Urgent Maxillectomy of Infants
(Melanotic Neuro-ectodermal Tumor of infancy)

Review of the Literature and cases Presentation

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Melanotic neuroectodermal tumor of infancy is a rare and mainly in children less than 1 year of age.

The lesion is generally accepted to be of neural crest origin.
Melanotic neuroectodermal tumor of infancy (MNTI): -

- Rare
- Mainly benign neoplasm
- Rapid expansile growth
- High recurrence rate

In the literature the tumor had a variety of names such as:-

- Congenital melanocarcinoma, (Krompecher et al., 1918).
- Retinal anlage tumor, (Clarke BE, Parsons H. 1951).
- Pigmented congenital epulis, (Lurie 1961).
- Melanotic progonoma. (Stowens D, Lin TH., 1974).

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The literature for the present analysis was selected according to the following criteria:

I - Inclusion criteria

1- These were all publications with the correct diagnosis
2- Histologic confirmation of the described MNTI from 1918 to 2009.

II - Exclusion criteria

1- Uncertain diagnosis
2- Missing of description of the tumor location
3- The lack of histology.

The review of the literature revealed:

- The total number of MNTIs published from 1918 until 2004 includes 355 cases.
- The additional one case from Kruse-Losler et al., (2006)
- One case from Heba Selim et al Department of Oral and Maxillofacial Surgery, Ain Shams University School of Dentistry, Cairo, Egypt (2008)
- 3 cases in Mansoura, Egypt (1988-2009)
- All are typical in the age of the patient, location of the lesion, clinical course, and histologic features.
- The total number of MNTIs published from 1918 until 2009 includes 400 cases.
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Origin of the tumor

Several theories concerning origin of the tumor have been proposed but:
There is general agreement that the tumor's origin is the neural crest. (Borello ED, Gorlin RJ. 1966), Cutler et al., 1981), (Misugi et al., 1965), (Jones et al., 1990), Bruce, etal, (1999) and Kruce-Losler, etal, (2006).

Sites of the tumor

- 92.8% of MNTIs occurred in the head and neck. (Kruce-Losler, etal, (2006).
  - The maxilla is main site (68%-80%) (Cutler et al., 1981). followed by:
    - The skull (10.8%),
    - Mandible (5.8%),
    - Brain (4.3%) (Cutler et al., 1981).

- 7.2% occurred in:-
  - Epididymis,
  - Mediastinum (Misugi K. et al., 1965).
  - Ovary, (Hameed K, Burslem MRG. 1970).
  - Uterus, (Schultz 1957).
  - Other sites (Johnson et al., 1983).

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This diagnosis is based on:

- Clinically presents as a rapidly growing, painless, expansile, partly pigmented mass, typically in the maxillary region,
- It tends to occur as a single lesion; however, multiple lesions have also been reported. (Steinberg et al., 1988), (Pontius et al., 1965).

The histological features include 2 cell populations:

1. Large polygonal epithelioid cells resembling melanocytes, with scattered intracellular brown granular pigmentation, (melanin)
2. Smaller neuroblast-like round cells in a stroma of fibrous tissue containing fibroblasts

- Immunohistochemical staining in MNTIs is essential.

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Imaging: -
- CT scans to reveal hyperdense masses, but hypodense variants have been reported as well.
- MRI shows a hypodense mass, with focal areas of hyperdensity in T1-weighted images and an isointense mass on T2-weighted images.


The biologic behavior of MNT:
- rapidly growing and invading
- So needs early diagnosis without unnecessary loss of time
- Late diagnosis may be the reason for difficulty in radical resection.
Malignant behavior

- Has been reported in 5 (3.1%) of the 158 cases of MNTI reviewed by Cutler et al. 1981.
- The literature review to 1989 increased the total number of malignant MNTIs to 15 (6.97%) of 215 reported cases, (Kruse-Losler, et al 2006)

The differential diagnosis of MNTI of Maxilla:

1. Neuroblastoma,
2. Ewing's sarcoma
3. Epithelioid Angiosarcoma
4. Rhabdomyosarcoma,
5. Peripheral neuroepithelioma,
6. Desmoplastic small round cell tumor,
7. Malignant melanoma,
8. Lymphoma.

Gajger de Oliveira et al., 2004
Surgical excision is the treatment for MNTI.

Recurrence:

- The local recurrence after conservative excision was 10% to 15%.
  (Pettinato et al., 1991), (Kapadia et al., 1993), (Judd et al., 1990), (Hoshina et al., 2000).

- Recurrences may be caused by incomplete removal of the primary lesion, dissemination of neoplastic cells during surgery, and multicentricity. (Nagase, et al., 1983)

- It might be due to the problem of finding the balance between radical surgical resection and the preservation of important anatomic structures. (Kruce-Losler et al., 2006).
Case 1

Need for close follow-up, especially within the first 6 months postoperatively.

Early detection and treatment of recurrence will avoid further complications and may support a favorable outcome for the patient.

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Case 2
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Case 3
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Conclusion

- MNTI is a rare and mainly benign neoplasm of early infancy.
- Rapid expansile growth and a high rate of recurrences.
- The most common site is the maxilla, but it may also occur in the mandible, skull, brain, epididymis, and other rare locations.
- The origin of the tumor is the neural crest.
- The expansive, destructive, and rapid growth of MNTI and its effects on the surrounding tissues are the most obvious clinical features.
- The definitive diagnosis should be by histopathological and immunohistochemical methods.
- Early diagnosis and radical excision can lead to excellent outcomes.
- Needs aggressive surgical treatment to avoid local recurrence.
- Proper follow up