

# Melanotic Neuroectodermal Tumor of Infancy —A Rare Case of an Encapsulated Tumor

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## Abstract

Melanotic neuroectodermal tumor of infancy (MNTI) is a rare benign, locally invasive neoplasm afflicting the infant more often in the craniofacial region. The current understanding is that this tumor's origin is neural crest cells. The typical presentation is that a rapidly growing non-ulcerated anterior maxillary mass occurs in an infant usually less than six months old. This tumor may involve other areas including the ovaries, epididymis, femur, mandible, and brain. We present that an 8-month-old infant with a maxillary lesion of MNTI appeared encapsulated, which is a hitherto unreported feature. Investigations leading to the diagnosis and the management of the case are also presented. The need to report cases of this rare entity cannot be overemphasized as this will go a long way in adding new knowledge about its biological nature.

## Keywords

Melanotic, Neuroectodermal, Tumor, Infancy

## 1. Introduction

Melanotic neuroectodermal tumor of infancy (MNTI) is a rare benign, locally invasive neoplasm afflicting the infant more often involving the craniofacial region. Since its first description by Krompecher in 1918 [1], just about 500 cases have been reported in the literature [2].

MNTI has been described under various names over the years, an attest to its confusing biological nature. Melanocarcinoma, retinal anlage tumor, melanotic progonoma, melanotic epithelial odontoma, melanoameloblastoma and melanotic adamantinoma are some of the names that have been used to describe

MNTI in the past. An in-depth understanding of the etiology of MNTI has led to its current name [3] [4] [5].

The typical presentation is that a rapidly growing non-ulcerated anterior maxillary mass occurs in an infant usually less than six months old. The tumor may be pigmented bluish black. The rapidly expanding maxillary lesion displaces teeth and causes bone destruction. Other sites of tumor affliction apart from the head and neck include the epididymis, femur, ovaries, uterus, mandible, and the brain [2] [6] [7].

The differential diagnoses of MNTI include rhabdomyosarcoma, Burkitt's lymphoma, malignant melanoma, and neuroblastoma [8]. The history, clinical features, histological appearance and pattern of positivity for Immunohistochemistry markers aid in ruling out the differential diagnosis. Some of the commonly used immunohistochemical markers include HMB-45, NSE, Synaptophysin and AE1/AE3.

The main modality of management of MNTI is surgery. However, this treatment is plagued with recurrence rates ranging between 15% and 30%. The role of chemotherapy as adjuvant/neo-adjuvant treatment of inoperable and metastatic tumors is a recent and promising development [9].

The aim of this case presentation is to add new knowledge of the existence of an encapsulated variety of MNTI.

## 2. Case Report

An 8-month-old female infant presented to the Gertrude's Children Hospital in Nairobi, Kenya on 15<sup>th</sup> November 2021 with a rapidly growing painless maxillary swelling of two-month duration. The onset was intraoral and around the left-maxillary anterior region. There was no pain, however, the growth suddenly became exponential, increasing to the size seen at the presentation. The swelling had grown to the extent of interfering with feeding. Her medical history was unremarkable. She had two older brothers aged 5 and 3 years, who are alive and well. The family history was devoid of any chronic or familial disease with both parents alive and well.

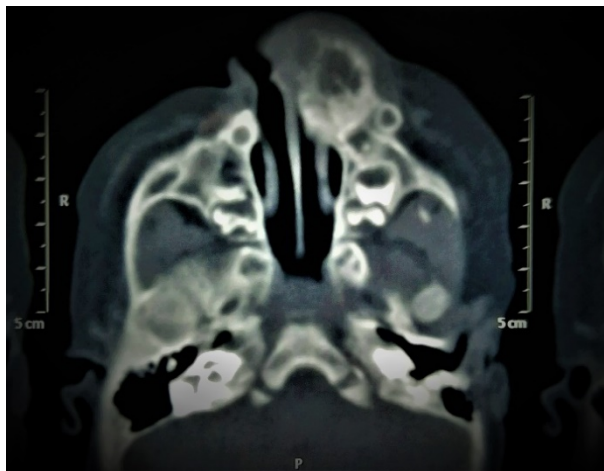
Physical examination revealed a child in distress with a chronologic age commensurate with the physique. There was no pallor, no jaundice and no cyanosis. The cervical nodes were not palpable and the abdomen was soft with no obvious organomegaly.

She had facial asymmetry with a left maxillary swelling extending from the region of the lateral incisor on the right maxilla to the commissure on the left side. The swelling was somewhat rounded and measured 5 × 4 × 4 cm in situ. The overlying skin was of normal colour and texture (**Figure 1**).

