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Case Report

Melanotic Neuroectodermal Tumor of Infancy: A Rare Case Report and Review

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Abstract

The melanotic neuroectodermal tumor of infancy is a benign but locally aggressive vanillylmandelic acid (VMA) producing neoplasm that originates from neural crest cells. It predominantly affects infants and is most common in the maxilla. Due the anatomical complexity of the head and neck region and the pathological nature of this tumor entity, the surgical therapeutic management poses considerable challenges upon surgeons and patients.

The purpose of this article is to report a rare case of a rapidly growing mass, diagnosed as melanotic neuroectodermal tumor of infancy (MNTI), in the right maxilla in a 3-month-old boy which was treated by a radical local resection and followed up after one year without any clinical and radiographical signs of recurrence.

Keywords: Benign Tumor of Infancy; Neuroectodermal Tumor; Neural Crest Cell; Tumor; Case Report

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Introduction

Although first described in 1918 by Krompecher as a "congenital melanocarcinoma", the tumor has been known by many other names in literature including melanotic epithelial odontoma, pigmented teratoma, retinal anlage tumors and melanocytoma [2]. Borello and Gorlin in 1963 finally suggested the term "Melanotic neuroectodermal tumor of infancy (MNTI)" which has now been globally accepted, for benign tumors predominately affecting the head and neck region [1] and arising from neural crest cells.

Clinically, the typical lesion appears as rapidly growing firm soft tissue mass in infants and newborns and is mostly observed in the maxilla. The aggressive behavior of the tumors causes a considerable bone destruction which often complicates the surgical intervention [3]. Considering the high recurrence rate and the aggressive nature of this tumor, the treatment modalities include wide local excision with or without adjuvant and neoadjuvant therapy options.

Case Report

A 3-month-old male child was presented to our clinic (Assendan Medical Center, Tripoli, Lybia) by his mother who noticed a rapidly growing mass for approximately 3 weeks in right maxillary region. The clinical examination showed a firm in consistency, non-fluctuant and non-compressible smooth swelling with a central ulceration in the overlying surface mucosa. (Figure 1). Conventional computed tomography with contrast media revealed a 3 x 2 cm measuring well-defined locally destructive hyperdense radiolucency with displacement of a dental follicle expanding into the right maxilla (Figure 2). Based on the clinical, radiographic appearance and considering the age of the patient, a rhabdomyosarcoma, Ewing sarcoma and a congenital teratoma were included in the initial differential diagnosis.

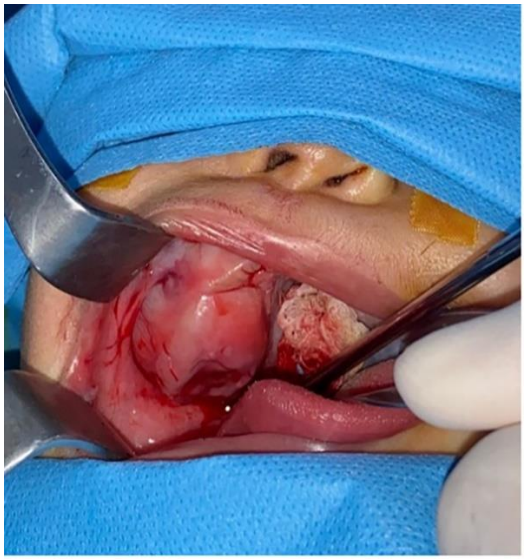

Figure 1: Preoperative intraoral view exposing a 2 x 3cm well demarcated swelling in the right maxilla with a central ulceration of the overlying mucosal layer.

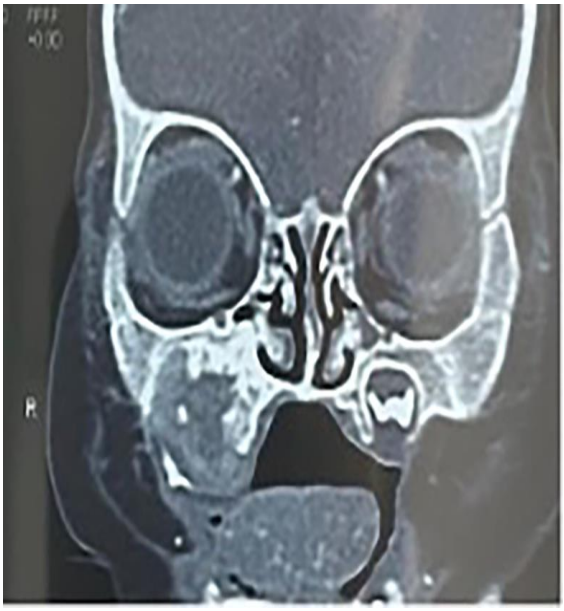

Figure 2: Pre-operative CT scan displaying a well-defined destructive hyperdense radiolucency with displacement of dental follicles in the right maxilla.

