

MELANOTIC NEUROECTODERMAL TUMOR OF INFANCY: CASE REPORT

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الورم القتاميني المتولد من الوديعة الخارجية العصبية في الرضع هو ورم سليم ونادر الحدوث وغالباً ما يصيب الفك العلوي .
وهذا تقرير عن حالة طفلة عمرها أربعة أشهر مصابة بهذا الورم في الفك العلوي الأيمن وقد تم استئصاله بنجاح وبدون أي إشارة لتكرار ظهوره وذلك خلال متابعة الأشهر الستة التالية . ومن ثم نوصي بالاستئصال الكامل لهذا الورم مبكراً . وذلك للحد من تلف الأنسجة المحيطة نتيجة تضخم الورم مع الحرص في المتابعة بعد ذلك .

Melanotic neuroectodermal tumor of infancy is a rare benign tumor that commonly affects the maxilla. This case is a report on a 4-month-old female infant with melanotic neuroectodermal tumor of the right maxilla. The tumor was successfully excised with no evidence of recurrence at 12 months follow-up. Early wide excision of the tumor to minimize tissue destruction by tumor growth and careful follow-up is recommended.

Introduction

Melanotic neuroectodermal tumor of infancy is considered as a rare tumor. Almost 95% of this tumor is found in infants less than one year of age and is equally distributed in both sexes. The most common site is the maxilla comprising 68.8% with the incisal area being the most frequently involved area.¹ Nevertheless, cases in different sites have been reported. The lesion is usually considered benign. Malignancy rate is approximately 1.9 - 3.2%.² Conservative excision is usually adopted but 10 - 15% of this tumor recur locally and may lead to death if it invades local vital tissues.²

Melanotic neuroectodermal tumor (MNT) was first described by Krompecher in 1981. Since that time, 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.

Case Report:

In this report, a 4-month-old Saudi female infant was referred to the Department of Pediatric Surgery, King Khalid University Hospital from a district hospital with a history of swelling in the right upper gum and maxillary region of two months duration but no associated symptoms.

The infant was a product of normal pregnancy and delivery. On examination, the child was healthy-looking and vital signs were stable with no lymphadenopathy. Chest, heart, and abdominal examinations were normal. However, examination of the lesion revealed a swelling in the anterior region of the right upper gum. The lesion was 3x4 cm in diameter, hard in consistency and obliterating the canine fossa on the right side. It was deviating and blocking the right nostril. There was no invasion of the soft and hard palate [Fig. 1].

Hemogram, serum electrolytes, renal and liver profiles were normal. Urinary Vanillylmandelic Acid (VMA) was normal. CT scan of the skull and face revealed a tumor invading the bone of the right maxilla and deviating the nasal septum [Fig. 2].

The child had a biopsy taken from the tumor site which was reported to be a pigmented neuroectodermal tumor of infancy. The child has under-

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gone surgical excision of the tumor. The tumor was enucleated and removed with the bone of the right premaxilla as well as the deciduous teeth. The area extending from the canine fossa to the infra-orbital margin was explored. The bone was shaved and smoothed down with an osteotome and the surgical specimen was sent to the pathology laboratory. The histopathology result confirmed a benign neuroectodermal tumor of infancy [Fig. 3a], The bone proved to be reactive calcification initiated by the presence of the tumor in the area^{3,4} [Fig. 3b]. Postoperative course was uneventful. A regular follow-up examination for 12 months revealed no evidence of recurrent tumor and the child appeared healthy [Fig. 4].



Fig. 1. Preoperative view showing swelling of the maxilla on the right side.

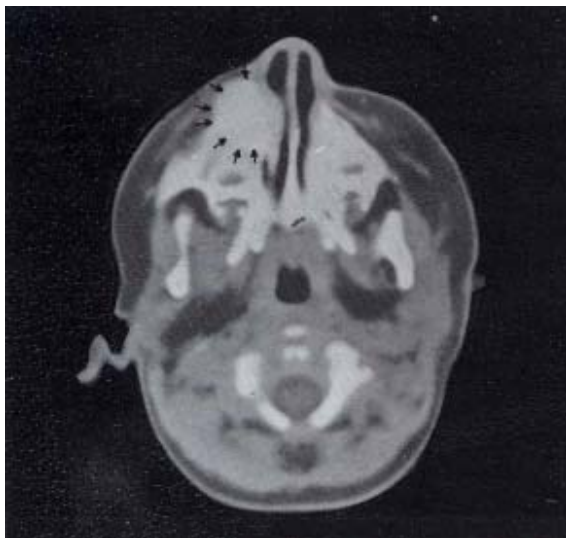


Fig. 2. CT scan of the skull and face showing a tumor invading the bone of the right maxilla and deviating the nasal septum.

