

| | |
|------------|--|
| 原文題目(出處)： | Melanotic neuroectodermal tumor of infancy in the maxilla. Case Rep Dent 2013, Article ID 726815 |
| 原文作者姓名： | Daniel Falbo Martins de Souza, Daniel Isaac Sendyk, Juliana Seo, Eduardo Vasques da Fonseca, Maria da Graça Naclério-Homem, Maria Cristina Zindel Deboni |
| 通訊作者學校： | University of Sao Paulo, Avenida, Brazil |
| 報告者姓名(組別)： | 陳苾溱 Intern E 組 |
| 報告日期： | 102/12/10 |

內文：

一、 Abstract

1. Melanotic neuroectodermal tumors of infancy (MNTIs) are rare fast-growing tumors with high recurrence rates.
2. These tumors, which originate in the neural crest, commonly occur in the anterior maxilla of children under the age of one.
3. Here, we describe an MNTI case in a two-month-old girl with increasing swelling in the left cheek. MNTI was diagnosed in this case following tomography and biopsy. The patient's histological and immunohistochemical profile indicated a remarkable combination of neural, melanocytic, and epithelial cell differentiation.
4. Despite complete surgical excision, careful followup is recommended. In addition, maxillary functional orthopedics and reconstruction may be necessary in cases of MNTI.
5. Approximately 260 cases of MNTI have been reported since this type of tumor was first described.

二、 Introduction

1. Melanotic neuroectodermal tumors of infancy (MNTIs) are rare, fast-growing, melanin-containing lesions that commonly occur in the head and neck regions of children under the age of one.
2. MNTIs are nonulcerative, painless, and pigmented lesions, but the pigmentation cannot always be observed through the covering tissues.
3. Uncertainties regarding the histogenesis of MNTIs have led authors in the literature to use a diverse nomenclature, and MNTIs have been described as congenital melanocarcinomas, atypical ameloblastomas and melanocytomas.
4. Neural crest is accepted to be the origin of these types of tumors.
5. MNTIs generally occur in the maxilla (68%–80%), but they can occasionally arise in the skull (10.8%), mandible (5.8%) or brain (4.3%). In addition to the head and neck region, other sites can be affected by the condition less frequently, including the femur, epididymis, ovaries, uterus and mediastinum.
6. The majority of publications reported no significant effects of gender.
7. MNTI lesions are regarded as benign tumors, although they can present locally aggressive behavior, including gradual invasion of the surrounding bone and sinuses. High recurrence rate that varies between 10% and 60% and the risk of malignant transformation is 6.6%.
8. MNTI appears as an intrabony expansive radiolucency, usually with poorly demarcated margins, which likely result from the rapid tumor growth of MNTIs and their tendency to be locally invasive.

三、 Case Presentation

1. A two-month-old girl was referred to the Oral and Maxillofacial Surgery Department of Conjunto Hospitalar do Mandaqui (São Paulo, Brazil) and

- presented with a mouth tumefaction with one-month evolution.
2. Clinical examination(extraoral)
 - a. facial asymmetry,
 - b. deletion of the left nasolabial folds
 - c. elevation of the left nasal alar base
 3. Clinical examination(intraoral)
 - a. A left premaxilla tumefaction was observed in the alveolar ridge near the canine region. The overlying mucosa was hyperemic, and the labial frenulum was distended.
 - b. Palpation revealed a lesion with well-defined limits, a smooth surface, and elastic consistency.
 4. Radiological examination: Tomography images revealed a homogeneous hypodense tumor that was associated with the upper left central primary incisor.
 5. Lesion aspiration produced negative results.
 6. Differential diagnosis: odontogenic tumor
 7. Histopathologically:
 - a. Fragments of tissue characterized by the proliferation of a dual population of cells arranged in solid nests or cords in the middle of dense, well-cellularized connective tissue.
 - b. The first cell type consisted of small rounded hyperchromatic cells with minimal cytoplasm that resembled neuroblast-like cells with delicate fibrils between them.
 - c. The second cell population consisted of epithelioid cells, some of which contained brown intracellular granules, similar to melanocytes.
 8. Definitive diagnosis:
To correctly identify the different cell types, an immunohistochemical panel of specific antibodies was performed.--> Melanotic neuroectodermal tumors of infancy (MNTIs)
 9. Operative course:
 - a. A peripheral ostectomy was performed to assure total tumor excision.
 - b. The surgical piece indicated a fibrous blackish-brown lesion containing two primary teeth within the tumor mass.
 10. Postoperative course:
At the time of the one-year follow-up appointment, clinical and tomography examinations did not reveal any tumor recurrence. The child was referred to a maxillary functional orthopedic professional for future attendance and treatment.