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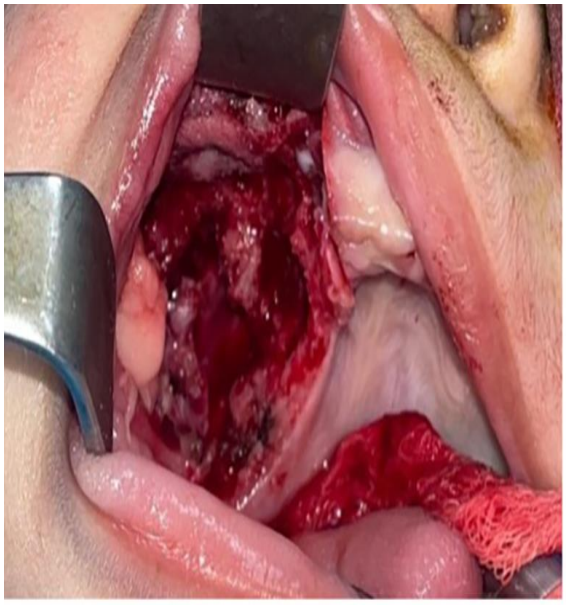

Figure 3: Intraoperative view laying out the surgical site after wide excision of the lesion.

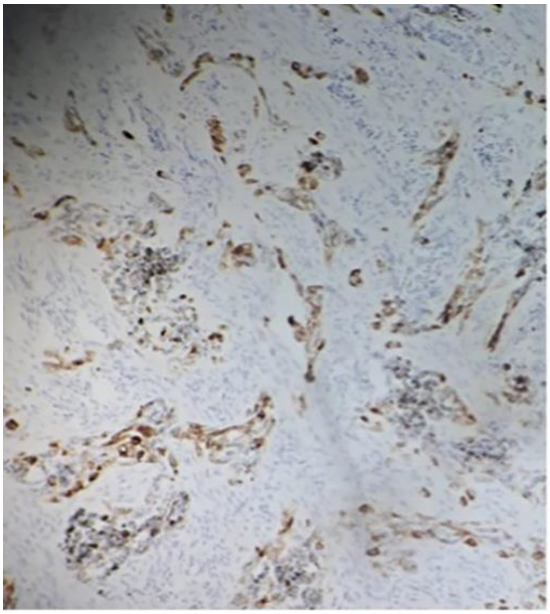

Figure 5a: Histopathology: Melanotic neuroectodermal tumor of infancy (Cytokeratin; AE1/AE3, 200x): The epithelioid cells population are positive.

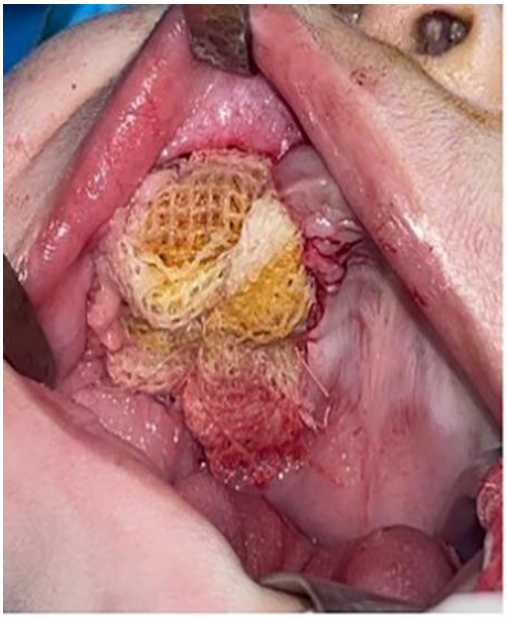

Figure 4: Intraoperative situs documenting the surgical defect coverage with gauze soaked in povidone iodine.

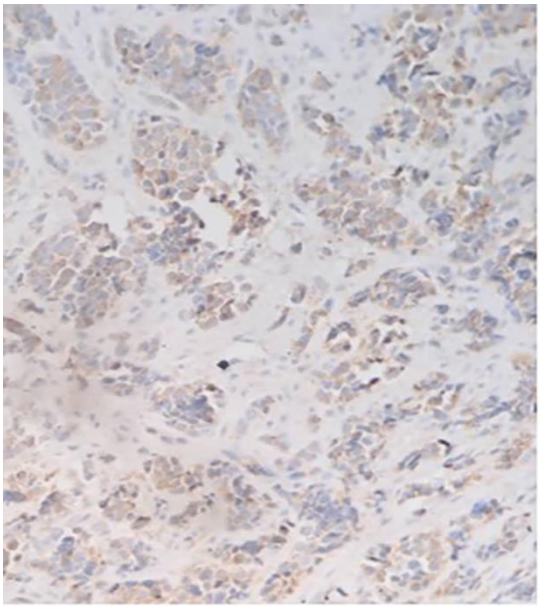

Figure 5b: Histopathology: Melanotic neuroectodermal tumor of infancy (Synaptophysin, 400x): The small round blue cells are positive.

