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Melanotic neuroectodermal tumor of infancy in the maxilla: A rare Case Report and literature review

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Abstract:

Melanotic neuroectodermal tumour of infancy (MNTI) is a rare neoplasm derived from neural crest cells. Although it is fundamentally benign, the tumour may present a locally aggressive behaviour, characterised by a rapid progression and a destructive invasion of adjacent structures, hence causing deformities. It primarily affects the maxilla of infants during the first year of life. Surgical excision is considered as the treatment of choice. The recurrence rate varies between 10% and 15%, and malignant behaviour has been reported in 6.5% of cases. We report a case of MNTI in 7-month-old male child. We discuss the clinical, radiographic and histologic features of this rare tumour, as well as its surgical management and the follow-up.

Introduction:

Melanotic neuroectodermal tumor of infancy (MNTIs) is an uncommon,rare pigmented neoplasm of neural crest origin(1).It was first reported in 1918 and designated with various synonym such as congenital melanocarcinoma, retinal anlage tumor, pigmented congenital epulis, or melanotic progonom(2), (3). Even if MNTIs are pigmented,the pigmentation cannot always be observed through the covering tissues (4).The majority of cases (90%) are located in the head and neck, generally, in the anterior region of the maxilla (about 70%) and may be associated with displacement of developing teeth (5).It can also occur in other locations, including the skull, mandible, brain and epididymis. (3) ;(4).

MNTI is found mainly in the first year of life. A small number of cases have been reported in older children and adults with no gender predilection (6). This benign entity is characterized with rapid expansile, local aggressivity and high recurrence risk rate between 10-15%. Malignant behavior has occasionally been reported in these tumors (5)(7).

Clinical and radiological findings as well as histopathological examination are required for accurate diagnosis (2). Complete surgical excision is generally the treatment choice for this tumor. (4)

We report a case of MNTI originating in the maxilla of a 7-month-old male infant, which was managed with complete excision with vital structures preservation.

Clinical case:

A 7-month-old patient was referred to the oral surgery department, Farhat Hached Teaching Hospital Sousse Tunisia with a chief complaint of swelling on the maxillary left anterior alveolar ridge limiting food intake because of pain, as well as mechanical difficulty for suction. His mother noticed ‘‘a mass in his upper jaw’’ which progressed rapidly since 1½ month.

No history of fever, cold, and cough was noticed. The child was immunized adequately. General examination was unremarkable.

The extra oral examination showed a deletion of nasogenian sulcus, there were no palpable lymph nodes.

Intraoral examination showed a mass which was soft to firm in consistency, compressible, non-fluctuant, painless, lobed sessile, well-defined measuring 3 cm x 2 cm, fixed to the maxillary anterior region and covered by intact mucosa. The tooth of Central Incisor -Primary was seen approximately in the center of the lesion. An incisional biopsy was taken and provisional diagnosis of fibro-osseous lesion of left maxilla was made

The swelling then began to increase rapidly in size and areas of ulceration were also present on account of trauma from biopsy.

Computed Tomography (CT) scan was conducted with three-dimensional reconstruction, showing a defined and expansive osteolytic lesion of isodense content measuring 27.4 mm x 13.7 mm x 17.6 mm [Figures 2 and 3], unerupted tooth buds were displaced laterally. Sclerotic expansion of the bone was noted and crossing the midline for a small extent.

A benign odontogenic tumor of the maxilla was the first diagnostic hypothesis. A surgical excision was performed under general anesthesia.

The surgical procedure included an intraoral approach from the maxillary anterior region with an incision made in the mucosa along the tumor boundaries. Total excision of the tumor, curettage and osteotomy of the bony structures adjacent to the tumor margins was made (Fig. 1C). Macroscopic exam showed a brownish firm lesion including unerupted primary tooth germs (51,52,53) (Fig. 1D).

The inner brown-colored aspect of the lesion raised suspicions of MNTI.

On histopathologic examination, the tumor was composed of a dual cell population arranged in solid nests in the middle of dense connective tissue. The first type was neuroblast like cells described as small rounded hyperchromatic cells with minimal cytoplasm. The second cell population consisted of epithelioid cells, some of which contained brown intracellular granules, similar to melanocytes. Approximately 1% of the tumor cells presented proliferative activity, as demonstrated by Ki67 staining

Histological features were consistent with a small round tumor, favoring the melanotic neuroectodermal tumor of infancy.

The post-operative period was uneventful. At one-year follow-up,IRM exam was performed and showed satisfactory healing and no signs of recurrence.

