

Case Report

Melanotic Neuroectodermal Tumor of Infant: Two Case Reports from Sulaimani

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Abstract

Melanotic neuroectodermal tumor of infancy is a rare osteolytic-pigmented neoplasm that primarily affects the maxilla of newborn infants. Two cases of melanotic neuroectodermal tumor of infancy were presented to the department of oral and maxillofacial pathology in Sulaimani Dental College, 2015. Clinical assessment and histologic diagnosis of the classic findings supported the final diagnosis, and no recurrence was detected after one year follow up.

Keywords: Melanin tumor, Maxilla, Benign, Infant.

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Introduction

Melanotic neuroectodermal tumor of infant (MNTI) originated from the neural crest cells and it was first described by Krompecher in 1918 as a congenital melanocarcinoma⁽¹⁾. Later on, a variety of different names were given to this lesion depending on the attempts of researchers to find the cell of origin such as pigmented ameloblastoma, retinal anlage tumor, melanotic progonoma, melanotic adamantinoma and pigmented epulis of infancy, all suspected origin from the odontogenic apparatus^(2,3).

Clinically, MNTI is a rare benign tumor but aggressive with a high recurrence rate. It is a rapidly growing, not capsulated mass and may cause tooth displacement⁽⁴⁾. It appears as soft dark pigmented, sessile, non-ulcerative and asymptomatic that primarily affects the maxilla of infants during their first year of life^(5,6). Primarily affects the calvarium or facial skeleton of children and the most common site of occurrence is the anterior maxillary alveolar ridge (70% of cases) followed by the skull, brain and mandible. The genital organs are the most frequent extracranial sites^(7,8). Regarding lesions affecting the mandible; are rare and account for 6% of all cases⁽⁹⁾.

Plain radiographs of skull always show an osteolytic lesion, i.e., intrabony radiolucent lesion with a poorly defined sclerotic margin and hyperostosis^(5,10). Computed tomography (CT) scans reveal mainly hyperdense masses, however, hypodense variants have been reported as well. Although the imaging procedures define the extent of the lesion and its osseous involvement in surgical planning, it is not specifically diagnostic⁽¹¹⁾. Maxillary lesions usually demonstrate the radiolucent bony lesion causing bony expansion, sometimes with “free-floating teeth”⁽¹²⁾.

Histopathologically, MNTI shows two distinct populations of cells that form nests or alveolar-like structures: large polygonal epithelioid cells, containing melanin, resembling melanocytes, and small, round lymphocyte or neuroblast-like cells⁽⁶⁾. Complete resection is the conventional treatment and recurrence rates vary from 10% to 60%⁽⁵⁾. The recurrent tumors grow more aggressively and can invade other anatomic structures, such as nasal cavity, orbit and the skull base⁽¹³⁾. In this article, we reported two cases of MNTI; the clinical and histopathological features were represented.

Clinical presentation

Case no.1: A 5-months old female referred to a maxillofacial surgery department with a swelling of the right upper face. Intra-orally, a firm non-ulcerated mass was observed having a reddish-blue surface in buccal mucosa side and pale-pinkish color on the palatal side. The mass was sessile of about 3 × 2.5 × 1 cm dimensions located on the right posterior maxillary-alveolar region (Figure 1A). The mass was attached firmly to the deeper structures. The parents noticed the swelling at the age of 3 months, which was slowly progressing initially and started growing rapidly for the next two months. A previous incisional biopsy suggested a fibrous or vascular lesion. Provisional diagnosis included: giant cell granuloma, a melanotic neuroectodermal tumor of infancy and congenital eruption cyst.

The CT and Cone beam computed tomography (CBCT) scans of the head region of case no.1 showed a large growing, well-defined hypodense lesion in the 2nd deciduous molar region of right maxilla with eroded, irregular and expanded cortical plates. Invasion of the lesion into the right nasal sinus is also obvious. The lesion leads to a buccal displacement of the deciduous 1st molar and destruction of the alveolar ridge and lateral posterior palate (Figure 1B, C and D).