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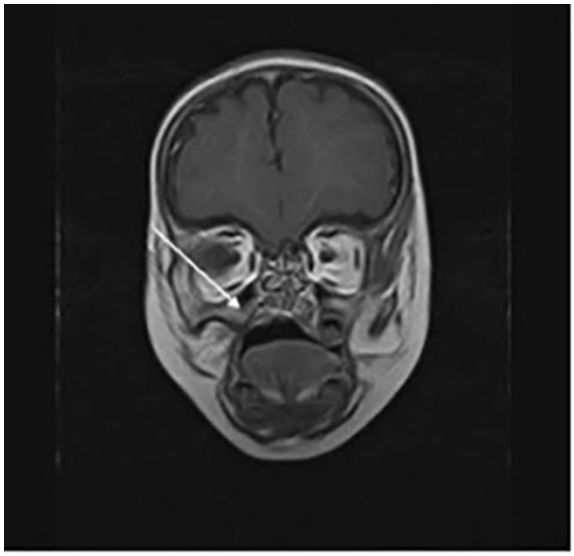

Figure 6: One-year postoperative follow-up head and neck MRI evidencing complete resolution of the tumor.

An incisional biopsy was performed under general anesthesia. The histopathology report revealed a pigmented melanotic neuroectodermal tumors of infancy. Due to the rapid growth and the destructive pattern of the tumor, the infant was scheduled for a surgical intervention. A wide local excision was made with a 5 mm circumferential safety margin and send for histopathological examination (Figure 3). Due to the bony infiltration, a peripheral ostectomy was done with a surgical diamond bur under rigorous irrigation. The surgical site was covered with gauze soaked in povidone iodine (Figure 4). The patient was extubated uneventfully and discharged from hospital the next day. The patient was followed up five days postoperatively for removal of the dressing. The wound healing by secondary intention was already in progress. The final histopathology report revealed a biphasic population of tumor cells composed of small round bule cells with hyperchromatic nuclei and scanty cytoplasm (called neuroblastoma-like cells) which showed a cross reactivity to cytokeratin. A second population of epithelioid cells with round vesicular nuclei and abundant cytoplasm containing melanin pigment were tested positive to Synaptophysin (Figure 5 A, B).

A postoperative follow-up head and neck MRI after the first postoperative year did not reveal any signs of a residual or recurrent tumor (Figure 6).

Discussion

MNTI is a rare neural crest derived benign tumor with an aggressive biological behavior and is characterized by a high recurrence (15-20 %), a malignancy transformation rate of 6%, and a metastasis rate of 3% [4-9].

The tumor mainly occurs in male newborns and infants. The anterior maxilla is the most affected tumor location (69%), whereas mandibular lesions represent only 6% [5 ,6].

This pathological tumor entity often initially presents as a painless rapidly growing firm soft tissue mass with a smooth bluish colored overlying mucosa. Considering the patient's age and the clinical appearance of the tumor, the differential diagnosis of such lesions may include congenital vascular lesions, primary bone malignancies like Ewing sarcoma and connective tissue malignancies such as rhabdomyosarcoma. Histologically, the tumor consists of two cell types, neuroblast like cells and epithelioid cells and melanin pigmented cytoplasm [7,8,10].

Many studies have shown that there is a strong relation between recurrence rate and age of the patient, the size of the tumor and the initial therapeutical management. Higher recurrence rates are seen more often in younger patients with bigger tumor sizes and in cases treated by a more conservative management.

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The anatomical complexity of the orofacial region and localized osseous infiltration are the main causes for incomplete resection of the tumor which may explain local recurrence and /or distant metastasis. Therefore, the therapeutic management of melanotic neuroectodermal tumor of infancy is mainly wide surgical resection with a 5mm safety margin.

Since aggressive surgical treatment and radiotherapy are usually associated with unpleasant cosmetic and functional outcomes in growing patients, some clinicians proposed systemic treatments options such as chemotherapy. However, systemic chemotherapy is associated with a high recurrence rate when used solely and a significant risk of long-term complications including subsequent malignancies and cardiovascular disease. Therefore most authors recommend that chemotherapy should be reserved for inoperable cases, early recurrence and distance metastases [4 ,5 ,13]. To date, two different chemotherapy protocols are described in the literature: Cyclophosphamide and vincristine for recurrent tumors and the OPEC/OJEC - protocol consisting of vincristine (O), cisplatin (P), etoposide (E), cyclophosphamide (C), carboplatin (J) which is reserved for persistently progressing tumors only [11,12].

Conclusion

MTNI is a very rare tumor that affects the craniofacial region mainly in the maxilla at an early age. Due to its rarity and complicated biological behavior, this category of tumors is posing multiple therapeutic challenges to surgeons and patients. Wide surgical resection with a safety margin is the most recommended treatment modality and the mainstay of treatment in these masses to avoid recurrence and distant metastasis. Even though there is no clear evidence of the role of adjuvant chemotherapy, it should only be proposed for aggressive malignant lesions, recurrent and distant metastatic cases. Owing to its risk of recurrence, proper treatment and close clinicoradiological follow-up are of paramount importance. More research on the histological and molecular pattern of MNTI is needed to better adjust the treatment to the biology of this tumor entity and develop more differentiated therapeutical approaches.

Conflict of Interest Statement

The authors have no conflict of interest to declare. All co-authors have agreed with the content of the manuscript and there is no financial interest to report.

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