CASE REPORT

A Rare Melanotic Neuroectodermal Tumor of Infancy in a 3-month-old Child: A Case Report

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Abstract

A rare case of melanotic neuroectodermal tumor of infancy (MNTI) occurring in a 3-month-old male child affecting the left maxilla is described. Computed tomography showed the benign expansile type of lesion affecting the left maxilla. The lesion was surgically excised completely. Histopathology confirmed the diagnosis of melanotic neuroectodermal tumor. Almost 6 months of follow-up showed no recurrence.

Keywords: Benign tumor, Case report, Infancy tumor, Neuroectodermal tumor.

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Introduction

A very rare benign malignancy called melanotic neuroectodermal tumor of infancy (MNTI) mostly affects the anterior maxilla in newborns under one year. This usually manifests as a pigmented swelling on the front maxilla that is nonulcerative, rapidly progressing, and painless.[1–4] Melanotic progonoma, pigmented epulis of infancy, pigmented ameloblastoma, and melanotic epithelial odontoma are other names for MNTI.[1] In addition to the maxillary region (68–80%), MNTI has also been reported to include the brain (4.3%), mandible (5.8%), and skull (10.8%).[5] According to published research, the patients' mean age is 4.3 months, and their gender predilection ratio is almost equal 6:7.[6–8] Here, we report a case of MNTI in a 3-month-old boy who was treated with a broad surgical excision, as well as a review of current histopathologic understanding and therapeutic approaches.

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Case Description

A 3-month-old child presented himself to the Department of Oral and Maxillofacial Surgery of Bharati Dental College and Hospital, Bharati Vidyapeeth (Deemed to be University) in Pune. The main complaint was swelling over the left maxillary region. After 1 month of the birth, the parents started noticing swelling, which at first grew slowly before starting to expand quickly after 1 and 1/2 months. The child's development was deemed to be typical for his age and neither medical nor family history was significant. Upon clinical examination, a solitary, pigmented, nonpedunculated swelling measuring approximately 4.5 × 3 cm was observed over the left maxillary anterior region without any ulceration or discharge (Fig. 1). This swelling has caused the left side of the nose to flare and has destroyed the labial vestibule, raising the upper lip. The skin surrounding the nose on the affected side turned pinkish red in the middle and adjacent to the ala. The swelling was nontender, nonfluctuant, compressible, and had a consistency ranging from soft to firm. A solitary expansile solid lesion with an epicenter located in the maxillary anterior alveolar ridge was visible in the left anterior maxilla on computed tomography images (Fig. 2). The tumor mass was sent for histological analysis after being surgically removed under general anesthesia (GA) (Fig. 3). Histopathological analysis revealed that the characteristics, which show a biphasic population of cells, were consistent with a small round cell tumor favoring MNTI (Figs 4 and 5). Confirmation was achieved through

Fig. 1: Clinical picture showing extraoral swelling

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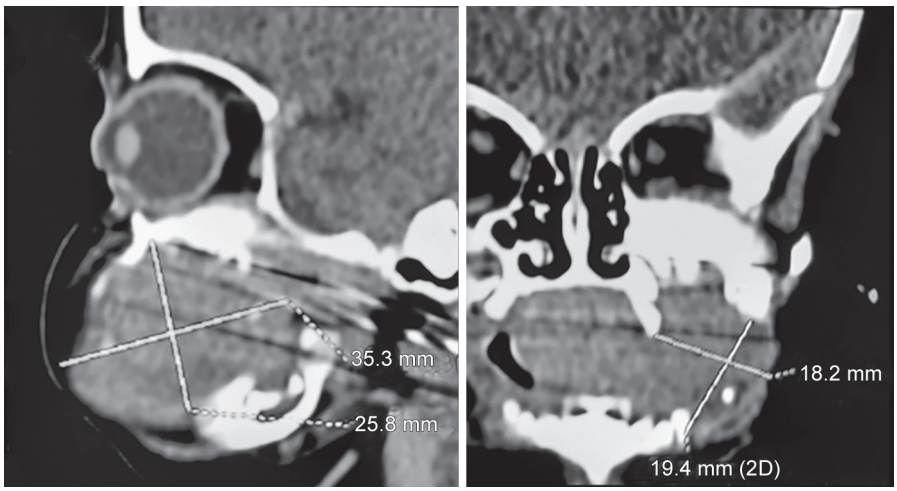

