The Turkish Journal of Pediatrics
Volume 24, No. 3, July - September 1982

MELANOTIC PROGONOMA OF THE MAXILLA*

(Report of a successfully treated case and a brief review of the literature)

*Sefa Kaya MD***, *Behçet Tınaztepe MD****

Key words: maxilla, melanotic ameloblastoma, melanotic neuroectodermal tumor, melanotic progonoma, retinal anlage tumor

Melanotic progonoma is a rare tumor, which characteristically has been found to occur in the jaws of infants. These tumors have also been reported to have been found elsewhere: in the shoulder epididymis, mediastinum, skull and brain[1-8].

These lesions have been designated melanotic ameloblastoma, melanotic neuroectodermal progonoma, retinal anlage tumor, melanotic ameloblastic odontoma, melanotic hamartoma[9,10].

The purpose of this paper is to present a case of successfully treated melanotic progonoma of the maxilla, and give a brief review of the literature.

Case Report

A 2.5-month-old female infant was admitted to the Hacettepe Children's Hospital in December 1975 because of a mass in the mouth and subsequent feeding difficulties. The mother had first noticed the mass as a small, firm, red colored lump in the intraoral region of the left upper jaw when the baby was twenty days old. The mass appeared to be growing rapidly and sucking and feeding were gradually impaired.

On physical examination, an obvious bulge in the left upper cheek was seen, but the overlying skin was intact. Intraorally, a firm, egg-sized mass in the region of the left upper jaw and the hard palate extending to the retromolar region was noted (Figure 1). There was ulceration of the mucosa but no drainage from the lesion.

Radiographic studies revealed a large tumor with destruction of the left maxillary and zygomatic bones. The results of the physical examination and laboratory studies were essentially unremarkable except for the tumor. Biopsy revealed a melanotic progonoma.

---

* From the Hacettepe University Faculty of Medicine, Departments of Otolaryngology and Pathology, Ankara.

** Professor of Otolaryngology, Hacettepe University Faculty of Medicine.

*** Professor of Pathology, Hacettepe University Faculty of Medicine.

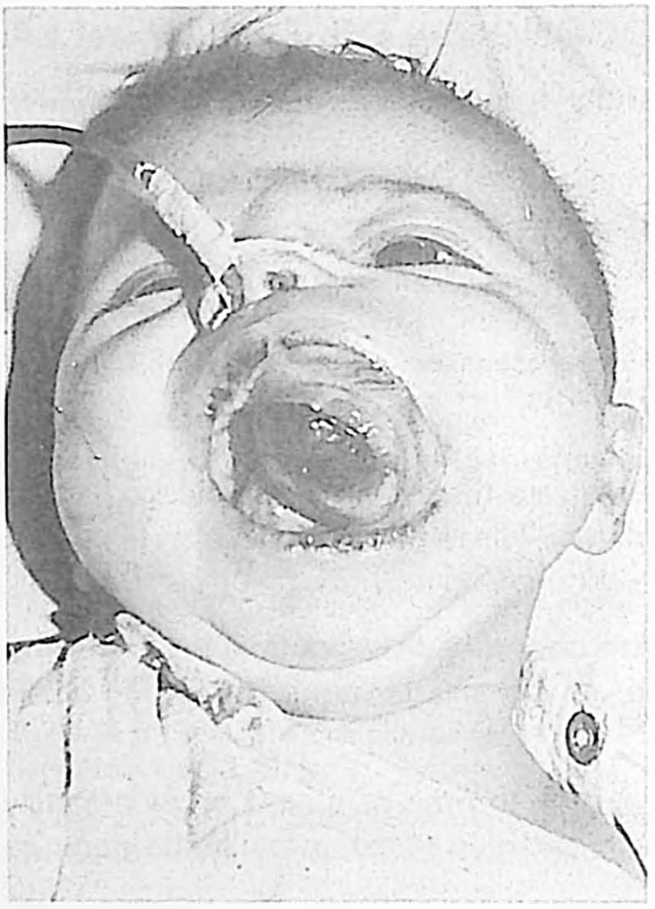

Figure 1.

Preoperative appearance of the patient.

The results of a total daily urine analysis for vanillylmandelic acid (VMA) were within normal limits.

