side of the anterior alveolus, measuring 2.5 cm × 2 cm spanning beyond midline. The swelling was non-tender with no surface erythema or ulceration.

Radiographically erupting tooth buds of the maxillary anterior teeth were appreciated. It appeared as a radiolucent, hypodense area with displaced tooth buds in relation to the right central incisor suggestive of eruption cyst. An ultrasonography report revealed a well-defined cystic lesion in the upper jaw, measuring around 2 cm × 2.5 cm on the right side of the maxilla, with calcifying foci inside and no signs of internal vascularity, indicating an eruption cyst.

A Computed tomography (Figure 2) scan showed 16.4 mm × 16.9 mm × 9.7 mm expansile lytic lesion at upper alveolus of right paramedian location with an unerupted tooth of deciduous left central incisor at the periphery of the lesion.

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Figure 1: Clinical photograph showing the lesion.

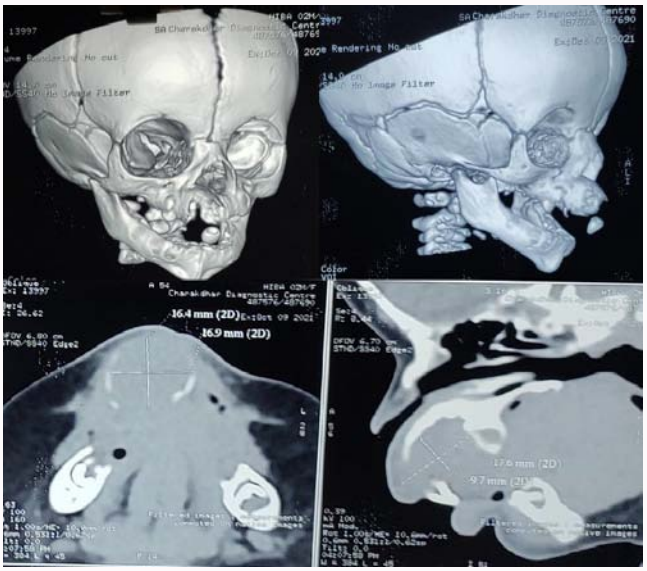

Figure 2: Computed tomographic scan of showing the extent of lesion.

Investigations included complete blood count, coagulation profile, liver and renal function tests, viral markers and a chest X-ray. Serum catecholamines, ultrasound of the abdomen and whole-body skeletal screening was within normal limits. Surgery performed with endotracheal anesthesia through intraoral approach. Aspiration with 16-gauge needle was negative lead to surgical exploration. Alveolar crestal incision was given, mucoperiosteal flap reflected and a well encapsulated dark solid tumor was identified. The tumor was excised maximally along with the sacrifice of associated deciduous teeth buds (right central incisor, lateral incisor, canine, and left central incisor). The tumor that was removed was firm, round, and grayish to black in color (Figure 3). Cavity was thoroughly curetted and hemostasis achieved. The cavity was then curetted to remove all traces of blue-black tissue, after which primary closure could be achieved. Mucoperiosteal flap was repositioned and closed with simple interrupted sutures by a resorbable suture.

Postoperatively the patient was allowed oral intake after recovery from anesthesia. The patient was discharged at 5 days after surgery and she had no feeding difficulty. The patient is in regular follow-up to detect any recurrences.

Microscopic examination with hematoxylin and eosin-stained sections revealed a non-encapsulated mass constituted of a dual

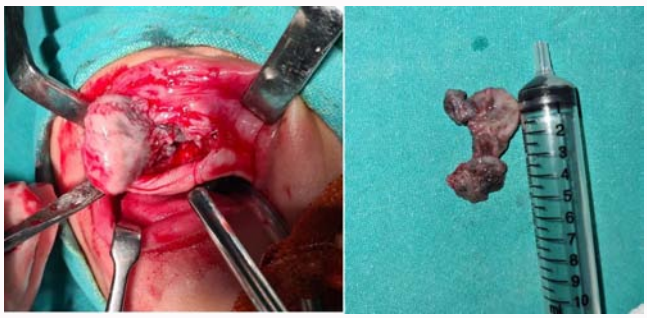

Figure 3: Excised lesion intraoperatively & the specimen.

composition of small round blue cells and larger melanin-containing epithelial cells, in a dense fibrocellular stroma. It was made up of tumor cell nests and clusters grouped in an alveolar pattern and separated by fibrovascular stroma. The central part of the alveoli is made up of small round cells with little cytoplasm and dark nuclei whereas peripheral part comprised of cuboidal, flattened epithelioid cells containing melanin pigments in cytoplasm. Bony trabeculae are found to be entrapped by the tumor. No necrosis was seen. Findings were reminiscent of Malignant Small Round cell tumor and further Immunohistochemistry (IHC) was warranted. Pathologist suggested differential diagnosis could include peripheral nerve sheath tumor, Ewing's sarcoma or neuroblastoma.