Case no.2: A 6-months old male presented with an extensive, firm, painless and bluish-purple mass located in the premaxilla. Facial asymmetry was obvious because the lesion lifted the right upper lip which was dry and ulcerated (Figure 2A). The mass surface appeared smooth but with grooves measuring 2 × 2.5 cm in size during the last three months. A curved neonatal tooth was merging through the mass from the palatal side (Figure 2B). The child parents notice growth of the mass rapidly in the last two weeks. Unfortunately, the surgeon didn't document any imaging on referral to the laboratory.

In both cases, there was no history of medication during pregnancy and no relevant medical or congenital problems were present. Growth and development of the infants were normal for their age. On palpation, the swelling was non-tender, firm in consistency and non-fluctuant. In fact, the tumor prevents baby feeding and the parents were overstrung and scared about their baby's condition. Provisional diagnosis included: congenital epulis, teratoma, congenital eruption cyst, and hemangioma.

Treatment and gross features

Under general anesthesia, excisional biopsy was performed with free margins and the involved teeth

were removed. The definitive diagnosis of MNTI is based on the histologic evaluation of a surgical specimen. Grossly, the specimen of case no.1 was intact

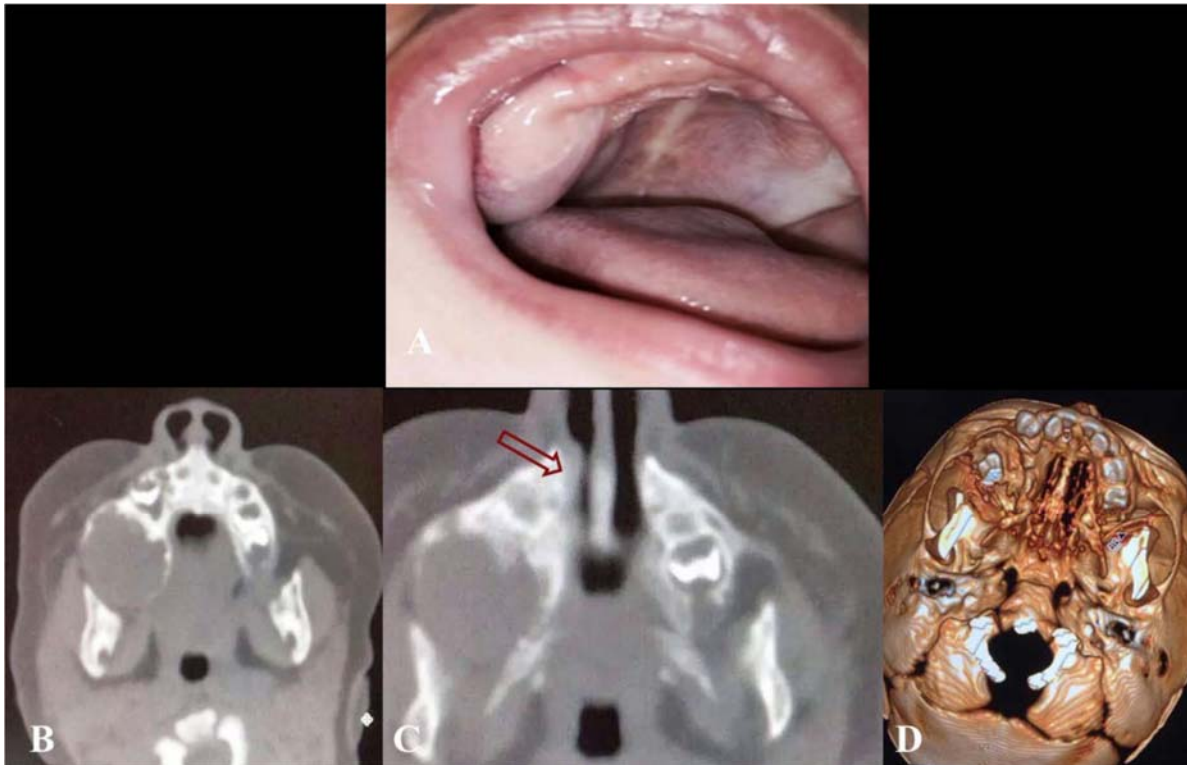


Figure 1: Clinical photograph of MNTI of case no.1, A- Shows a sessile mass in the right posterior maxillary region, B- CT scan demonstrates expansive lytic lesion surrounded by the eroded cortical plates of the right maxilla, C- A deeper section of the CT image showing invasion of the lesion into the right nasal sinus (arrow), D- CBCT axial image showing the lesion enclosing the 2nd molar and a buccal displacement of the 1st molar and causing destruction of the alveolar ridge and lateral posterior palate.



Figure 2: Clinical pictures of MNTI of case no.2. A- Bluish mass on premaxilla lifting upper lip which appears ulcerated and fissured, B- Oral view of the lesion with a neonatal tooth merging on the palatal side.

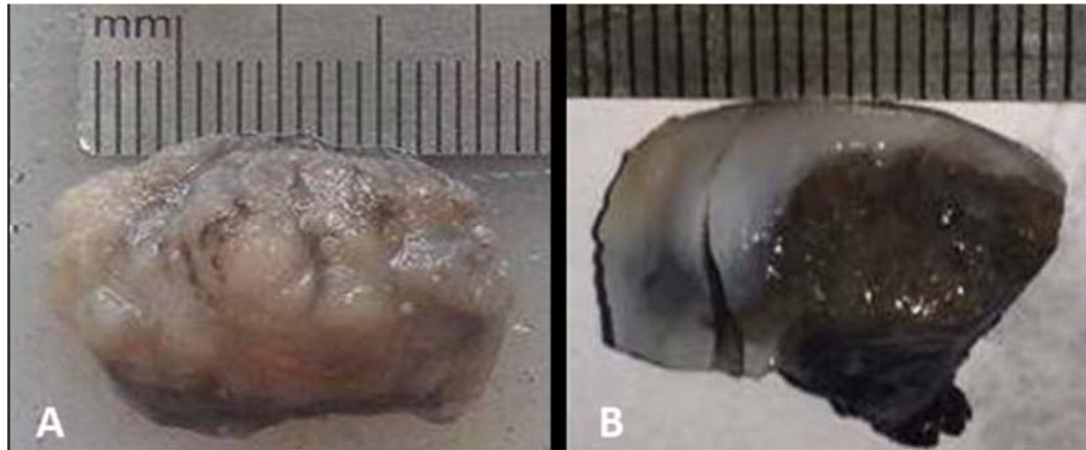


Figure 3: Gross specimens, A- Case no.1 shows a well-circumscribed mass with foci of blue-black pigmentation, B- Case no.2 shows a cut-piece mass with white- grayish surface covering a gray to bluish-black core.

well-circumscribed mass, firm-rubbery inconsistency with foci of blue-black pigmentation, measuring $2.3 \times 2 \times 2$ cm. The cut surface was grayish-white in color with black pigmented areas (Figure 3A). The sample of

case no.2 was cut-piece (remaining tissue sent to another histopathological lab) firm, and smooth white grayish surface mass, measured $2 \times 1.3 \times 1$ cm.

