Reviews Of Progress
ISSN:-2321-3485
ORIGINAL ARTICLE
DOI : 10.9780/2321-3485/1322013/74
Vol - 1, Issue - 33, Dec 11 2013

"Melanotic Neuroectodermal Tumour of Infancy (MNTI) – A Review"

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Abstract:

The melanotic neuroectodermal tumour of infancy (MNTI) is a fairly rare, benign, pigmented lesion that commonly occurs in the anterior maxilla of infants younger than 1 year and mainly in the first 6 months of life.Like melanocytes and nevus cells, these cells have their origin in the neural crest.This lesion has been treated with surgical excision with good results.

KEY WORDS:
Melanotic Neuroectodermal Tumour Of Infancy, Synonyms, Etiopathogenesis, Managment.

INTRODUCTION

The melanotic neuroectodermal tumour of infancy is a lesion which has had a long and controversial history concering the nature of the tumour . The tumor was first described by Krompecher in 1981[1] as a congenital melanocarcinoma.The multiplicity of terms that have been used throught the years mirrors the confusing and conflicting pathogenectic theories that have been advocated for this lesion : melanotic epithelial odontome , melanotic progonoma , pigmented adamantinoma, melanotic Ameloblastoma, congenital pigmented epulis , retina anlage tumour, melanocytoma, and pigmented neuroectodermal tumour of infancy.A complete list of synonyms can be found in a report by Nozicka and Spacek[2]. The current consensus favours the use of the term melanotic neuroectodermal tumour of infancy , a designation accepted by and used in the 1992 World Health Organization (WHO) classification of odontogenic tumours[3] .As the end of 2000, an estimated 250 cases of MNTIs have been published in the literature .

CLINICAL FEATURES

Melanotic neuroectodermal tumour of infancy almost always develops in young children during the first year of life; only 9% of cases are diagnosed after the age of 12 months .There is a striking predilection for the maxilla, which accounts for 61% of reported sites include the skull (16%),epididymis and testis (9%), mandible (6%) , and brain (6%).A slight male predilection has been noted[4].

The MNTI usually presenta as a rapidly enlarging exophytic mass most often localized in the anterior alvelolar ridge of the maxilla of an infant .Lesions are occasionally described in the mandible and extragnathic sites such as brain , epididymis ,uterus , ovary and mediastinum . The MNTI usually represents as a single lesion , but multiple lesions have been reported[5,6].The lesion often appears to have irregular pigmentation, although this pigmentation is not always clinically evident .The nontender growth is of a rubbery consistency , may contain prematurely erupted or displaced primary teeth and may have an

ulcerated surface .Most tumours measures from 1 to 4 cm at their greatest diameter.The case of a gigantiform MNTI occurring in a 7 – month-old girl and measuring 18 cm in greatest diameter was recently published by Bouckaert and Raubenheimer[7].

The typical radiographic appearance of the MNTI is that of an intrabony radiolucent lesion with poorly demarcated borders, presumably caused by rapid growth and a tendency to locally invade bone. The area of bone destruction may be transerved by bone septa . Teeth invoved in the lesion may appear to be floating within the radiolucent area of the tumour because they are displaced from their normal developmental sites .This radiographic appearance can understandably mislead a clinician into making a provisional diagnosis of malignancy .Another potentially misleading feature is a common osteogenic reaction that exhibits a "sunray" radiographic pattern which may be mistaken for an osteosarcoma.Computed tomography and magnetic resonance imagery[8,9] are able to define the extent of the lesion precisely , localize possible multiple sites , and thus greatly assist in surgical planning . These radiographic methods subject very young patients to significant radiation exposure, but they can provide important information and are diagnostically superior to other routine imaging methods.

PATHOGENESIS

Five principal theories regarding the origion of MNTI s have arisen from the cases reported in the literature. They may be described as

- Malignant transformation of odontogenic epithelium1
- Ameloblastoma variant[10]
- Origin from Jacobson's vomeronasal organ[11]
- Origin from retinal anlage(Progonoma)[12]
- Origin from neuroectodermal rests [13]

Little if any evidence exists to support the first four theories, whereas the fifth has considerable merit .Evidence for this derivation stems from tissue culture and immunohistochemical and ultrastructural studies[13], and the neural crest is currently the most commonly accepted tissue of origin for MNTI s.

HISTOPATHOLOGIC FEATURES

The tumor cosists of a biphasic population of cells that form nests, tubules, or alveolar structures within a dense, collagenous stroma.The alveolar and tubular structures are lined by cuboidal epithelioid cells that demonstrate vesicular nuclei and granules of dark- brown melanin pigment.The second cell type is neuroblastic in appearance and consists of small,round cells with hyperchromatic nuclei and little cytoplasm[4].

IMMUNOHISTOCHEMICAL FINDINGS

Immunohistochemically , the large melanin containing epithelium – like cells react with a monoclonal antibody directed against antihuman melanoma HMB-45[13].The same authors also found that the small lymphocyte – like cells were positive for neuron-specific enolase (NSE) .Bouckaert and Raubenheimer7 confirmed these findings and also found that vimentin was focally expressed in the pigmented cells.De Souza et al[14] studied the immunohistochemical expression of several cell cycle proteins (p53,MDM-2,cyclinD1,cyclinA, and proliferating cell nuclear antigen (PCNA) in three cases of MNTI.

