Case Report

Melanotic neuroectodermal tumour of infancy: Case report of an unusual tumor

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ABSTRACT

The melanotic neuroectodermal tumour of infancy is a rare, pigmented neoplasm of neural crest origin occurring in infants before 1 year of age. We report a 6-month-old male baby with a similar tumour involving superior maxillary alveolar ridge and most of the hard palate. A partial maxillectomy was performed by oral approach. One-year follow-up of the patient showed no recurrence.

KEY WORDS

Neuroectodermal, Melanotic, Tumour.

INTRODUCTION

The melanotic neuroectodermal tumour of infancy (MNTI) is a rare, pigmented neoplasm of neural crest origin occurring in infants before 1 year of age. It is a rapidly growing tumor that most frequently affects the craniofacial skeleton. The most common site of occurrence is the anterior maxillary alveolar ridge (70% cases) followed by the skull, brain and mandible. The genital organs are the most frequent extra cranial sites. It usually follows a benign course but inadequate excision, occasional multicentricity, and a small malignant potential result in a fairly high recurrence rate. Long-term follow-up is imperative. This tumour should be included in the differential diagnosis of head and neck neoplasms in infants and young children.

CASE REPORT

A 6-month-old male baby presented with a smooth, firm, painless, slow growing swelling involving the superior maxillary alveolar ridge and most of the hard palate. (Figure 1) A radiograph revealed a radiolucent lesion in the left anterior maxilla, and CT scan showed a solid hyper dense mass. (Figure 2) The pre-operative diagnosis of melanotic neuroectodermal tumour of infancy was made by fine needle aspiration cytology, showing a dual population of small neuroblastic cells.
and large melanin-containing epithelial cells. Urine catecholamine excretion was normal. A partial maxillectomy was performed by oral approach. A small split thickness skin graft was applied to the raw area and cavity was packed with liquid paraffin soaked roller gauge. After 2 weeks, a palatal prosthesis with maxillary extension incorporated into dental cap splint was made and applied. (Figure 3) Histological examination of the excised mass confirmed the initial diagnosis. Post-operative course was uneventful. One-year follow-up of the patient showed no recurrence.

DISCUSSION

Krompecher first described the tumor in 1918, since then only about 250 cases have been reported in the world literature. Difficulty in deciding the cellular origin of this tumour has led to numerous names, including congenital melanocarcinoma, melanotic epithelial odontoma, melanotic ameloblastoma, and retinal anlage tumour.

The pre-operative distinction of this tumour from other small round cell tumours of infancy (rhabdomyosarcoma, neuroblastoma, melanoma and lymphoma), is essential in order to plan the most complete resection and therefore reducing the possibilities of tumour recurrence. Though high levels of urinary excretion of vanilmandelic acid and serum alpha-fetoprotein are characteristic of this tumor their not always present. Histologic appearance is distinctive, with tubular or alveolar formations of large melanin-containing cells around nests of smaller neuroblastic cells possessing fibrillar cytoplasm. Immunohistochemical and ultrastructural studies reveal two types of cells: small, poorly differentiated cells that were positive for neuron-specific enolase protein and vimentin, and larger epithelial cells that were positive for melanoma antigen (HMB45) and frequently contain large and elongated melanosomes, similar to those described in retinal pigmented epithelium.

An aggressive surgical approach consisting of complete surgical excision is advocated when vital structures are not involved. It is a rapidly growing tumor with potential for local invasion; therefore, radical surgery is associated with a favorable outcome and offers the potential for long-term cure. Carnevale et al suggested the use of operating microscope for surgical excision to remove unseen remnants of the pigmented lesion. In continuity, reconstruction is recommended in newborn and infants. However, long-term follow-up is necessary for secondary corrective surgery at early skeletal maturity, if required. Currently, there is no effective adjuvant therapy for recurrent and residual tumour. Radiation is precluded by the patients’ young age, and chemotherapy trials have not demonstrated long-term efficacy.

REFERENCES

1. Kaya S, Unal OF, Sarac S, Gedikoglu G. Melanotic