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Melanotic Neuroectodermal Tumor of Infancy

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Keywords: Infants, Locally aggressive, Maxilla, Malignant potential

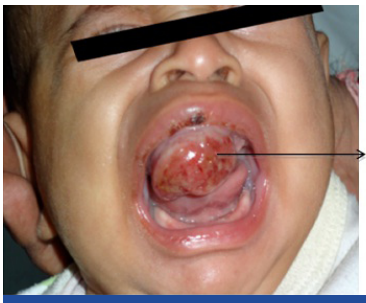
A three-months-old infant was brought to the Department of Pedodontics and Preventive Dentistry, Maulana Azad Institute of Dental Sciences, Delhi by his parents with chief complaint of swelling in upper front gums since 20 days. The onset of swelling was insidious and gradually increased in size. The delivery of child was normal and medical history was non contributory. Developmental milestones were adequate for child's age.

Intraoral examination revealed firm, tender, nodular swelling on right maxillary alveolus near midline of size 3x 2cm. The mass was tender on palpation and firm in consistency with bluish hue in color [Table/Fig-1]. CT scan of maxilla was done [Table/Fig-2-4]. and surgical excision was done under general anaesthesia [Table/Fig-5].

Histopathological examination of the excised tissue revealed the characteristic biphasic pattern of cell distribution: large epithelioid melanin containing cells and small neuroblastic cells with minimum cytoplasm [Table/Fig-6,7].

The immunohistochemical stains were positive for the following markers: Cytokeratin and HMB 45 (in larger cells), neuron specific enolase and synaptophysin (in smaller cells) while they were negative for S 100 protein.

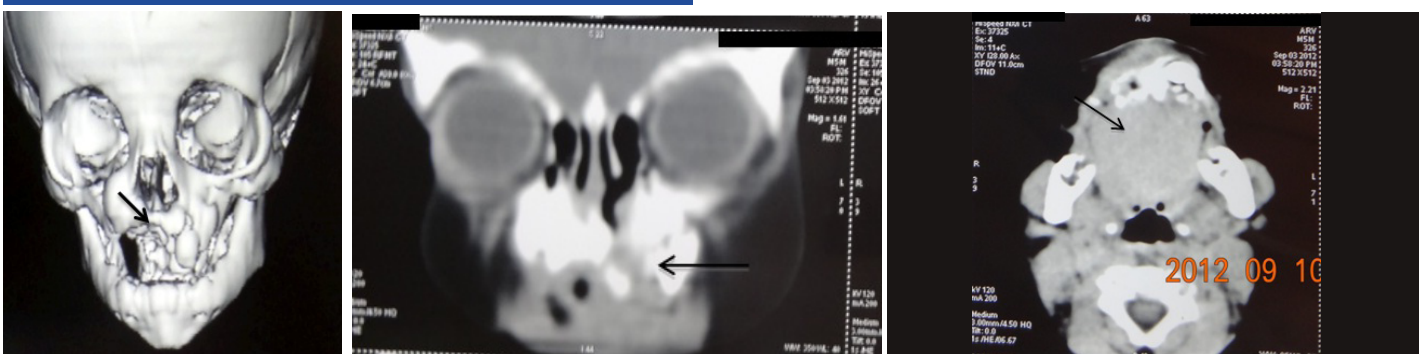
Histopathological examination and immunohistochemistry confirmed the diagnosis of Melanotic Neuroectodermal Tumor of Infancy. The swelling reappeared after three months in anterior maxilla and left infraorbital region and this time the swelling grew rapidly [Table/Fig-8]. A wide surgical excision was performed to achieve complete tumor remission [Table/Fig-9]. The patient was kept on regular follow-up till two years and no further recurrence was seen [Table/Fig-10].



