原文題目(出處)：	Melanotic neuroectodermal tumor of infancy in an African-indigenous patient from the Amazon: a case report Head & Face Medicine 2013, 9:35.
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內文：

Abstract:

1. Melanotic neuroectodermal tumor of infancy (MNTI) is a rare condition that occurs normally in the anterior maxilla of infants aged <1 year.
2. This paper describes a case of a 2-month-old male of African-indigenous descent and Brazilian Amazon residency, who presented to our unit in 2009 with a history of an expanding mass involving the anterior maxilla.

Background:

1. Melanotic neuroectodermal tumor of infancy (MNTI) is an extremely unusual, benign osteolytic neoplasm of neural crest origin
2. previously referred to as melanoameloblastoma, pigmented congenital epulis, melanotic progonoma, and other synonymous terms , and is classified as a congenital anomaly of the face and neck according to WHO ICD-10
3. MNTI affects infants in the first year of life with no sex predilection , and since the time it was first reported by Krompecher in 1918
4. 237 cases (65.5%) involving the maxilla alone

Case presentation:

1. A 2-month-old normally developed male of African indigenous descent (a background in Brazil referred to as “cafuzo”) with no relevant medical history presented to the oral surgery department at Ophir Loyola Hospital (Belem, Para, Brazil) in 2009 with a history of an expanding mass that involved the anterior maxilla, observed since his birth, that interfered with breathing and feeding.
2. Oral examination showed prematurely erupted teeth and a firm, non-ulcerated, reddish-bluish tumor of approximately 4 cm× 4 cm in size, extending from the alveolar ridge to the hard palate, displacing the overlying cheek and lip and covered by intact mucosa.
3. Computed tomography (CT) in the soft tissue window showed an expansive mass measuring 4 × 4 × 3 cm that involved the middle of the anterior maxilla region with bone destruction, extending superiorly and medially, just inferior to the ethmoid air cells. Several unerupted tooth buds were displaced laterally.

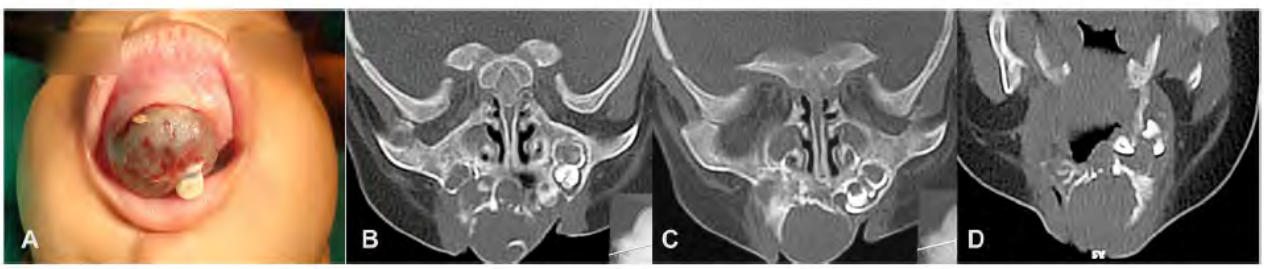

Figure 1 Prematurely erupted teeth with a firm, non-ulcerated, reddish-bluish tumor approximately 4 cm × 4 cm in size, extending from the alveolar ridge to the hard palate and covered by intact mucosa. A, B, C, D: Pre-operative axial computed tomography scans showing an erosive and expansive lesion extending from the alveolar ridge to the hard palate and regional displacement of teeth.

4. The chosen treatment was enucleation.

5. The lesion was large and well bonded to the maxilla, permitting a two-piece excision of the mass.
6. The facial growth has been normal thus far, but long-term oral rehabilitation will be necessary because of teeth extraction. No recurrence was detected.
7. A microscopic biopsy later showed that the tumor was composed of two different types of cells.
8. One portion of the lesion presented smaller round cells with minimal cytoplasm and hyperchromatic nuclei
9. The other portion exhibited larger cells with vesicular nuclei and eosinophilic cytoplasm containing typically abundant brown intracellular melanin granules.
10. The first cell population (neuroblast-like cells) was arranged in nests or in alveolar patterns surrounded by the larger ectodermal cells, separated by fibrovascular stroma; this biphasic pattern characterized MNTI.