The infant underwent surgery on February 2, 1976. Using a Ferguson incision, the left cheek was elevated and the tumor exposed. After exposure, the tumor was found to be extended to the ethmoidal region, the orbital floor, and the underside of the zygomatic arch. Complete removal of the tumor by left total maxillary resection was performed. The periosteum of the orbital floor was found to be intact and was not resected. In order to reconstruct the orbital floor, the inferior concha was elevated into a pedicle flap and sutured to the periosteum. The surgical cavity was covered with a split-thickness skin graft.

The postoperative course was uneventful and the patient was discharged in good health on March 18, 1976 (Figure 2).

In a follow-up control examination, 5 years after the operation, there was no sign of recurrence of the tumor.

Pathological examination. The specimen was fixed in formalin: it consisted of various sized pieces of firm tissue, grey-white with patchy areas of black, measuring

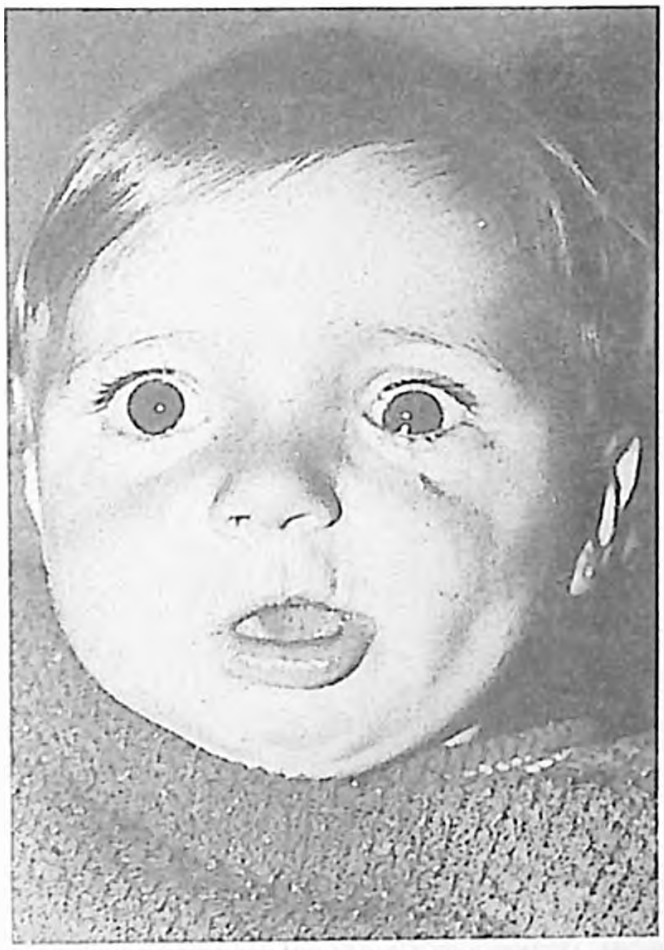

Figure 2.

Postoperative appearance of the patient.

