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內文：

**Introduction**

Melanotic neuroectodermal tumor of infancy (MNTI) is a rare tumor of neural crest origin. It was first described in 1918 by Krompecher and 356 patients have been reported in the English language literature to date.

- Incident Age: 1 year old.
- Incident Location: Maxillary alveolus
- Clinical Presentation: firm, painless, rapidly growing mass, may elevate the upper lip and can interfere with nursing.
- Treatment: Surgery.
- Local recurrence rate: 10% to 15%.
- Incidence of metastasis: 3%.

This article reports experience with the management of this tumor in 18 patients. This is the largest number of patients with this tumor from a single institution.

**Patients and Methods**

A retrospective analysis based on medical records was performed for patients with MNTI admitted to the Department of Pediatric Surgery, King George Medical University, between October 1984 and January 2004.

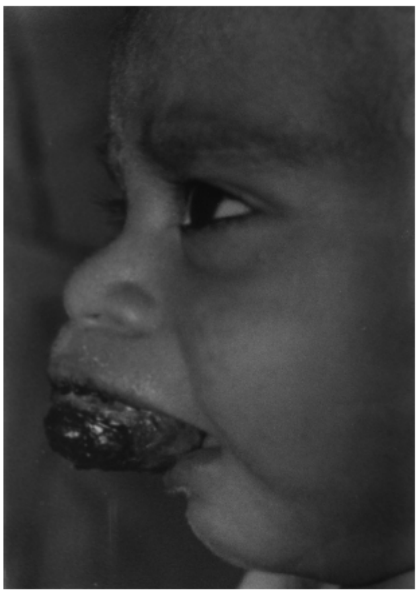

**FIGURE 1.** Photograph showing clinical appearance of melanotic neuroectodermal tumor of infancy.

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The mean age was 4.3 (±2.1) months. All the babies presented with a painless slowly growing swelling in the maxillary alveolus. There was no family history of similar tumors in any of our patients.

On examination, in all patients the swelling was smooth, hard, bluish black in color, and fixed to the bone. The overlying mucosa was thin and slightly mobile; none of the patients showed ulceration of the mucosa.

**Table 1. EIGHTEEN PATIENTS WITH FOLLOW-UP AND OUTCOME**

| Case | Age (mo) | Gender | Site | Follow-Up (mo) | Outcome |
|---|---|---|---|---|---|
| 1 | 8 | Male | Rt max alveolus | 198 | Alive and well |
| 2 | 2 | Male | Lt max alveolus | 152 | Alive and well |
| 3 | 3 | Female | Lt max alveolus | 202 | Alive and well |
| 4 | 6 | Female | Ant max alveolus | 206 | Alive and well |
| 5 | 5 | Female | Lt max alveolus | 102 | Alive and well |
| 6 | 2 | Male | Lt max alveolus and palate | 98 | Expired |
| 7 | 4 | Male | Lt max alveolus | 156 | Alive and well |
| 8 | 2 | Male | Rt max alveolus | 172 | Alive and well |
| 9 | 6 | Male | Rt max alveolus and palate | 164 | Alive and well |
| 10 | 8 | Male | Rt max alveolus | 152 | Alive and well |
| 11 | 2 | Male | Lt max alveolus | 134 | Alive and well |
| 12 | 3 | Female | Ant max alveolus | 94 | Alive and well |
| 13 | 6 | Female | Rt max alveolus | 74 | Alive and well |
| 14 | 5 | Female | Lt max alveolus | 188 | Alive and well |
| 15 | 2 | Male | Rt max alveolus and palate | 90 | Alive and well |
| 16 | 4 | Male | Lt max alveolus | 88 | Lost to follow-up |
| 17 | 2 | Male | Rt max alveolus | 72 | Alive and well |
| 18 | 6 | Male | Lt max alveolus | 12 | Alive and well |

Abbreviations: Ant, anterior; Lt, left; Rt, right; Max, maxillary.