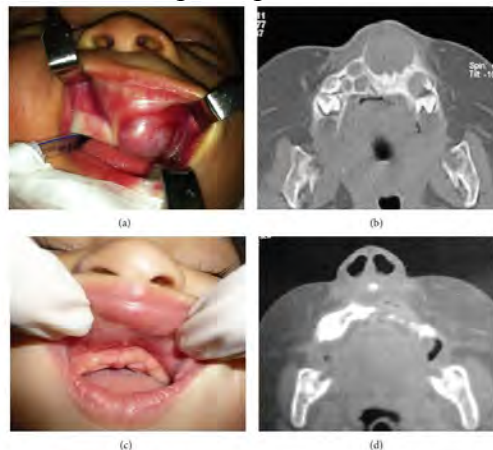


FIGURE 1: Upon an intraoral assessment, swelling in the left premaxilla alveolar ridge near the canine pillar (a) was observed. Preoperative tomography images (b) revealed a homogeneous hypodense tumor associated with the upper left central primary incisor. An image showing the one-year postoperative intraoral aspect (c). Postoperative tomography image presenting a maxilla defect but no lesion recurrence is shown (d).

四、 Discussion-MNTI

1. Characteristic: Rare, fast-growing, melanin-containing lesions. Nonulcerative, painless, and pigmented lesions. Described as congenital melanocarcinomas, atypical ameloblastomas and melanocytomas. No significant effects of gender. Regarded as benign tumors.
2. Radiographs: Intrabony expansive radiolucency, usually with poorly demarcated margins, which likely result from the rapid tumor growth of MNTIs and their tendency to be locally invasive.
3. Histology: Uncertainties regarding the histogenesis of MNTIs
4. Pervallence : Head and neck regions of children under the age of one. MNTIs generally occur in the maxilla (68%–80%), but they can occasionally arise in the skull (10.8%), mandible (5.8%) or brain (4.3%). High recurrence rate that varies between 10% and 60% and the risk of malignant transformation is 6.6%.
5. Differential diagnosis: odontogenic tumor

五、 Conclusion

| 題號 | 題目 |
|--------|--|
| 1 | 下列關於 Melanotic neuroectodermal tumors of infancy (MNTIs)的敘述何者錯誤? (A) 此疾病與色素沉積有關 (B) 好發年齡小於 1 歲 (C) 最常在 Ant. maxilla 發現 (D) 局部侵犯性高,可能侵犯到周圍的 bone 和 sinuses,故定義為 malignant tumor |
| 答案 (D) | 出處：Oral and maxillofacial pathology 3 rd edition , Neville, et al p.533-535 |
| 題號 | 題目 |
| 2 | 下列何者為 Melanotic neuroectodermal tumors of infancy (MNTIs)的特徵? (A) 阻生牙(impacted tooth) (B) 咖啡牛奶斑(café'-au-lait spot) (C) 黑色素沉積(melanin) (D) 與陽光曝曬有關,常發生在下唇 |
| 答案 (C) | 出處：Oral and maxillofacial pathology 3 rd edition , Neville, et al p. 533-535 |