# Melanotic Neuroectodermal Tumor Of Infancy (MNTI): A Case Report With Historical Insights And Review In Relation To Its Origin

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**Citation**

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**Abstract**

The melanotic neuroectodermal tumor of infancy is an uncommon, extremely rare neoplasm typically of early childhood which has a predilection for the head and neck region, particularly the maxilla. Prompt recognition of such a case is essential for the Pediatric Dentist. Presented here is case of a four month old child with a tumor involving superior maxillary alveolar ridge. The clinical assessment, histological diagnosis, and management are reviewed, with an emphasis on historical insight and reviewing its origin for proper treatment.

## INTRODUCTION

Melanotic neuroectodermal tumor of infancy (MNTI) is a rare, benign neoplasm of neural crest origin, chiefly composed of relatively primitive pigment-producing cells. It's a pigmented tumor, although the pigmentation cannot always be observed through the covering tissues.[7]

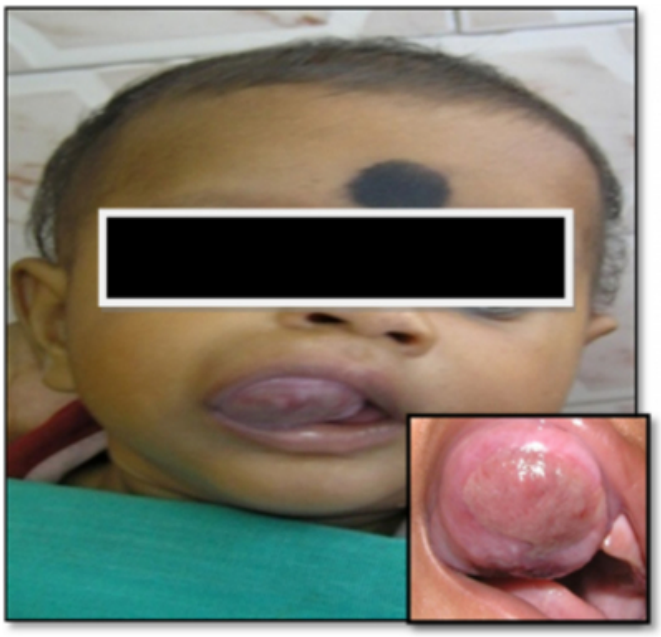
MNTI was first described by Krompecher in 1981. Since then, more than 100 cases have been reported, under a variety of names that includes melanotic prognoma, pigmented ameloblastoma, pigmented congenital epulis, pigmented teratoma, atypical melanoblastoma, congenital melanocarcinoma and melanotic epithelial odontoma.[5] The terminological variations undergone by this entity with 23 different denominations reported illustrates the uncertainties about its histogenesis.[9]. It usually follows a benign course but inadequate excision, occasional multicentricity and a small malignant potential result in a fairly high recurrence rate.[3] The expansive clinical presentation in a young child could induce the pediatrician to think in the direction of neoplastic lesions also. Thus, the present case report highlights the clinical, radiographic and histopathological palms of a case diagnosed as MNTI.

## CASE REPORT

A four-month-old infant reported to the outpatient department with a 4-month history of a smooth surfaced, pinkish- coloured, rapidly growing/protruding mass localized to the upper right anterior region since birth which continuously went on increasing to attain the present size.There was no history of airway or feeding difficulties. Maternal, birth and family history were all within normal limits. On physical examination, the child was healthy looking and vital signs were stable with no lymphadenopathy. Chest, heart and abdominal examinations were normal. Intraoral examination of the lesion revealed a firm pinkish mass with brownish pigmentation involving the anterior maxilla. The lesion was 3×4 cm in dimension, non-tender, non-pulsatile, non-reducible, non-compressible, non-fluctuant swelling with well defined border.