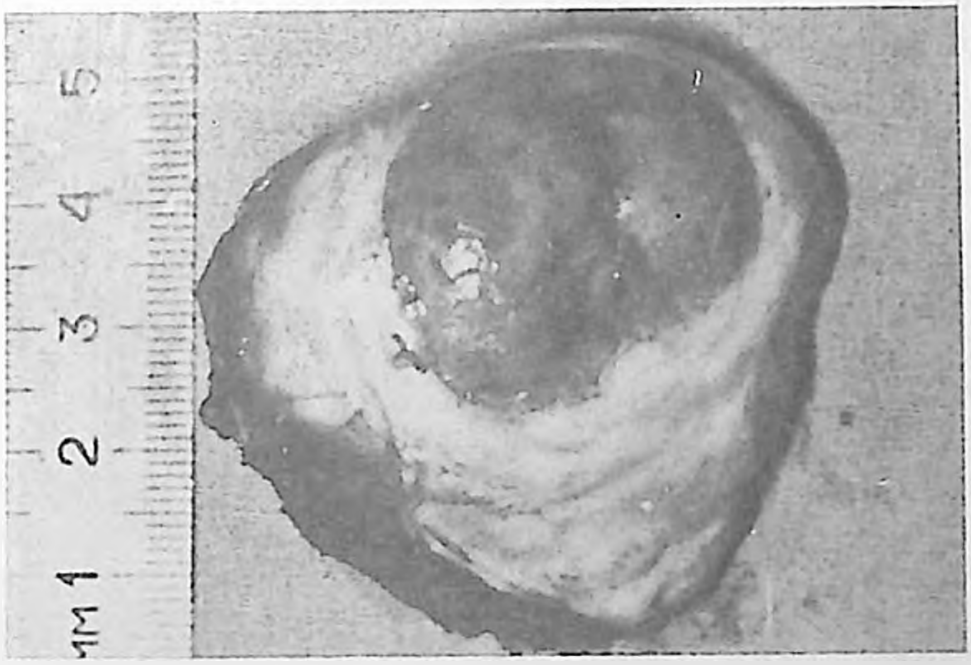

Figure 3.

Total resection of maxillary tumor.

in aggregate 10×7×4 cm. The largest piece was 5.5×5×3.5 cm firm, roughly semiglobular with a base of hard bone (Figure 3). The cut surfaces were grey-white, light brown, and black.

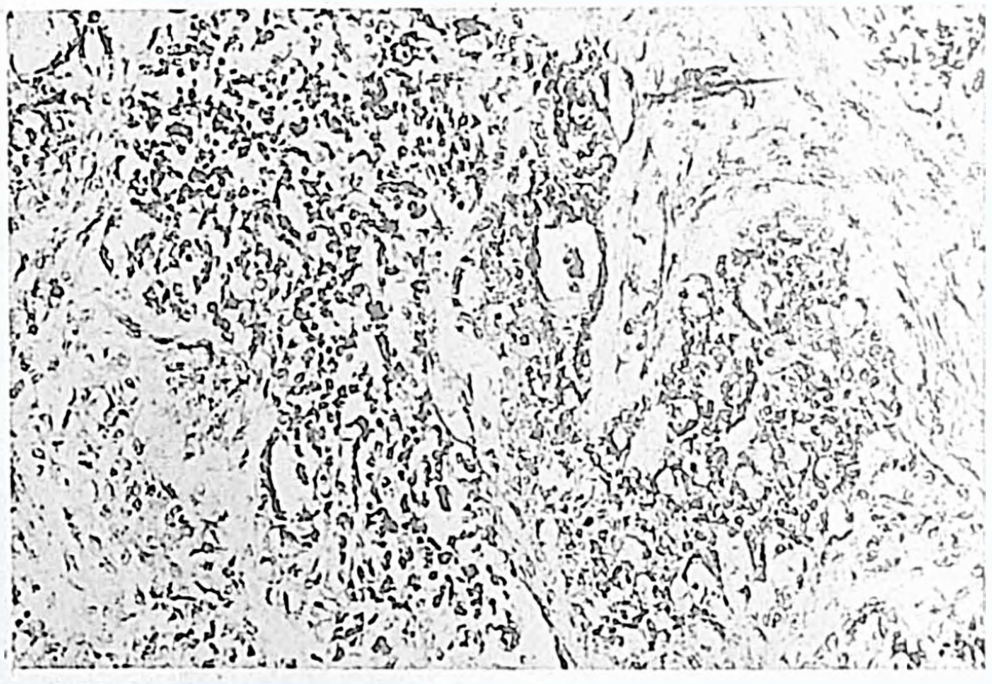

Figure 4.

Tubular arrangement of tumor cells containing abundant melanin pigment (H + E × 400).