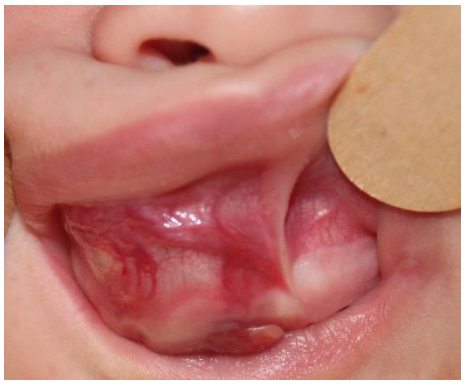

FIG 1: Intraorally appearance: Deformed contour on anterior alveolar maxillary region With firm, pigmented, ulcerated reddish-blue tumor in the maxilla

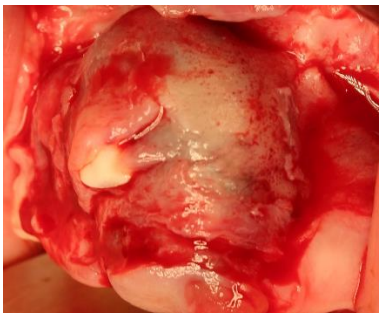
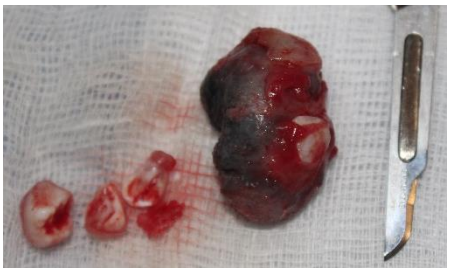
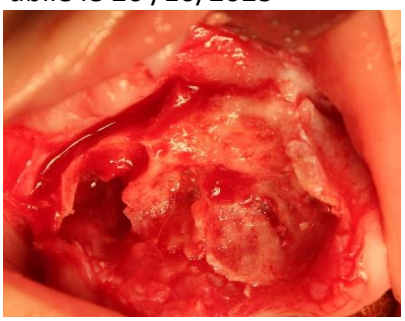
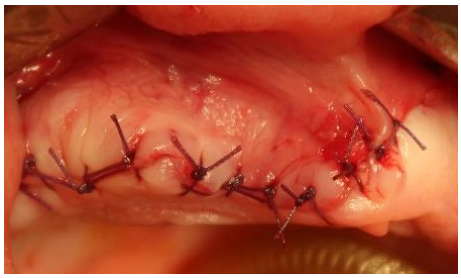
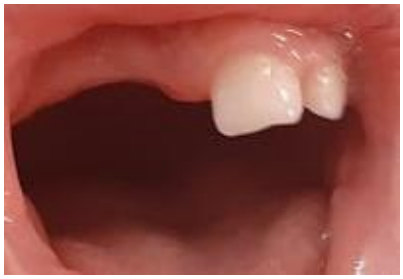

FIG 2: Surgical steps: **A**: Preoperative appearence of the lesion in maxilla:well-circumscribed soft mass,**B**: Image of the surgical specimen containing melanin pigment and primary teeth**. C**: immediate Cavity of enuclated lesion's appearance. **D**: Tension-free flap fixation with transmucosal sutures,**E:**Follow-up: intra oral view shows satisfactory healing with an erupting teeth and no signs of recurrence.

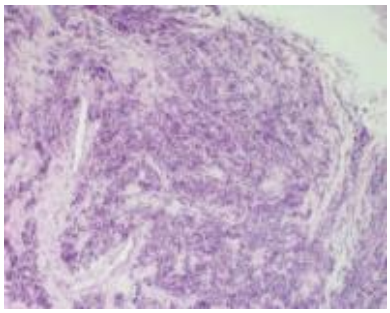
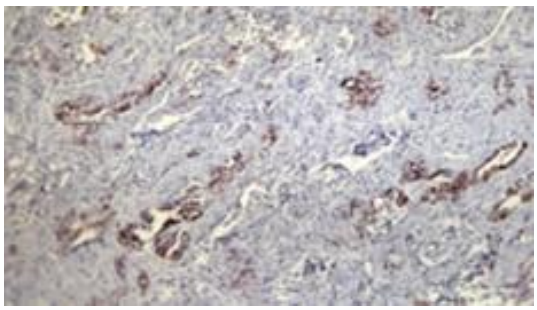

FIG 3: A:Histopathologic section of melanotic neuroectodermal tumor of infancy : small, round, melanin-containing cells,neuroblastic cells in the fibrovascular stroma (hematoxylin and eosin staining), B: Positive marker for immunohistochemistry protein S100, melanocytic marker. Cytoplasmic marker

