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# Melanotic Neuroectodermal Tumour of Infancy- A Short Communication

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**Abstract**

Melanotic Neuroectodermal tumour of infancy is rare occurring in infants less than one year old. Early diagnosis with prompt treatment is desirable as it can take malignant transformation. Surgical excision being the treatment of choice we hereby report a case of a 4 month old infant who reported to us where after excising the tumour amniotic membrane was used for closure. Follow-up revealed no morbidity.

**Keywords**: Amniotic membrane, Infancy, Malignant, Rare

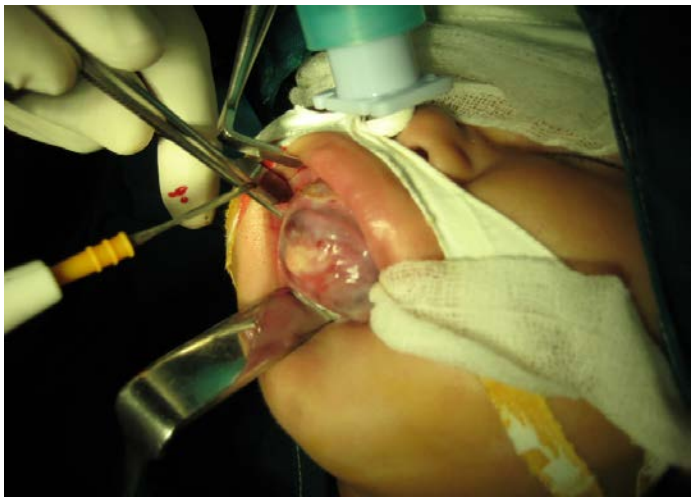
## Introduction

Melanotic neuroectodermal tumour of infancy (MNTI) is a very rare benign neoplasm that occurs in early infancy, first described by Krompecher in 1918. In 1966, Borello and Gorlin suggested that the tumour originated from neural crest cells as some of these tumours excreted large amount of vanillylmandelic acid (VMA) that is assosciated with other neuroectodermal tumours as well.[1] Tumour contains melanin and primarly affects the maxilla of infants during first year of life. It presents clinically as rapidly growing, painless expansile, partly pigmented mass. Although overall recurrence rate after surgery does not exceed to 15-20%, it has been reported to be as high as 60% after incomplete resection.[2]

The purpose of this present paper was to present a case report of such a tumour in 4 month old child and the management of the same.

## Case Report Study

A 4 month old child presented with a mass in the maxilla that had been growing since 1.5 months. Initially it was a small swelling in the anterior maxillary alveolar ridge since then it was growing rapidly to the present size (3×3 cm) that it almost obstructed the oral cavity and caused difficulty in feeding.

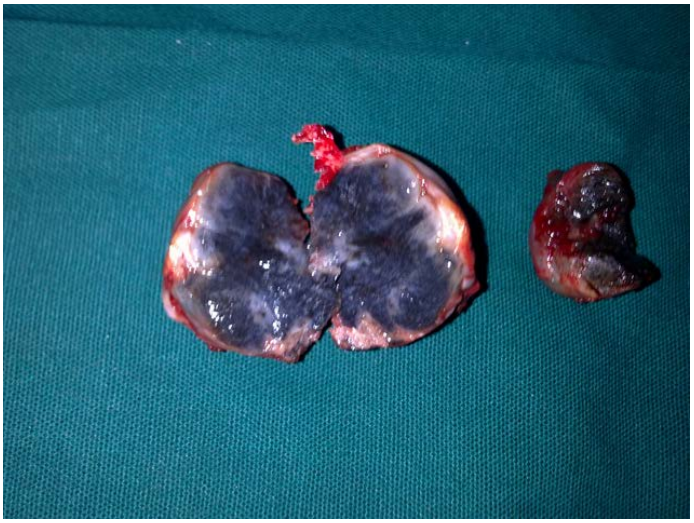


On physical examination loss of external facial symmetry was noted. Almost rectangular sessile growth was presented on the anterior maxillary alveolar ridge and hard palate region crossing the midline. Rapidly

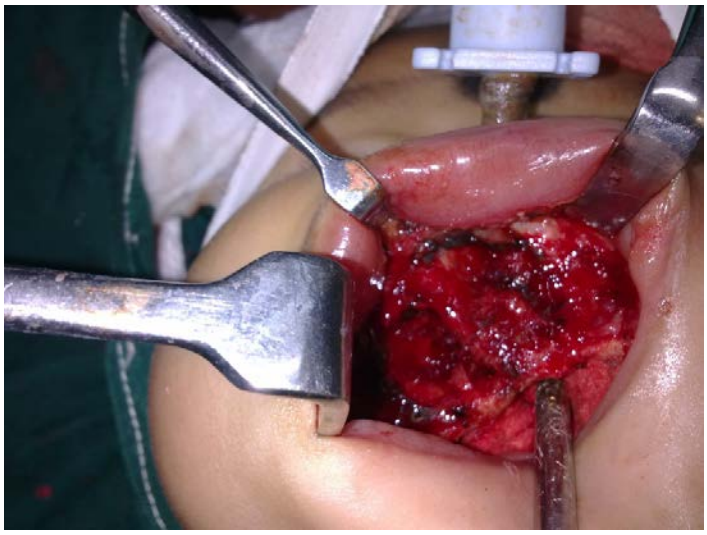
increasing size of the lesion caused risk of airway patency and aspiration.

Contrast enhanced CT revealed an osteloytic lesion (3×4 cm) present in the anterior premaxillary region.

Specimen obtained during biopsy revealed macroscopic bluish coloured gelatinous deposits. The cells were positive for histochemical markers like HMB 45, synaptophysin. Thus definitive diagnosis of MNTI was made.



Patient was taken under general anaesthesia. Incision was made in the oral mucosa along the boundaries of the tumour while maintaining a sufficient margin. Mucosa was reflected along the margin to expose healthy bone margins and the tumour was resected. Revised soft and hard tissue margins were send for frozen section and result was negative. No oroantral or oronasal communication was found to be present. Defect was large so decided to allow healing by secondary granulation. Raw surface was covered with amniotic membrane by suturing it to the surrounding soft tissue margins. Post operative recovery was uneventful. At 6 months follow up patient showed satisfactory healing and no signs of recurrence.



**Discussion**

Halpert and Patzer in describing this type of lesion was the first to introduce the term retinal analge tumour, stating that the folded, pigmented, cuboidal epithelium resembled the ciliary process of the eye and the sheets of small hyperchromatic cells mimicked the nuclear layers of retina. A number of investigators have challenged this theory, stating that retina is well organized before the maxilla begins to form in embryo. And also, no tissue of sclera or choroid has been identified and the eyes of these patients are normal.[4]

Two theories concerning the histogenesis of this tumour are the odontogenic theory put forward by Mummery and Pittis based on the site of the tumours in proximity of the developing teeth and tooth structures and with their apparent involvement in the tumourthe second is neuroectodermal theory.[5]

Today the most commonly accepted theory is one that puts forward its neuroectodermal origin, being based on 5 parameters: the tumour cells are similar to the neuroblast; microscopically it includes the presence of melanin, by electron microscopy we could distinguish neurosecretory granules; there might be an increase in VMA in urine and the presence of a high content of tyrosine hydroxylase in these cells.[6]

**Conclusion**

MNTI is usually a benign tumour arising from cells of Neuroectodermal origin. Due to its rapid growth potential and locally destructive behaviour early diagnosis and resection is required to reduce the morbidity and increase the prognosis.

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