Fig. 2: Computed tomography showing expansile lesion

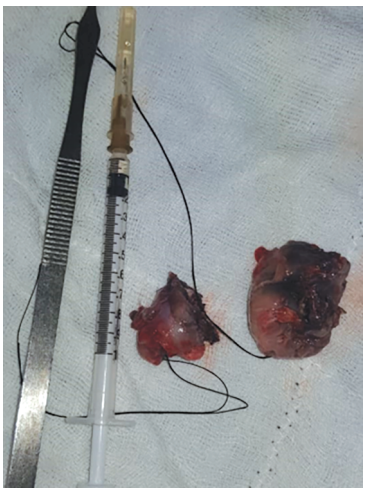

Fig. 3: Surgically excised tumor mass

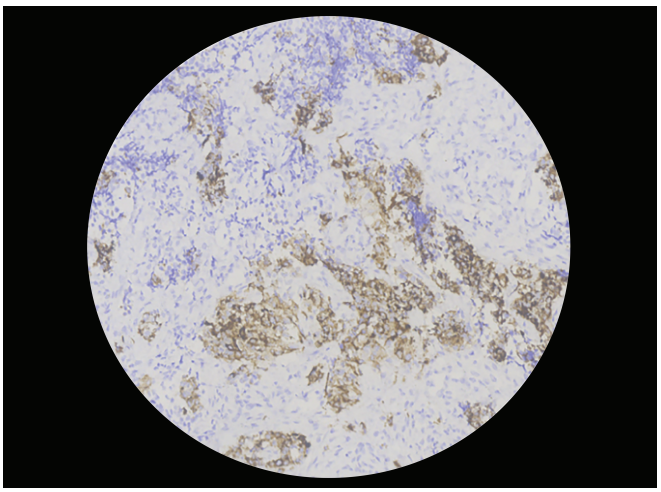

Fig. 5: Epithelioid cells are positive for HMB-45

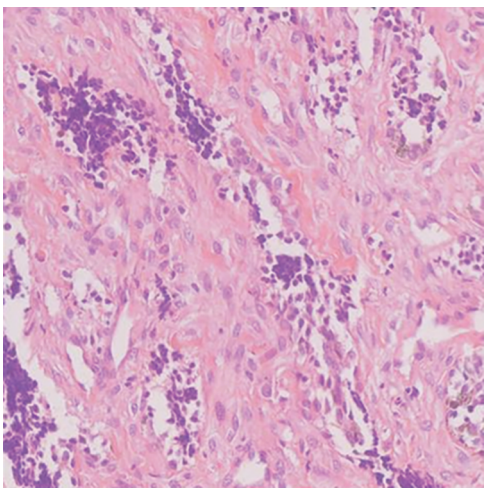

Fig. 4: Small round tumor cells arranged in diffuse and alveolar patterns separated by thin fibrovascular stroma (hematoxylin and eosin, ×10)

immunohistochemistry (IHC) investigations. Thus, based on clinical, surgical, histological, and immunohistochemical results, a final diagnosis of MNTI was obtained.