BIOCHEMICAL FINDINGS

Borello and Gorlin[13] reported high urinary vanillyl mandelic acid (VMA) levels in a patient with an MNTI .The VMA levels returned to normal after excision of the tumour.In patients with MNTI s, VMA is believed to be strongly circumstantial evidence of a neuroectodermal origin.

DIFFERENTIAL DIAGNOSIS

Few other lesions present in this age group and in this characteristic location . Malignancies of early childhood such as neuroblastoma, rhabdomyosarcoma, or histiocytic tumors might be considered. Odontogenic cysts and tumors can be considered in differential diagnosis.[15]

TREATMENT & PROGNOSIS

Despite their rapid growth and potential to destroy bone, most melanotic neuroectodermal tumours of infancy are benign .The lesion is best treated by surgical excision.This treatment can usually be

accomplished with a partial maxillectomy usually curative.Many clinicians advocate a 5mm margin of healthy tissue to be included with the surgical specimen.

The average recurrence rate is 15-20 %.Approximately 1% of tumors is malignant with only rare tumors producing metastasis.[16]

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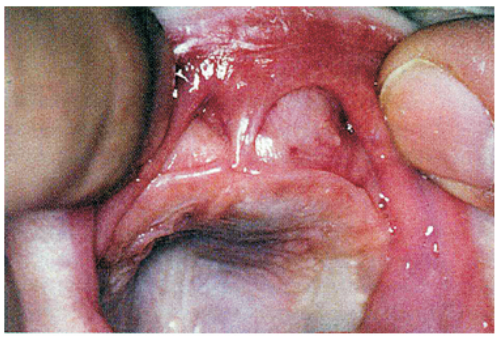
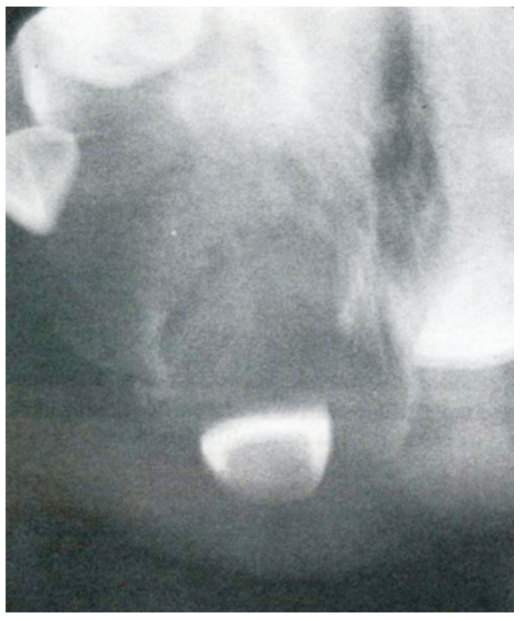
LEGENDS FOR FIGURES:

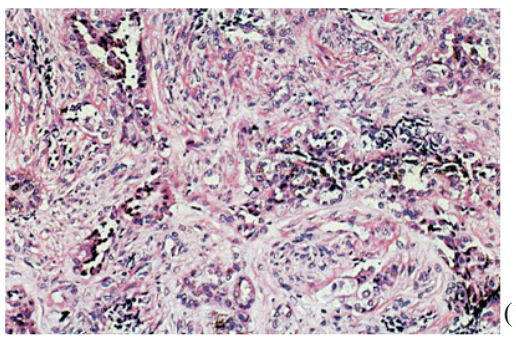
1 Photomicrograph showing.Melanotic neuroectodermal tumour of infancy .Infant with an expansile mass of the anterior maxilla (From text book Oral and Maxillofacial pathology – 3rd edition, Neville, Damm Allen Bouquot , 533-535)
2 Photomicrograph showing radiolucent destruction of the anterior maxilla associated with displacement of the developing teeth (From text book Oral and Maxillofacial pathology – 3rd edition, Neville, Damm Allen Bouquot , 533-535)
3 Photomicrograph showing nests of round cells with peripheral pigmented cells (H& E, 10x & 40x) respectively [From text book Oral pathology – clinical pathologic correlations 4th Edition, Regezi, sciubba, Jordan 201-203.]



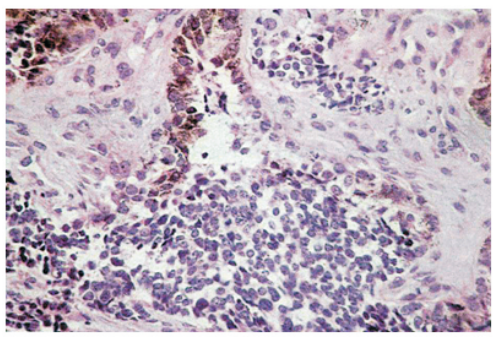
Photomicrograph – 1.Infant with an expansile mass of the anterior maxilla.



Photomicrograph – 2. Showing radiolucent destruction of the anterior maxilla associated with displacement of the developing teeth.



(10x)



(40x)

Photomicrograph – 3 Photomicrograph showing nests of round cells with peripheral pigmented cells (H& E, 10x & 40x) respectively.