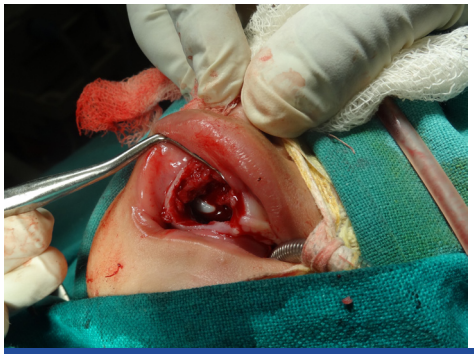
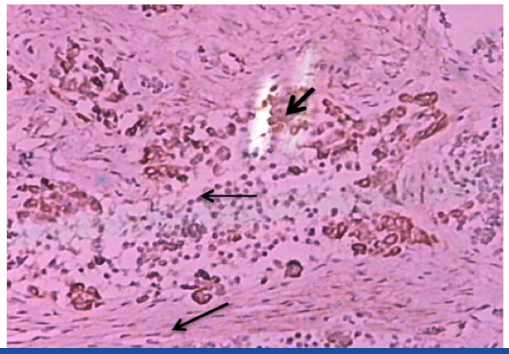
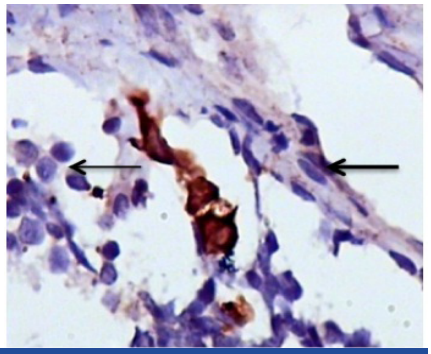
[Table/Fig-1]: Picture of patient showing swelling on maxillary alveolus.

DISCUSSION

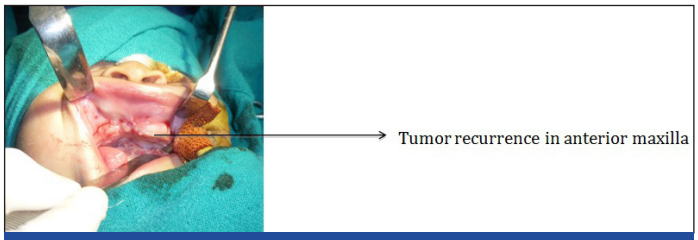
Melanotic neuroectodermal tumor of infancy (MNTI) is a rare tumor mostly involving jaw bones and arises from neural crest cells. In the medical literature, the tumor has been referred with a variety of synonyms such as congenital melanocarcinoma, pigmented ameloblastoma, retinal anlage tumor, pigmented epulis, melanotic epithelial odontome or melanotic prognoma which reflect multiple histogenesis. The mean age of affected patients is 4.3 months. It is a benign tumor which is locally aggressive with rapid onset and alarming growth rate. Patients typically present with a rapidly growing, non ulcerated mass, affecting the craniofacial region in 90% of cases [1]. Local invasion by tumor can lead to bony destruction, tooth displacement and feeding difficulties. Approximately 1% of tumors are malignant, with only rare tumors producing metastases. Plain dental radiography, Computed Tomography and MRI have been used to evaluate the site and extent of MNTI. Computed tomography imaging typically reveals a well demarcated, hyperdense lesion with contrast enhancement and hyperostosis of adjacent bone [2]. However, imaging is seldom diagnostic and biopsy of tissue is required. Histopathologically, MNTI shows a characteristic biphasic cell distribution of large epithelioid cells and small neuroblastic cells with scattered melanin pigment. Immunohistochemical stains are extremely important for confirming the diagnosis of MNTI. The larger, epithelioid cells stain positive with cytokeratin, epithelial membrane antigen, vimentin and HMB-45 and are usually non-reactive with S-100 protein. The smaller neuroblastic cells in MNTI are often positive for neurogenic markers such as synaptophysin, neuron-specific enolase and glial fibrillar acidic protein [3]. Treatment of choice for MNTI is surgical excision, with a 5mm healthy margin and it is usually curative [4]. The tumor has high recurrence rate and requires mandatory follow up for first few years after surgery.