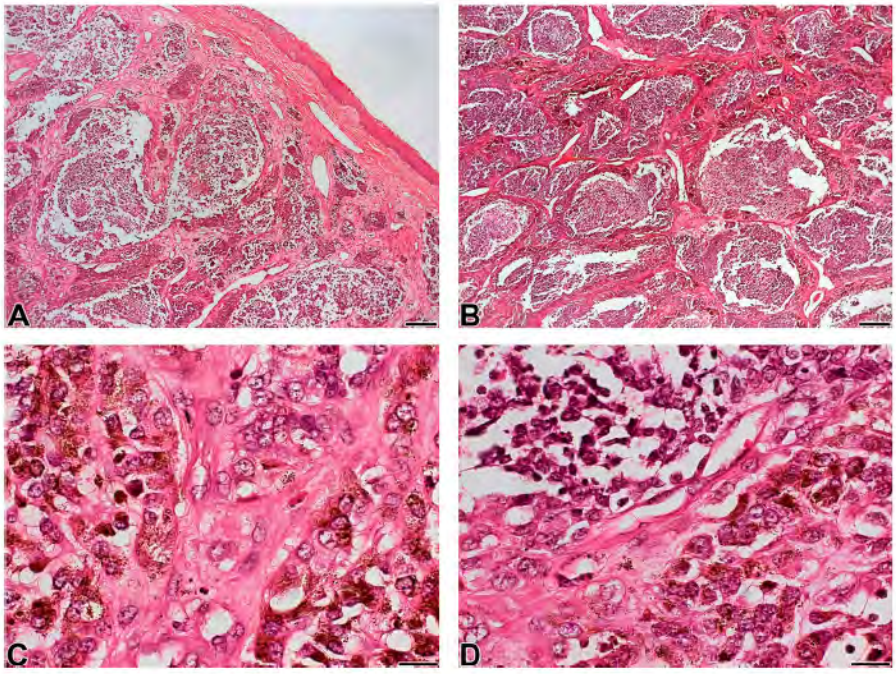

Figure 3 A, B: Postsurgical facial appearance. A, **B**: Note the alveolar pattern and the fibrous stroma (hematoxylin and eosin, scale bars 200 µm); **C**: Larger cells with intracellular melanin granules (Hematoxylin and eosin, scale bar 20 µm); **D**: The biphasic microscopic pattern (Hematoxylin and eosin, scale bars 20 µm).

Discussion:

1. Lesion involving the maxilla, which affects 68–80% of children in the first year of life.
2. The lesions are usually benign, presenting as an aggressive, quick growing, and painless mass in the anterior maxilla with irregular reddish-bluish pigmentation swelling on the top of the alveolar crest.
3. This mass may have a rubbery consistency and non-ulcerated surface, containing prematurely erupted or displaced primary teeth
4. Generally, these tumors are asymptomatic lesions, noticed by parents because of asymmetry caused by a growing mass in the midface region [12] that displaces teeth and deforms the patient's face.
5. In our reported case, the patient's mother had noted the lesion since birth, and it was treated 2 months later, affecting function as well as the facial features in the breastfeeding child.
6. The child had 5 teeth missing up to the age of 10 years. This condition could lead to facial development problems; therefore, frequent dental follow-up examinations are required, with oral rehabilitation treatment.
7. Despite the classic features mentioned, this patient was of "cafuzo" descent living in the Brazilian Amazon. Among all the cases described in Table 1 between 2000–2011, this is the

only report of MNTI in a patient with this specific ethnic background and residence.

8. We suggest that the behavior of MNTI in “cafuzo” patients is similar to that in individuals of other races, showing the influence of familial ethnicity (a mix of African and the indigenous Brazilian population)
9. Radiographs and CT images have revealed the following characteristics of MNTI: tissue destruction, poorly demarcated radiolucency, hypodense underlying bone, occasional faint “sunburst” appearance resulting from mild calcifications along vessels radiating from the center of the tumor.
10. The surgical approach for lesion removal was enucleation. This conservative surgical approach consists of local excision and curettage, and prevents aesthetic sequelae with reduced manipulation of the lesion.
11. While extent of surgical excision is debatable in the literature , the chosen surgical method seems essential to reduce the possibility of local recurrence rates to approximately 10–15% and usually leads to a good prognosis
12. In spite of the classical clinical and imaging features presented in our case report, only a histopathological evaluation could confirm MNTI

Conclusion:

1. The low number of MNTI cases reported in “cafuzo” patients and in the residents of the Brazilian Amazon region demonstrates the need to register such cases and improve the local dentists’ knowledge about the classical features of MNTI to facilitate a preliminary diagnosis.
2. Careful management of post-surgical consequences such as the loss of teeth and bone support is needed in patients of mixed ethnic backgrounds such as our patient.
3. When designing facial orthopedics to help with prosthetic rehabilitation and/or reconstructive surgery in adults, the integrity of the facial pattern in the region should be preserved, providing an aesthetic face.

題號	**題目**
1	一位出生半年的嬰兒，媽媽發現在小孩的上顎前區（anterior maxilla）出現一個外觀呈藍黑色且生長快速的腫塊，X 光檢查發現顎骨有破壞情形，且發育中的牙齒發生位移現象。請問小孩可能罹患下列何種疾病: (A) Pindborg tumor (B) ameloblastoma (C) melanotic neuroectodermal tumor of infancy (D) melanoma
答案(C)	出處：Oral and maxillofacial pathology 3rd edition , Neville, et al p.533-535
題號	**題目**
2	下列關於 Melanotic neuroectodermal tumors of infancy (MNTIs)的敘述何者錯誤?
	(A) 好發於不足一歲的嬰兒 (B) 最常在 lower jaw bone 發現 (C) 局部侵犯性高，且生長快速 (D) 大多為良性
答案(B)	出處：Oral and maxillofacial pathology 3rd edition , Neville, et al p.533-535