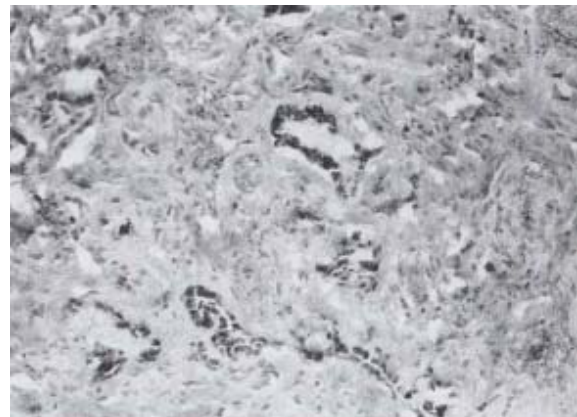


Fig. 3a. Histopathological section showing neoplastic epithelial-like cells in nests or cords growing freely within dense connective tissue stroma. Islands of non-pigmented epithelial-like cells reminiscent of odontogenic rest cells are also evident. (H&E x 250).

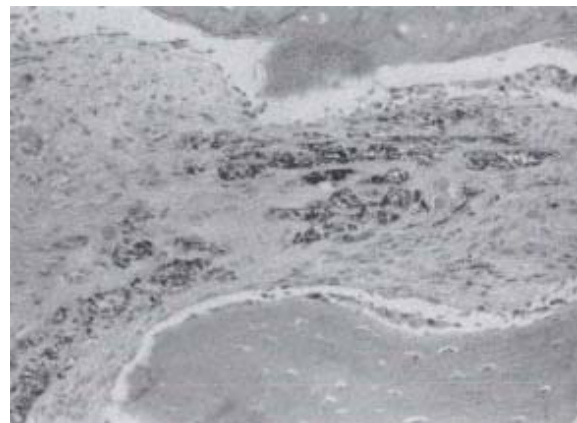


Fig. 3b. Histological section showing a reactive calcification initiated by the presence of the tumor. (H&E x 250).



Fig. 4. The patient 12 months after surgery, showing no evidence of recurrence.

Discussion

Cutler² published an extensive review of the literature since the first case report of MNT by Krompecher³ in 1918. The review showed that MNT is a tumor of childhood and the majority of patients were below the age of one year. Nevertheless, adult cases have been reported.² The majority of cases were benign in nature and commonly located in the maxilla. However, other locations have been reported. The lesion is a fast growing swelling of the incisor region of the maxilla with brown pigmentation commonly seen on the overlying soft tissues. The origin of this tumor is not well known. However, ultra structural studies,^{2,4} histochemical studies,⁵ and elevated VMA⁶ suggested neural crest origin.

Our patient was below six months of age, female, and had right maxillary tumor which proved to be benign in nature clinically and histopathologically and similar to others reported. Wide surgical excision with removal of some of the tooth buds to avoid local recurrence was performed in our patient as has been reported in the literature.⁴ Some authors advocated conservative excision and attributed the success of this procedure to the debunking effect and initiation of bodily defense that resulted in destruction of any residual cell of the tumor left behind.^{7,9} However, the growth rate of the recurrent lesion mass may change from what is initially a simple localized tumor into a more invasive one that causes extensive destruction to healthy tissues. The use of CT scan has been helpful in assessing the extent of the disease and insured a careful excision without interfering with

important structures in the area like the infra-orbital nerve, the orbit, or nasal floor.

This case report stresses the fact that wide local excision is needed to avoid local recurrence which is associated with such tumor. Careful follow up is required in such patients to detect any early recurrence.

References

1. Johnson RE, Scheithauer BW, Dablin DC. Melanotic neuroectodermal tumor of infancy: a review of seven cases. *Cancer* 1983;52(4):661-6.
2. Cutler LS, Chaudhry AP, Topazian R. Melanotic neuroectodermal tumor of infancy: an ultrastructural study, literature review, and re-evaluation. *Cancer* 1981 ;48(2):257-70.
3. Krompecher E. Zur Histogenese und morphologie adamantinome und sonstiger kiefergeschwulste. *Beitr Pathol Anat* 1981 ;64 165-97.
4. Nikai H, Ijuhim N, Yamasaki A, Niitani K, Imai K. Ultra structural evidence of neural crest origin of melanotic neuroectodermal tumor of infancy, *J Oral Pathol* 1977;6:221-232.
5. Koudstaal J, Oldhoof J, Panders AK, Mardonk MJ. Melanotic neuroectodermal tumor. *Cancer* 1968;22:151-61.
6. Borello ED, Gorlin RJ. Melanotic neuroectodermal tumor of infancy - A neoplasm of neural crest origin. Report of a case associated with high urinary excretion of vanillylmandelic acid. *Cancer* 1966; 19:196-206.
7. Claros P, Claros A Sr, Claros A Jr, Claveria A. Melanotic neuroectodermal tumor of infancy: a case report. *Int J Pediatr Otorhinolaryngol* 1989;17(1):65-73.
8. Hupp JR, Topazian RG, Krutchkoff DJ. The melanotic neuroectodermal tumor of infancy. Report of two cases and review of the literature. *Int J Oral Surg* 1981; 10(6):432-46.
9. Hellstrom KE, Hellstrom I. Immunity to neuroblastomas and melanomas. *Annu Rev Med* 1972;23:19-38.