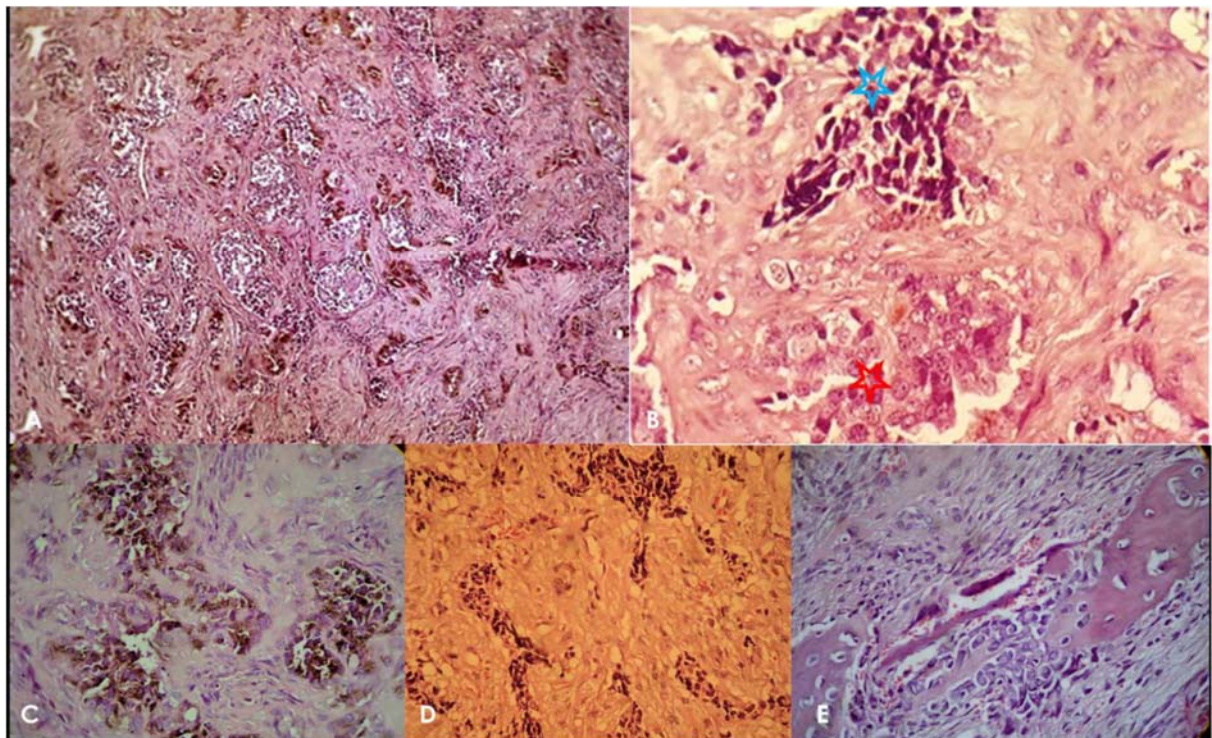


Figure 4: Histopathological pictures (H&E): A- An irregular tubular or alveolar pattern of tumor cells in a vascularized dense fibrous stroma ($\times 100$), B- Two population of the tumor cells within the fibrous stroma, large pigmented cells (red star), and small lymphocyte-like cells (blue star) with scanty cytoplasm and hyperchromatic round nuclei ($\times 400$), C- Large cells with pale abundant cytoplasm and round vesicular nucleus with brown pigment ($\times 400$), D- Small collections of the lymphocyte-like cells ($\times 400$), E- Local infiltration into the adjacent bone ($\times 400$).

The inner side of the sample was gray to bluish-black (Figure 3B).

The infants were re-examined at 1, 3 6 months, and one year after surgery and no recurrence was evident clinically. All soft tissues had healed well. The parents were reminded and motivated about the necessity of follow up every six months to monitor any recurrence and to correct any functional and cosmetic deformity along with the ongoing development and successful growth.

Histopathological evaluation

The histological appearance of MNTI was unique and characteristic as follow; the tumor was composed of epithelioid cells arranged in an irregular tubular or alveolar pattern in a vascularized dense fibrous stroma (Figure 4A). Two distinctive types of cells were seen; large cells with pale abundant cytoplasm and round vesicular nucleus with brown pigment and small lymphocyte-like or neuroblast-like cells with scanty or fibrillar cytoplasm and hyperchromatic round nuclei (Figure 4B and C). These small cells were occasionally seen as small collections in the fibrous stroma throughout the specimen (Figure 4D). This non-encapsulated tumor showed local infiltration into the adjacent bone which appeared irregularly eroded (Figure 4E).