Immunohistochemistry (IHC) revealed positive staining of Cytokeratin (CK), Homatropine Methylbromide 45 (HMB 45), Cluster of Differentiation 99 (CD99) and synaptophysin markers. HMB 45 (cytoplasmic) and CK are only found in epithelial cells (both membranous and cytoplasmic). Neuroblastic cells are negative for CK (cytokeratin) and HMB 45 staining. Nuclear expression of Ki67 was also observed. Hence, histopathological differential diagnosis given was MNETI or malignant melanoma. Finally, IHC positivity for HMB 45, synaptophysin in the present case indicated that the tumor cells are melanocytic and confirmed the final diagnosis as MNETI.

Discussion

Krompecher was the first to describe melanotic neuroectodermal tumor of infancy in 1918 [6]. MNETI is an uncommon but distinct tumor that affects children under the age of one year. The etiology of MNETI is unknown; it may develop from enclosed epithelial remnants, odontogenic epithelium or a phylogenetic origin which is linked to the median or pineal eye of various lower vertebrates [6,7]. The tumor is generally accepted as being of neuroectodermal origin on the basis of ultrastructural, immunocytochemical and electron microscopic studies [7,8].

Overall 90% percent of the tumors were seen in the head and neck region, maxilla being the most common site (68.8%), followed by skull (10.8%), mandible (5.8%), and brain (4.3%) [8,9]. Most of the patients have involvement of the maxillary alveolus without gender predilection [6,9].

The tumor is unique in its ability to synthesize melanin and may also elaborate Vanillylmandelic Acid (VMA) in urine [5]. Chaudhary et al. [9] observed exclusive production of VMA by male patients having MNETI. The expression of VMA is characteristic of tumors arising from neural crest cells however its association with male patients could be coincidental [9].

Fine needle aspiration cytology could be inconclusive and

preoperative diagnosis usually relies on the clinic features of this rare entity. The presence of a central region of radiolucency with strong borders displacing the surrounding bone and tooth buds may be typical radiographic features. This would probably suggest a diagnosis of eruption/dentigerous cyst of newborn, but a negative aspiration would rule out the possibility. Computed tomography scans may prove very helpful in planning surgical excision. The symptoms of the patient could be due to physical effect of the swelling and cosmetic disfigurement.

The histopathological differential diagnosis of MNETI involves other pediatric 'small round cell' neoplasms. These include neuroblastoma, Ewing's sarcoma, and rhabdomyosarcoma, and peripheral neuroepithelioma, desmoplastic small round cell tumor, malignant melanoma, peripheral primitive neuroectodermal tumor and lymphoma [10].

There are reports of wide excision, subtotal maxillectomy, and use of titanium miniplates for reconstruction [11,12] in the literature but these were not required in our patient. A thorough local excision and curettage of the cavity to remove all traces of bluish black tissue could suffice for cure. However, most authors favor a conservative approach of local excision and curettage, in as much as remnants of tumor may not necessarily cause recurrence. The debulking action may trigger tumor regression by body defenses, resulting in the involution of remaining tumor [13].

Although the majority of tumors are benign, a local recurrence rate of 10% to 15% and a malignancy rate of 3.2% are reported [10]. Histopathology and IHC are confirmatory diagnostic of MNETI. Our patient is free of any local recurrence or distant metastasis. Based on our experience we feel that the diagnosis of MNETI is mainly clinical. Physical findings are typical, whereas radiographic characteristics are distinctive. IHC clarifies the neural crest as the cell of origin of MNETI and characterizes the tumor but no prognostic benefit is available from this investigation at present. In neuroblastoma the expression of neuron-specific enolase is a poor prognostic feature but this was not seen to apply to MNETI. For individuals with MNETI, early conservative surgical excision could yield satisfactory results with a good prognosis [10].

The quick and invasive expansion of MNETI emphasizes the need for early diagnosis to save inordinate amount of time spent in treatment [14]. Late diagnosis may be the reason for difficulty in treatment and subsequent facial deformity.

Following surgery, a close follow-up especially within the first 6 months is very important. Early tumor detection and treatment can help the patient to minimize subsequent complications and a better prognosis.

Conclusion

Melanotic Neuroectodermal Tumors of Infancy (MNETI) are very rare but have deleterious effects on the condition of the child ranging from having airway obstruction to malignant transformation. In our case, the clinical, radiographic, ultrasonographic, and histopathological findings assisted us in making the diagnosis, as well as the proper treatment plan and postoperative follow-up, which helped us confirm the diagnosis cumulatively without the use of adjuvant chemotherapy or radiotherapy. In the absence of metastatic disease MNETI can be successfully managed by local excision or debulking.

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