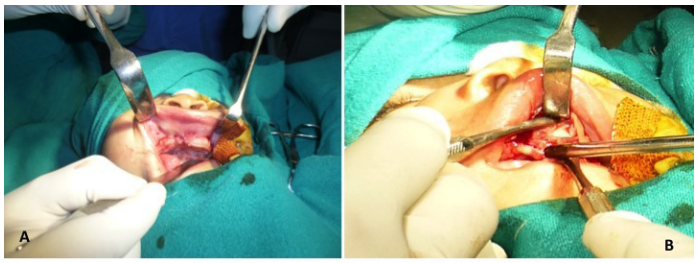
[Table/Fig-2]: 3D CT image reconstructed using Volume Rendered Technique showing osseous erosion involving maxillary alveolus of right side (arrow).
[Table/Fig-3]: Contrast enhanced CT scan- coronal image shows soft tissue swelling overlying maxillary alveolus with presence of bone erosion (arrow).
[Table/Fig-4]: Contrast enhanced CT scan- axial image shows heterogenously enhancing soft tissue mass lesion in the region of maxillary alveolus (arrow).

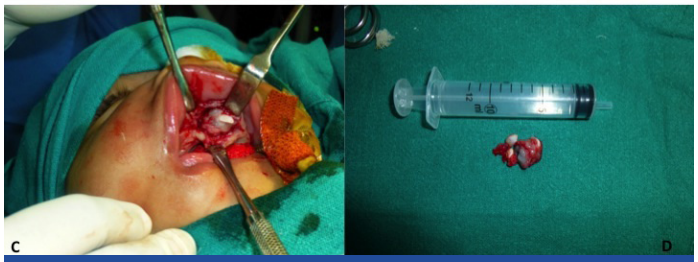




[Table/Fig-5]: Surgical excision of tumor mass. **[Table/Fig-6]:** Histopathological picture (Magnification 40X) showing biphasic pattern with small neuroblastic cells with minimum cytoplasm (thin arrow), large epithelioid melanin containing cells (thick arrow) and melanin pigment (thickest arrow). **[Table/Fig-7]:** Histopathological picture (Magnification 400X) showing biphasic pattern with small neuroblastic cells with minimum cytoplasm (thin arrow) and large epithelioid melanin containing cells (thick arrow).



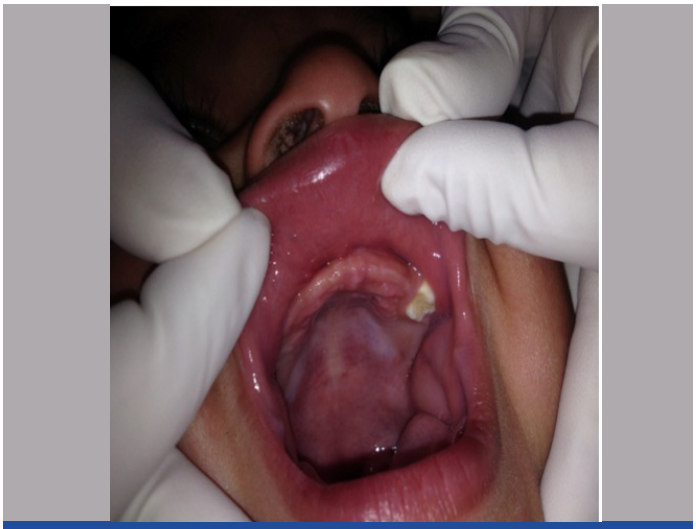
[Table/Fig-8]: Recurrence of tumor.





[Table/Fig-9]: Step by step excision of tumor mass during second surgery.

Differential diagnosis includes other round cells tumors of infancy like rhabdomyosarcoma, neuroblastoma, Langerhans histiocytoma and Ewing's sarcoma. MNTI does not express diffuse reactivity with S-100 protein unlike these round cell tumors, lacks other markers of neuroendocrine differentiation, myoglobin, myogenin, and muscle specific actin reactivity.



[Table/Fig-10]: Two years post operative picture showing no further tumor recurrence.

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