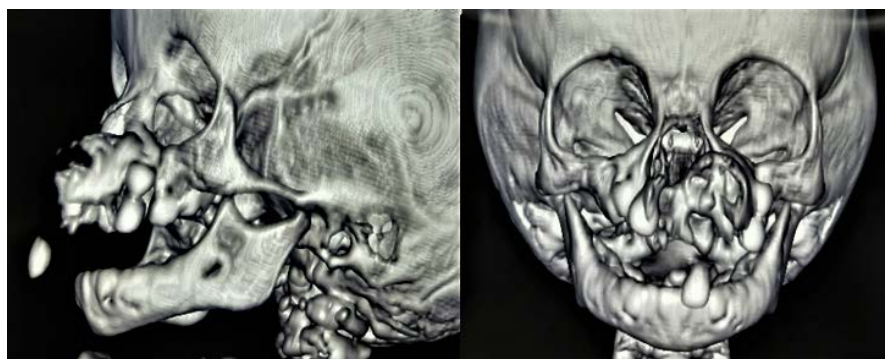
A computerized tomographic scan examination revealed a well-defined mixed radiolucent and radiopaque lesion involving the left anterior maxilla (**Figure 2** and **Figure 3**). There appeared to be septation within the lesion. There were no signs of active invasion of the contiguous structures and the features seen were suggestive of a benign process.



**Figure 1.** Frontal view of the patient showing the extent of the swelling.



**Figure 2.** Axial scan showing a well-defined lesion involving the left anterior maxilla.



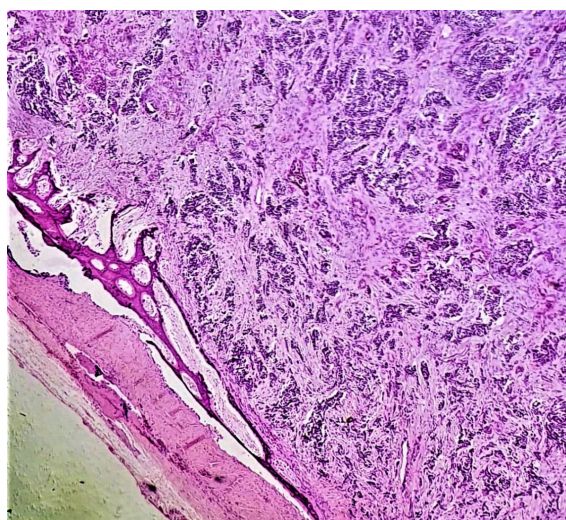
**Figure 3.** 3D reformatted images from the computerized tomographic scan showing the well-defined left anterior maxillary mass with the associated displacement of the 61.

An incisional biopsy was done and the histological features were suggestive of Pigmented Neuroectodermal Tumor of Infancy (**Figure 4**).

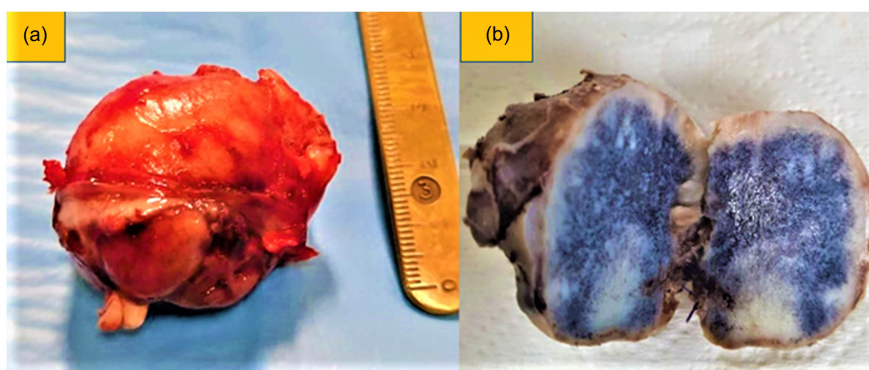
The lesion was then extirpated under general anesthesia via an intraoral approach and a total mucosal closure of the residual defect was achieved by mobilizing the contiguous mucosa. It was noted that the lesion was encapsulated (**Figure 5**). The transected sections of the specimen showed a surface with a bluish black pigmentation pathognomonic of MNTI (**Figure 5**).

The specimen was subjected to immunohistochemical tests that confirmed positivity for the immunohistochemical markers HMB45, synaptophysin and AE1/AE3 thus confirming the diagnosis of Pigmented Neuroectodermal Tumor of Infancy (**Figure 6**).

The patient went through an uneventful recovery period and continues on four months of follow-up post-surgery and was free from the disease (**Figure 7**).