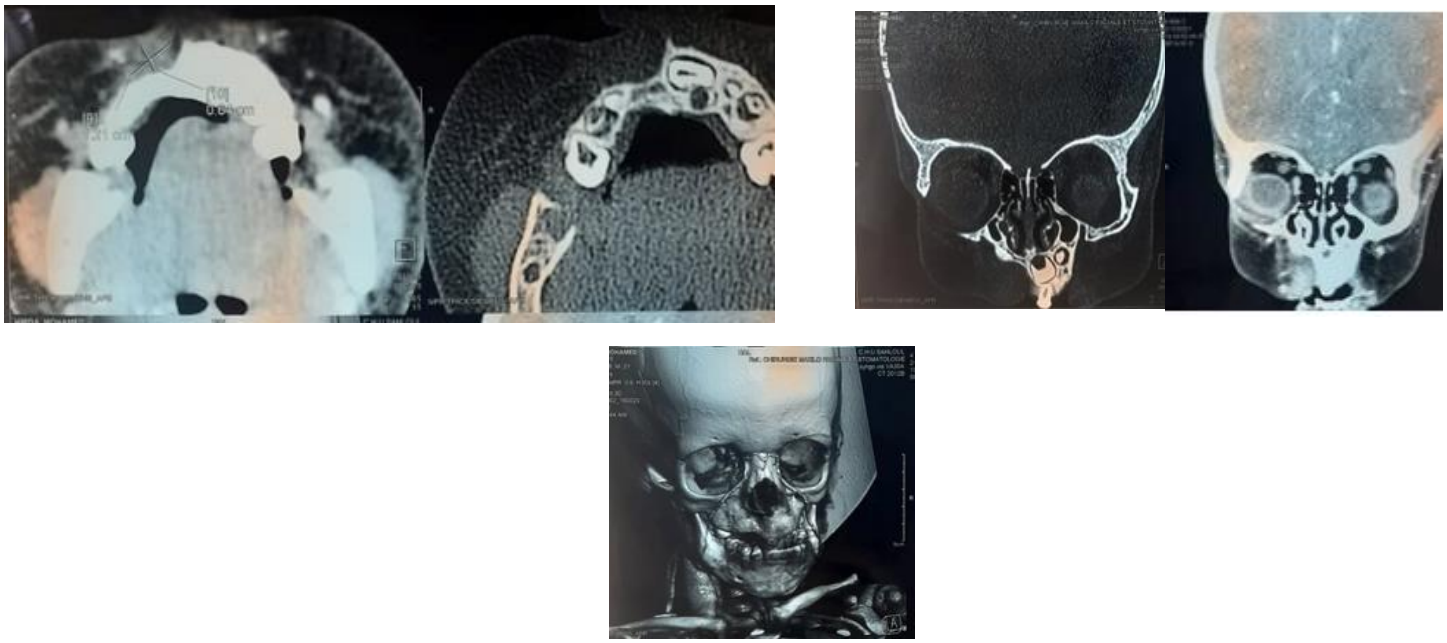

FIG 3: One year follow up IRM exam presenting a maxilla defect but no lesion recurrence is shown A: Axial ct. B: Coronal cut, C:3D reconstruction

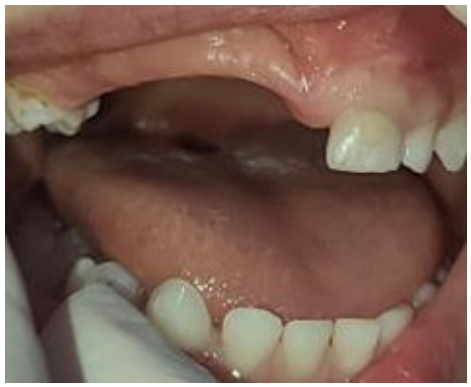

FIG4: Postoperative intraoral view (3 Years later) shows satisfactory healing with an erupting teeth and no signs of recurrence at three-years follow-up

Discussion:

MNTI is an unusual and rare type of benign, neoplasm that represents less than 1% of all benign maxilla infancy tumors. The clinical, radiological and histological findings observed in our case report were in agreement with those that have been described in the literature (2).

MNTI was previously considered as an odontogenic tumor, but histopathological examination and VMA production have confirmed the neuroectodermal origin of this tumor (3),(4).

There is a general agreement that the MNTI derived from the neural crest for several following reasons: the cells resemble neuroblasts and neurosecretory granules can be observed under electronic microscope (5),(7),(8).Moreover for diagnosing neural crest tumor origin, the catecholamine metabolite VMA levels are measured ,it usually increases in the urine and gradually returns to normal after tumor resection.