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The swelling was present on:

- Lateral maxillary alveolus: 13 patients (left: 8; right: 5)
- Midline maxillary alveolus: 2 patients
- Maxillary alveolus and hard palate: 3 patients

Investigations included hematocrit, coagulation profile, and serum creatinine. Urinary vanillyl mandelic acid expression was positive in 6 of the patients. Hematological screening for catecholamines was within normal limits in all.

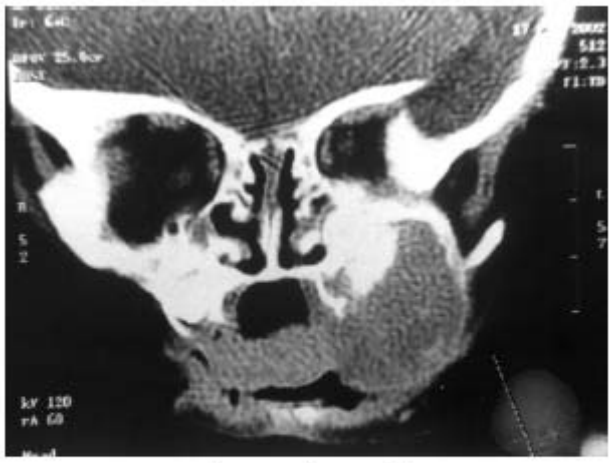

**FIGURE 2.** Computed tomography scan of the patient in Figure 1, showing radiolucent lesion expanding the maxillary alveolus.

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X-ray of the maxilla in all patients showed expansion of the bone by a radiolucent lesion.

Computed tomography scans were performed for 14 patients admitted after 1990 and uniformly revealed a central area of radiolucency with sharp margins displacing the surrounding bone and tooth buds.

Disease-free and overall survival was calculated from the time of diagnosis. Statistical analysis for overall survival was performed using the Kaplan-Meier method with SPSS statistical software. Mean follow-up time was 130.8 months

**SURGERY AND PATHOLOGIC DETAILS**

The average age at surgery was 9 months. Surgery was performed under endotracheal anesthesia by an intraoral approach. A mucoperiosteal incision was made and the mucosal flap elevated to remove the tumor by enucleation; the tumor was well-circumscribed and could be easily enucleated out of the alveolar crest; however, the related primary teeth and tooth buds had to be sacrificed. The cavity was then

curetted to remove all traces of blue black tissue, after which primary closure could be achieved in only 6 patients.
In the remaining patients a local mucoperiosteal flap and costocondral graft was used to cover the defect. The costocondral graft was harvested by removing the costal cartilage of the right sixth rib subperiostially. The cartilage was then trimmed to bridge the defect in the maxilla and sutured in place with absorbable suture to the mucoperiosteum. A mucosal flap taken from the cheek or inside of the upper lip was then used to cover the cartilage graft putting the epithelial surface outward.