Generally, the lesion had a normal appearance, mitotic activities were scant and no atypia was identified. Cellular pleomorphism was also scant (Figure 4E). Based on the above histopathologic findings, the lesion was finally diagnosed as MNTI.

Discussion

The rare MNTI clinically presents as a rapidly growing soft tissue swelling. It is painless, expansile and un-encapsulated partly pigmented mass typically affects the maxillary region (69% of reported cases) and more than 90% appear in the first year of life usually from age 1-6 months with a peak between the second and sixth month of life⁽¹⁴⁾. Although the tumor cells produce melanin, pigmentation may not be clinically evident^(15,16). Our first infant tumor showed slight pigmentation in comparison with that of the second case which showed a total extreme pigmentation.

The tumor tends to occur as a single lesion. However, multiple lesions have also been reported⁽¹⁷⁾. The tumor causes compression rather than infiltration of adjacent structures with local invasion causing bone destruction, teeth displacement and feeding difficulties⁽¹⁸⁾. All these clinical findings were not far from our cases description like the expansion of the alveolar ridge, and

displacement and floating of the teeth just in the affected area.

It is typical of CT scans to reveal well- demarcated hyperdense masses, but hypodense variants have been reported as well with a displacement of teeth as in 1st case. This tumor requires multimodality imaging with magnetic resonance and CT but they are complementary. The CT can accurately define the extent of the lesion, or whether is it single or multiple and thus provides a good basis for surgical planning⁽¹⁹⁾.

In addition to the typical clinical presentation, the histology was distinctive, showing the general tubular or alveolar distribution of cells, which were very easily classified into a population of small neuroblastic cells and larger melanin-containing epithelioid cells⁽²⁰⁻²²⁾. This allows its differentiation from other pediatric neoplasms like congenital epulis, teratoma, neuroblastoma, Ewing's sarcoma, rhabdomyosarcoma, melanoma, congenital eruption cyst, Burkett's lymphoma, Langerhans cell histiocytosis, and hemangioma^(23,24). The general normal looking of the lesion in our cases were: scarce mitotic figures and absence of necrosis have ruled out the suspicions of malignancy. Therefore, our cases showed a clear MNTI histopathological similarity to what had been documented in the literature.

Early detection and prompt treatment prevent complications. Most authors agreed on complete surgical excision as the treatment of choice and it was quite enough in our cases as no recurrence was reported in both cases after one year follow up. There is be no need for other modalities of treatment if the MNTI treated with surgery alone^(17,25,26). Most children adapt to the conservative surgery very well, however, some will require follow-up treatment plans like speech therapy and orthodontic care⁽²⁷⁾.

It has been reported that recurrence of the tumor may be as a consequence of incomplete removal seeding during surgery or tumor multicentricity of the primary tumor⁽²⁸⁾. It was documented that about 10-15% local recurrence rate and a 3% metastatic spread can occur. Moreover, most recurrences appear to occur within four weeks after the operation^(5,29,30). Furthermore, a malignancy rate of 6.5% had been reported for these tumors, with most malignant cases occurring in the skull and brain. Only very few tumors produce metastases and death⁽⁵⁾. Therefore, it needs a follow up for at least six months postoperatively with a clinical and radiographic evaluation to rule out recurrence. The present cases were followed for one year with no recurrence. The maxillary defect became relatively smaller as the child had grown.

Conclusions

The diagnosis of MNTI is mainly dependent on clinical and histopathological findings. Presence of two types of tumor cells, large epithelial melanin-containing and the small lymphocyte-like cells, which are embedded in the fibrovascular stroma are specifically diagnostic and separating this lesion from other similar infantile pathological entities. It is worthy to say that these conditions are rare but not uncommon and should be treated as soon as possible to prevent the aggressive expansible behavior of this tumor and restore functional activities for infants with a reduction in recurrence chances. We have to emphasize on receiving all the information of any biopsy referral from the dental surgeons who are in charge. The two reported cases were followed up to one year and reveal no signs of any recurrence.

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