The MNTI clinically appears as a sessile-based, asymptomatic, unique, non-ulcerated, unencapsulated partly pigmented mass (2),(9) .It tends to occur as a unique lesion. However, multiple locations can also be observed

It shows an expansive rapid growth producing destruction of underlying bone, and can possibly be associated with displacement and alteration in dental development, invading neighboring anatomical structures such as the nasal cavity, orbit and base

The main differential diagnoses of MNTI includes nasopalatine cyst, ameloblastic fibroma, globulomaxillary cyst, ameloblastoma, odontogenic myxoma, rhabdomyosarcoma, Burkitt's lymphoma. (12) However, based on the typical clinical findings and radiographic appearance as well as histopathologic hallmarks, this list can be reduced. (9)

Radiographic examination of MNTI, shows as a defined isodensal lesion, causing the expansion and destruction of cortical bone, simulating "floating" teeth when presented in the maxillaries. In some cases, an associated osteogenic reaction, which exhibits a ''sun ray'' radiographic pattern, may be seen and could lead to a

misdiagnosis of osteosarcoma (8). The soft tissue component appears slightly hyperdense, which is attributed to melanin. (13)

In magnetic resonance, this tumor appears isointense or hypointense in the T1 and T2-enhanced sequences, showing improvement in contrast after gadolinium injection; the large amount of melanin pigment explains a T1- enhanced hyperintense sequence (13).The tumor may also contain areas of T1 and T2-enhanced hypointense sequences, corresponding to hyperostosis and calcifications (14).

Microscopically, MNTIs are biphasic tumors composed of small-cell and large-cell components that are arranged in nest or cord set in a vascularized fibrous stroma (10).The large, cigar-shaped, elongated melanin granules that are commonly observed in MNTI differ from those observed in melanoma, in which the granules are smaller or are unpredictably sized inclusions in melanophages (15).

The neuroblast-like cells are positive for markers like neuron-specific enolase, CD 56, the melanogenic cells are positive for HMB-45, epithelial antigen membrane antigen, cytokeratin, and vimentin (7). Expression of Ki-67/CD99 in MNTI, which is tumor cells proliferation marker is quite uncommon, might be correlated with more aggressive growth of the tumor.

There is no real consensus regarding the management of MNTI, Surgical excision is the treatment of choice for MNTI.Because of the benign nature of the tumor, most authors agree on a conservative surgical treatment (16). Some clinicians prefer simple curettage, although others advocate that a 5-mm margin of normal tissue should be obtained. (11)

The surgeon may be limited in balancing resection with an adequate margin in an attempt to preserve anatomic structures and dental germs. The biologic behavior of MNTI is of a rapidly invading lesion, which highlights the importance of prompt diagnosis to reduce the extent of radical resection (17).

Recent studies have indicated that the local recurrence rate following conservative resection varies between 10%-60%. It tends to occur as a single lesion. However, multiple lesions have also been reported (11). Recurrence may be observed if the invasion of the tumor occurred into the bone with difficulty in complete resection

(10). It can also be the consequence of incomplete removal, seeding during surgery, or tumor multicentricity of the primary tumor (17),(18).

The management of recurrent and extensive MNTI may also include adjuvant chemotherapy. Chemotherapy is not a usual treatment procedure except for in patients with confirmed metastatic diffusion. According to the literature, chemotherapy in combination with resection has also been described as a successful therapeutic option in patients with widely extended tumor or when recurrence is identified (13). Radiotherapy in combination with resection or radiotherapy in combination with chemotherapy and resection has also been described as successful therapeutic options in a few patients in the literature (15).

Postsurgical effect usually has serious consequences as it disturbs both the form and function of the normal stomatognathic system. Prosthetic rehabilitation at this stage is essential to restore the masticatory function, deglutition, speech and to prevent development of deleterious oral habits. Removable prosthetic oral rehabilitation is mandatory for patients as early as 3 years of age to stimulate a normal oral functional development. The ability of young children to adapt to removable denture prostheses is rapid, likewise oral rehabilitation at a young age can prepare them for more definitive prosthetic treatment once growth is completed.

Children with premature loss of primary teeth should be followed up for many years. Periodic recall visits should be advised, as prostheses should be evaluated and refabricated annually to account for growth and eruption of permanent teeth.

CONCLUSION:

Although MNTI is usually a benign tumor, due to its rapid growth potential and locally destructive behavior, early detection and treatment can avoid further complications and this will support a favorable outcome for the patient. However, the rarity of the tumor often leads to a delay in diagnosis, resulting in less than the desired outcome. In the present case, early diagnosis and treatment prevented further complications and the patient was followed up for one year without any recurrence.

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