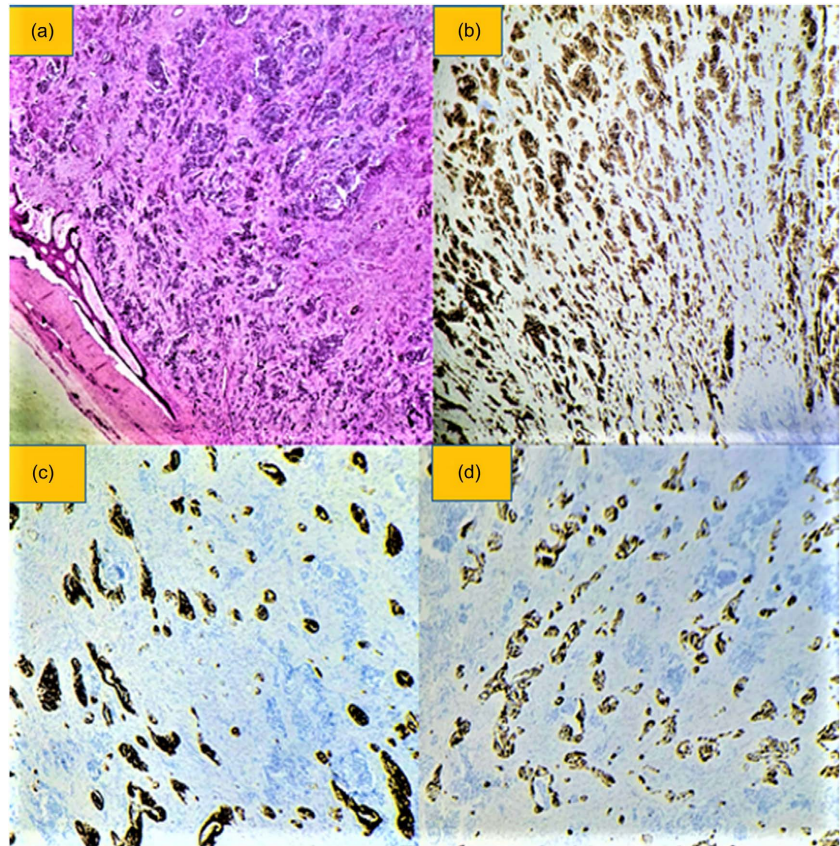


**Figure 4.** The histology under Hematoxylin-Eosin (Magnification  $\times 10$ ) showed a biphasic cellular pattern with nests of the smaller neuroblast-like cells interspersed with the large-melanin containing epithelioid cells in a solid alveolar pattern suggestive of MNTI.



**Figure 5.** (a) The lesion showing its intact capsule after extirpation in its entirety. (b) The picture on the right showing the blue-black pigmentation that is pathognomonic of MNTI. The intact capsule is also evident in the transected specimen.





**Figure 6.** (a) Hematoxylin-Eosin staining (H&E Staining) showed a biphasic cellular arrangement with the larger cells possessing abundant cytoplasm and pale nuclei surrounded are nests of smaller cells resembling neuroblasts. The capsule of the lesion is evident in the lower left corner (Magnification  $\times 10$ ). (b) Synaptophysin staining of the neuroblast-like cells (Magnification  $\times 10$ ). (c) Intense staining of the melanin bearing nests with HMB45. (d) Intense staining of the epithelial pigmented nests with AE1/AE3 (Magnification  $\times 10$ ).



**Figure 7.** Patient showing recovery three-month following surgical treatment.

### 3. Discussion

MNTI is a rare but significant neoplasm affecting the head and neck region with features like the rapid growth and destructive nature closely resembling malignancies in the region. It is therefore imperative that in infants, it is considered as a differential diagnosis to the rapidly growing neoplasia in the maxillary region [9]. The onset of disease in this case was insidious at first followed by rapid growth to the time of diagnosis, a feature that is consistent with some reported cases [6] [8]. In the case reported, the finding of an encapsulated lesion is a new finding hitherto unreported and may point towards the heterogeneous nature of this entity while at the same instance portending a better prognosis. The presence of a capsule may insulate the surrounding tissues from seeding during surgery and prevent a recurrence. The reported recurrence rate of 10% - 15% following surgery in MNTI makes the need for follow-up of treated cases imperative [8].

High levels of vanillyl mandelic acid in the urine of those afflicted by MNTI is indicative of the neural crest origin of the tumor, however, this finding is not usually consistent for all MNTI thereby making it a tool with low diagnostic value [6] [8]. The use of immunohistochemical markers is valuable in reaching a definitive diagnosis. The immunohistochemistry profile for our case was positivity for immunohistochemical markers HMB45, synaptophysin, and AE1/AE3. These mirrored what is already known hence confirming the diagnosis [5]. The age of this infant at the inception of the tumor is 6 months falling squarely at the age-bracket associated with a better prognosis. Removal of the lesion by intra-oral approach guaranteed a good esthetic outcome and reduced morbidity. This particular approach has been the method of choice in some case reports [6]. We managed to attain primary closure by mobilization of mucosa that was contiguous to the defect. In the case that this is not adequate, the raw surface may be covered by a collagen membrane reinforced with sofrattouille [6].

In conclusion, the need to report these cases remains a priority enabling the understanding of the biological nature of this disease [1] [4] [6] [7] [8] [9].

### Authors' Contributions

Dr Osundwa Tom was the primary surgeon and performed the surgery. Dr Safari Karume and Dr Nelson Oduor assisted with the surgery and follow-up. Dr Mary Mungania was the histopathologist for the case.

### Ethical Approval

Ethical approval was sought from the Gertrude's Children Hospital Ethical Review Board.

### Consent

Consent for publication including the use of pictures and any other material has

been obtained from the mother to the case and is readily available in case needed.

### Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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