Microscopic examination. Sections of the tumor showed epithelial-like cells arranged in tubules containing abundant brown pigment (Figure 4). The pigment was bleached with acid potassium permanganate; iron and PAS reactions were negative. The pigment was more readily seen with acetic acid-acidified toluidine blue (pH 3.5) than with the H + E stain. The second type of cells were aggregated into groups of various sizes and shapes, not unlike neuroblasts (Figure 5). There were also cells

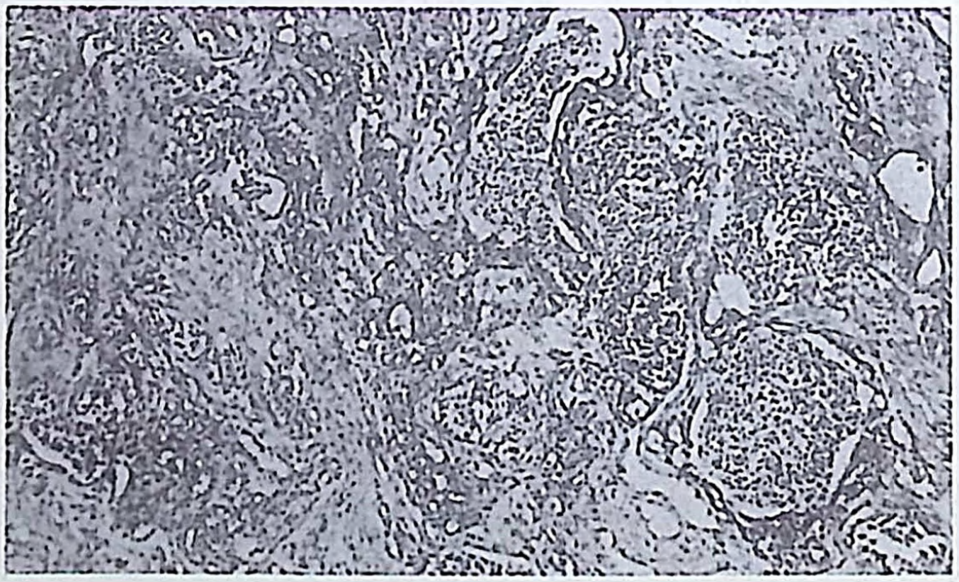

Figure 5.

Neuroblast-like cells arranged in groups (H + E × 150).

suggestive of a transition of both of these types of cells. Fibrocollagenous stroma varied in amount in different areas. A section of decalcified bone was also found to be infiltrated by the tumor (Figure 6).

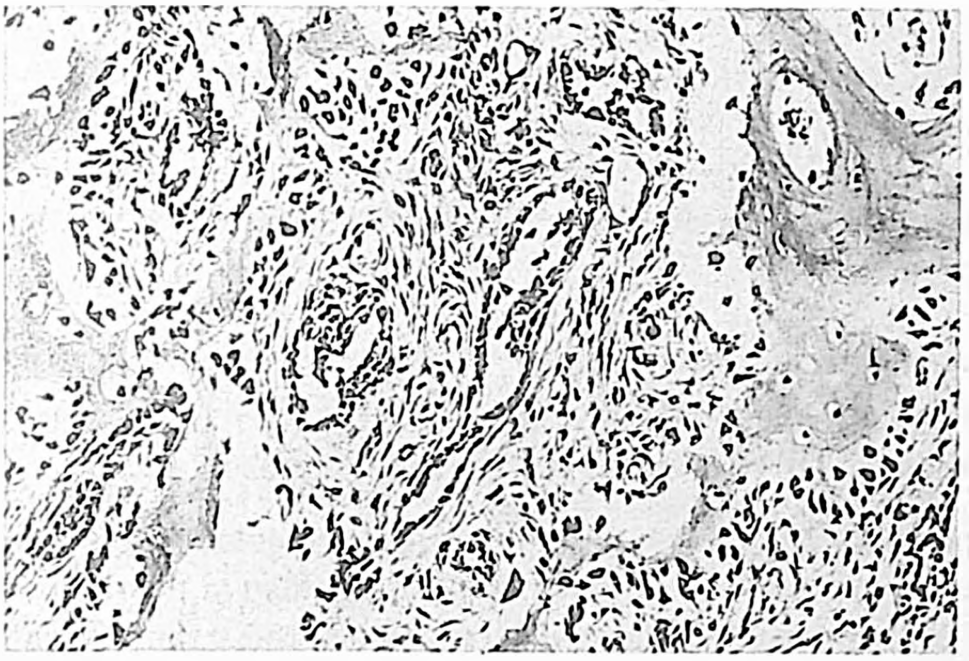

Figure 6.

Infiltration of bone by tumor (H + E × 400).