Discussion

The first documented case of MNTI was described as "congenital melanocarcinoma" by Krompecher in 1918.[9] He reported a 2-month-old baby's pigmented lesion that was associated with a developing tooth. This tumor's source has many names, including pigmented ameloblastoma, melanotic adamantinoma, melanotic epithelial odontoma, retinal anlage tumor, retinal choristoma, melanotic progonoma, atypical ameloblastoma, pigmented teratoma, retinoblastic teratoma, and pigmented epulis, have been associated with this condition, which is believed to originate from odontogenic epithelium or retinal rests. However, investigations using electron microscopy, IHC, and ultrastructural methods, as well as sporadically elevated vanillylmandelic acid (VMA) levels in the urine,[10] It is now established that neural crest-derived cells are the source of this tumor. In 1966, Borello and Gorlin[10] proposed the term "MNTI," which is currently recognized globally. In their case series, Johnson et al.[11] said that a tumor of neural crest origin can be diagnosed with great significance when there is a large urine output of VMA. This tumor presents clinically as a nonpedunculated solid swelling involving the premaxilla. Melanin pigment, which gives some areas of the enlargement a bluish hue, may be the cause of the surface discoloration. Asymmetry

of the face, vestibular obliteration, tooth displacement, and feeding difficulties are obvious but noteworthy observation times. Although it grows quickly, pseudo-encapsulated bone production is thought to be benign and may occur as a result of reactive bone formation. The nasopalatine cyst and globulomaxillary cyst are two examples of developing cysts that are included in the differential diagnosis of MNTI. Odontomas, ameloblastomas, ameloblastic fibromas, odontogenic myxomas, adenoameloblastomas, and odontogenic keratocysts are examples of odontogenic lesions. Arteriovenous malformation, fibrous dysplasia, and central giant cell granuloma are examples of nonodontogenic nonneoplastic lesions. Nonodontogenic neoplastic lesions—Burkitt's lymphoma, Ewing sarcoma, Langerhans cell histiocytosis, and rhabdomyosarcoma. Nevertheless, a small number of options remain after a review of the clinical and radiological results. With or without well-defined margins, radiolucent lesions can be seen on conventional radiographs. Although hypodense masses have also been described, hyperdense masses are typically seen by computed tomography scans. On computed tomography, the lesion's margin and extent are visible, assisting the surgeon in performing their work with accuracy. More accurately, IHC stains aid in the diagnosis of MNTI. Commonly, epithelioid cells stain positively for vimentin, cytokeratin, epithelial membrane antigen, and Human Melanoma Black-45 (HMB-45), which indicates melanotic and epithelial differentiation. When it comes to protein S-100, which aids in the differentiation of tumors like melanoma, these cells are frequently nonreactive. Neurogenic markers such as neuron-specific enolase, glial fibrillar acidic protein, and synaptophysin are positive in smaller cell nests. Staining variability demonstrates the tumor's multiphenotypical nature.[1] Studies using electron microscopy reveal that neural crest cells are the source of a subset of neuroectodermal tumors. Similar to previous tumors, elevated α flavoprotein levels, and positive c-myc staining were also linked to this one tumor neuroectodermal.[1] Despite the absence of comparable genetic alterations that can connect other neuroectodermal tumors to MNTI. Current research indicates a 10–60% recurrence probability after conservative surgical excision. Around 6.5% of cases exhibit distant metastases. Multicentric development, penetration of the tumor edge into the bone, and inadequate encapsulation are possible causes of recurrence.[2] Surgical excision combined with a 5 mm removal of safety margins of the tumor. The ideal course of action to stop a tumor from returning is to treat the healthy tissue surrounding the tumor. The reported average recurrence rate for MNTI is about 20%.[5] When MNTI approximates vital structures chemotherapy or radiotherapy can be the treatment of choice. Waiting until the patient reaches skeletal maturity allows for the reconstruction of missing parts, such as missing teeth or portions of the alveolus. For the time being, a removable prosthesis might fulfill a functional need for patients.

Conclusion

Even though MNTI is a benign lesion, early detection and diagnosis is the key factor as it can cause local destruction. As the occurrence of this tumor is in growing age careful follow-up becomes necessary to check for recurrence and also to see any effect of surgical trauma on growth and development of maxilla-facial structures.

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