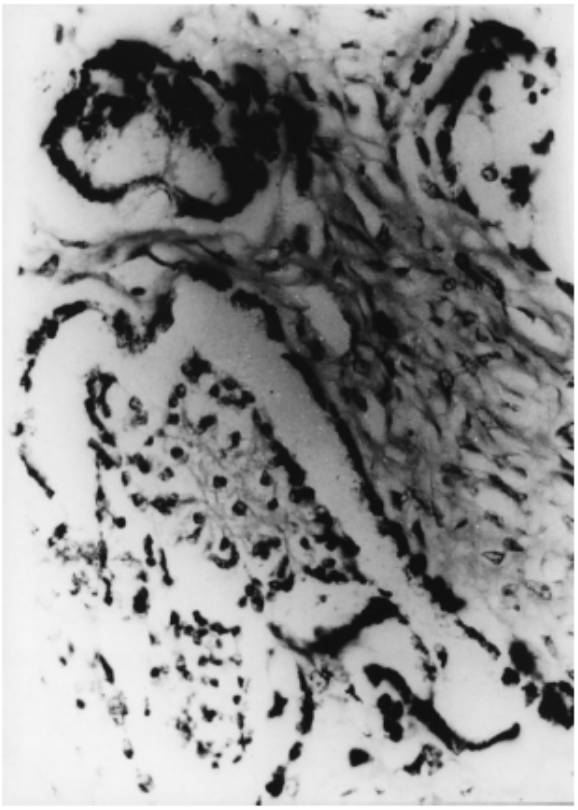

**FIGURE 3.** Histology of the excised tumor showing biphasic cell population.

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On gross examination the excised specimens were bluish black or brown in color and well encapsulated. The histological features showed cells arranged in clusters or lines with cleft-like spaces within a fibrous stroma. A biphasic cell population consisting of large epitheloid cells with cytoplasm containing melanin pigment and small ovoid, undifferentiated cells closely resembling neuroblasts were seen.

**IMMUNOHISTOCHEMISTRY**

Immunohistochemistry showed, the tumor was positive for neuronspecific enolase, synaptophysin, S-100.

**Results**

Before discharge all patients were referred for pedodontic consultation. None required any dental prosthesis.
We found clear-cut male predilection of the disease (12 males: 6 females) and all the patients who expressed vanillyl mandelic acid (VMA) in their urine were males.
All the patients are in regular follow-up except 2 and there is no any local recurrence or distant metastasis. One patient was lost to follow-up at 88 months without disease while 1 patient expired outside the hospital (cause, not known). Overall survival at 17 years was 86.7%, while mean survival time was 189.7 months.

**Discussion**

90% of the Melanotic Neuroectodermal Tumors are seen in the head and neck region,

- Maxilla :68.8%

- Skull: 10.8%
- Mandible: 5.8%
- Brain: 4.3%

All of our patients had involvement of the maxillary alveolus. No definite gender predilection exists in the literature, though 12 out of our 18 patients were male.
The etiology of MNTI is unknown. The tumor is unique in its ability to synthesize melanin and may also elaborate VMA in urine. Six of our patients, all male, tested positive for VMA in urine. The expression of VMA is characteristic of tumors arising from neural crest cells but we do not know why VMA expression was limited to male patients in this study.
All 18 patients except 2 were otherwise healthy and well except for the symptoms due to physical effect of the swelling and cosmetic disfigurement. A thorough local excision and curettage of the cavity to remove all traces of bluish black tissue sufficed for cure in all 18 patients. Although the majority of tumors are benign, a local recurrence rate of 10% to 15% and a malignancy rate of 3.2% are reported in the literature.
Histopathology and immunohistochemistry was diagnostic of MNTI. The diagnosis of MNTI is mainly clinical. The physical findings are classical and the radiological features are characteristic. Immunohistochemistry clarifies the neural crest as the cell of origin of MNTI and characterizes the tumor but no prognostic benefit is available from this investigation at present.
We conclude that early conservative surgical excision provides an excellent result with good prognosis for patients with melanotic neuroectodermal tumor of infancy.
Questions

| 題號 | 題目 |
|---|---|
| 1 | What is the origin of melanotic neuroectodermal tumor?<br>(A) Melanocytic origin<br>(B) Epithelial origin<br>(C) Neural crest origin<br>(D) Fibrous cell origin |
| 答案<br>(C) | 出處：*Melanotic Neuroectodermal Tumor of Infancy: 2 Decades of Clinical Experience With 18 Patients*<br>*J Oral Maxillofac Surg 67:47-51, 2009* |
| 題號 | 題目 |
| 2 | Which is not a common clinical presentation of melanotic neuroectodermal tumor?<br>(A) Bluish-black color<br>(B) Ulceration<br>(C) Hard<br>(D) Smooth surface |
| 答案<br>(B) | 出處：*Melanotic Neuroectodermal Tumor of Infancy: 2 Decades of Clinical Experience With 18 Patients*<br>*J Oral Maxillofac Surg 67:47-51, 2009* |