Discussion

Melanotic progonoma may be found at any age from stillborn to old age[6]. Five of the 158 cases reported in the English language review literature were malignant, producing a malignancy rate of 3.2%. The most common sites of origin were the maxilla (68.8%), the skull (10.8%), the mandible (5.8%) and the brain (4.3%) [11]. An additional two cases appeared in the Turkish literature, both of which were treated successfully [12].

According to Best[7], the first case to appear in the literature, reported by Krompecher in 1918, was regarded as a congenital melanocarcinoma.

Opinion is still divided regarding the histogenesis of this tumor. The main theories are that it arises from paradental epithelium[7], an atavism of sensory neuroectodermal [10] and neurogenic origin[4], like retinal anlage[13] and one-sided teratoma[5].

Borello and Gorlin[14] recorded a case in which an excessive urinary excretion of vanillylmandelic acid returned to a normal level after removal of the maxillary tumor. These authors explain the absence of demonstrable neural elements by an analogy to neuroblastoma in which neurofibrils are not always found. Similarly we could not demonstrate neurofibrils in the case presented as we did not find high VMA in the urine to support the theory of neurogenic origin.

The clinical presentation and the rapid growth of the tumor were suggestive of malignancy. Although this tumor has a distinct histologic appearance, in cases

where only a small gingival biopsy is performed, the sections show only poorly pigmented, immature, neuroblast-like cells. In these cases difficulties of diagnosis may arise.

The biologic behavior of the tumor in infants is usually benign. Therapy is local excision. If not totally resected, the tumor tends to recur. The recurrence rate is about 15%.

In the case presented, although the tumor was extensive and required total maxillary resection, follow-up studies up to 5 years in duration did not reveal any recurrence of the growth.

Summary

In this article a 2.5-month-old female with melanotic progonoma of the left maxilla, who was treated by a total resection of the maxilla, is presented.

Melanotic progonomas are rare, benign melanin-pigmented tumors, which usually occur in the jaws of infants, and few cases are reported in the medical and dental literature. The importance of wide surgical excision of these tumors is stressed.

REFERENCES

1. Blanc WA, Rosenblatt P, Wolff JA. Melanotic progonoma (retinal anlage tumor) of shoulder in an infant, a case report. *Cancer* 11:959, 1958.
2. Lurie HI, Isaacson C. A melanotic progonoma in scapula region. *Cancer* 14:1088, 1961.
3. Frank GL, Koten JW. Melanotic hamartoma ("retinal anlage tumour") of the epididymis. *J Path Bact* 93:549, 1967.
4. Misugi K, Okajima H, Newton WA Jr, et al. Mediastinal origin of a melanotic progonoma or retinal anlage tumor: ultrastructural evidence for neural crest origin. *Cancer* 18:477, 1965.
5. D'Abrera VS, Burfitt-Williams W. A melanotic neuroectodermal neoplasm of the posterior mediastinum. *J Pathol* 111:165, 1973.
6. Schulz DM. A malignant melanotic neoplasm of the uterus resembling the retinal anlage tumours. *Am J Clin Pathol* 28:524, 1957.
7. Best PV. Pigmented tumour arising in the skull of an infant. *J Pathol* 107:69, 1972.
8. Stowens D, Lin TH. Melanotic progonoma of the brain. *Hum Pathol* 5:105, 1974.
9. Tiecke RW, Bernier JL. Melanotic ameloblastoma. *Oral Surg* 9:1197, 1956.
10. Stowens D. A pigmented tumor of infancy, the melanotic progonoma. *J Path Bact* 73:43, 1957.
11. Cutler LS, Chaudhry AP, Topazian R. Melanotic neuroectodermal tumor of infancy: an ultrastructural study, literature review, and reevaluation. *Cancer* 48:257, 1981.
12. Tel N, Tel E, Tınaztepe K, Tınaztepe B. Melanotik nöroektodermal tümör (melanotik progonoma). *Çocuk Sağlığı ve Hastalıkları Dergisi* 15:351, 1972.
13. Cholnoky T. Melanotic progonoma (retinoblastoma of maxilla). *Plast Reconstr Surg* 46:600, 1970.
14. 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* 19:196, 1966.