**Figure 1**

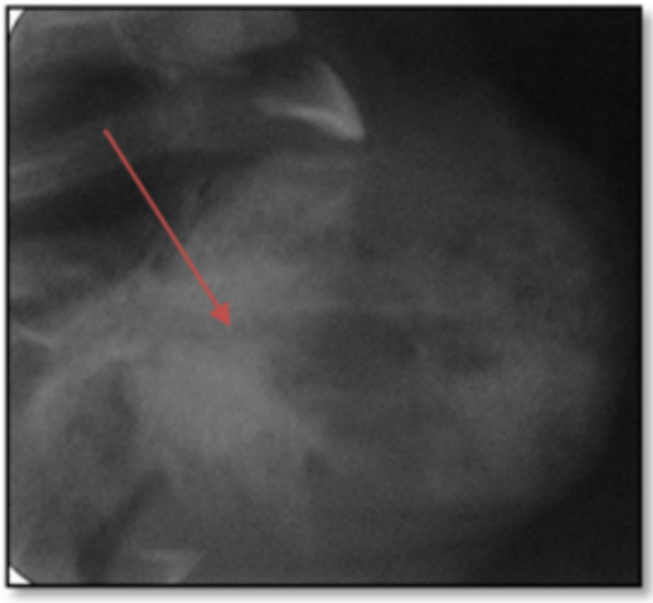
Fig. 1-Smooth surfaced, pinkish swelling in upper right anterior region



Radiographic examination revealed: hazy radio-opacity in maxillary anterior region with crown of maxillary right deciduous central incisor.

**Figure 2**

Fig. 2-Hazy radio-opacity in maxillary anterior region

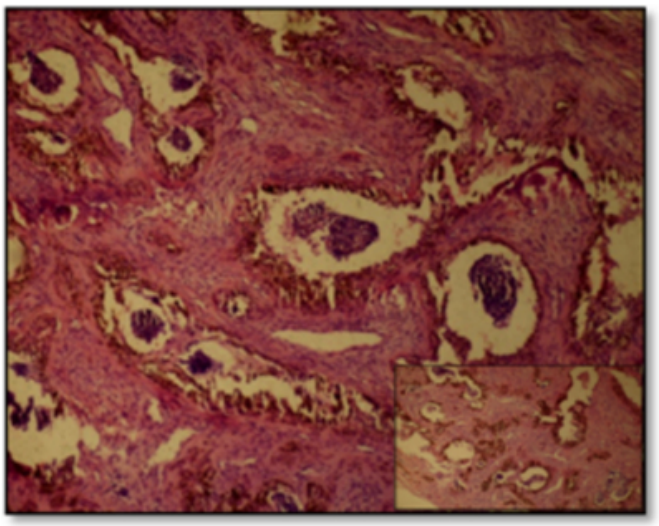


Grossly the tumour mass was grayish-to bluish-brown in colour, due to the varied amount of melanin pigmentation present. The histopathology (Fig. 4) revealed the presence of neoplastic cells which were arranged in alveolus pattern.

Each alveolus were lined by cuboidal cells, many of these contain melanin pigment and central portion of alveolus contain small round less differentiated neuroblast- like cells which show little cytoplasm and round to oval intensely staining nucleus.

**Figure 3**

Fig. 4: shows irregular alveolar spaces containing clusters of neuroblast like cells, and lined by cuboidal cells containing varying amount of melanin pigments.



## DISCUSSION

Oral and maxillofacial tumours rarely occur in the paediatric population compared with the adult population. Most jaw swellings that occur in the infant are usually benign odontogenic cysts or tumors.[5] Cutler published an extensive review of the literature since the first case report of MNT by Krompecher in 1918.[1] According to Krompecher, this tumor is derived from epithelial islands trapped during the embryonic fusion of the facial buds, but their aggressive behaviour urged him to propose the Latin term of "Melanocarcinoma Congenitum Processus Alveolaris"[9] agreeing with the benign nature of tumor commonly seen in childhood which rarely metastatize.[1] Since its description, there have been approximately 250 cases of MNTI reported in the world literature.[2] The origin and pathogenesis of this tumor is as confusing as its nomenclature and has been a matter of debate. The principle theory of origin as proposed by Krompecher (1918) that it is congenital melanoma or melanocarcinoma derived from odontogenic epithelium or from epithelial rests enclaved in the process of fusion of the maxillary process. Mummery and Pitts (1926) regarded the tumor to be a type of epithelial or melanotic odontoma or of ameloblastic origin. This view of origin of the tumor is

derived from its anatomical relationship to the tooth bearing tissues and from the fact that in a few cases, the tumor tissue was found to be closely related to the developing teeth and even connected with dental tissues. Halpert and Patzer (1947) put forward the retinal anlarge theory which is based on its hypothetic resemblance to the ciliary body and the nerve cells of the retina, but this could not explain its occurrence at multiple sites outside the oral cavity. Slowene (1957) proposed the derivation of this tumor from Jacobson's vomeronasal organ or from misplaced sensory neuroectoderm. He considered that the spindle cells, bipolar neurons and other cells may correspond to the cells of the olfactory epithelium. Misuge and colleagues (1965) suggested on the basis of ultrastructure studies that the growth is derived from the neural crest cells (NCC).[6-8] In 1966, Borello and Gorlin reported a case characterized by a high urinary excretion of vanillyl-mandelic acid classically found in a phaeochromocytoma as well as in other neuroectoblastic tumors (retinoblastoma, ganglioneuroblastoma, neuroblastoma) suggesting a neuroectodermal origin for this lesion.[9] Reviewing all the theories of pathogenesis, the NCC theory seems to be the most acceptable.[6-8.] Ultrastructural studies with electron microscopy have been used alongside histochemical staining to support the theory that MNTIs belong to a group of neuroectodermal tumors. Electron microscopy also has demonstrated that the cells appear to of NCC origin and are like melanocytes.[6-8,(10s-6),12] Elevated urinary vanillymandelic acid and the ability to differentiate primitive NCC to neuroblastic and melanocytic lineages provide additional support. Increased alpha-fetoprotein levels and positive staining for the c-myc antigen (characteristic of other neural crest tumours) have also been described.[4] Actually, taking into account the frequency of MNTI in the jawbones, the neural crest cells destined to the dental lamina should be considered in the first place. A differentiation pathway, under mesenchymal direction, could influence the tumour stroma morphogenesis: by a progressive transition from isolated fusiform cells within common fibroblasts to clusters of large pigmented cells.[16] The large cells with melanin resemble a neuroepithelium, while the small non-pigmented cells resemble immature neuroblasts, or differentiating neuroblasts.[13]. MNTI commonly invades overlying the maxilla as found in more than 70% of cases with the incisal area being the most frequently involved area, although mandibular, cranial, cerebral and genital involvement have also been described . The mean age of patients at diagnosis is 4.3 months, with a near-equal male to female ratio of 6:7.[4] Melanin is produced by the tumor; pigmentation may not be clinically evident. Local invasion may be accompanied by bony destruction, tooth displacement and feeding difficulties. Despite this locally aggressive behaviour, MNTI is generally classified as a benign tumor. A review by Cutler et al demonstrated malignant features in only 1.9% of MNTI cases, with estimates from more recent publications as high as 6%. Unfortunately, it is difficult to determine the potential for malignancy or recurrence based on clinical assessment, imaging or histopathology.[4] The pre-operative distinction of this tumour from other small round cell tumours of infancy (rhabdomyosarcoma, neuroblastoma, melanoma and lymphoma), is essential in order to plan the most complete resection and therefore reducing the possibilities of tumour recurrence[11]. Histologic appearance is distinctive, with tubular or alveolar formations of large melanin-containing cells around nests of smaller neuroblastic cells possessing fibrillar cytoplasm.[11] Immunohistochemical and ultrastructural studies reveal two types of cells: small, poorly differentiated cells that were positive for neuron-specific enolase protein and vimentin, and larger epithelial cells that were positive for melanoma antigen (HMB45) and frequently contain large and elongated melanosomes.[14] An aggressive surgical approach consisting of complete surgical excision is advocated when vital structures are not involved.

Treatment generally needs to involve a comprehensive surgical excision along with reconstruction of the involved area to marginalize the defect in the soft tissue structure of the child. Thus, the use of surgical microscopes which aid in the precisional workout of the surgery and also facilitate in the removal of all minor tissue bits related to the tumor mass is the need of the hour. Eventually though well treated, the cases need to have a thorough follow up, since disfigurement in children could be a bane to their social acceptance if surgical reconstruction is not of high standards.

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