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Melanotic neuro-ectodermal tumour of infancy arising from the squamous and occipital bone

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Abstract
The melanotic neuro-ectodermal tumour of infancy is an uncommon benign tumour of neural crest origin occurring in infants with a predilection for the anterior maxilla. This lesion shows a good response to conservative surgical excision with few recurrences but it should be followed up for long periods of time because of the rare malignant change possible in the tumour.

Key words: Ameloblastoma, pigmented; Neural crest

Introduction

The melanotic neuro-ectodermal tumour of infancy was first reported by Krompecher in 1918 as 'congenital melanocarcinoma' in a two-month-old infant. This tumour is also known as pigmented ameloblastoma, retinal anlage tumour, pigmented congenital epulis *etc*. As the names suggest there have been various theories of origin that it was odontogenic or of neural crest and germ cell origin. In 1966, Borello and Gorlin reported increased levels of 3-methoxy-4-hydroxymandelic acid (VMA) in a three-month-old child with this tumour, suggesting a neural crest origin. They gave the name 'melanotic neuro-ectodermal tumour of infancy'. Ultrastructural studies have demonstrated the presence of both melanotic and neurogenic cells.

Case report

A three-month-old male child was brought to the ENT clinic at the All India Institute of Medical Sciences with a history of a right-sided swelling behind the ear since birth which was rapidly increasing in size. Apart from this swelling the child was well preserved. General and systemic physical examinations of the patient were normal. Examination of the head and neck region revealed a firm mass (6 × 6 cm) in the right postauricular region which had well-defined margins and was fixed to the occipital bone. The overlying skin was free and non-ulcerated but the local temperature was raised and there were dilated veins over the tumour. The mass was not tender. There were no cranial nerve palsies or signs of increased intracranial tension. There were no palpable nodes in the neck. The remainder of the ear, nose and throat examination was within normal limits.

The patient underwent routine clinical laboratory studies which were within normal limits: a test for the VMA level of the urine was not carried out. The patient was subjected to a fine needle aspiration test which was inconclusive. A CT scan was done under sedation which showed a well-defined lesion of high density causing erosion of the outer table of the occipital bone in

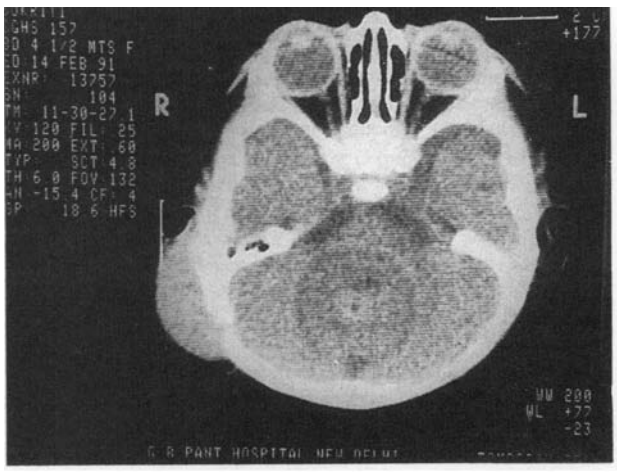

FIG. 1
Axial CT scan showing tumour arising from the squamous and occipital bone.

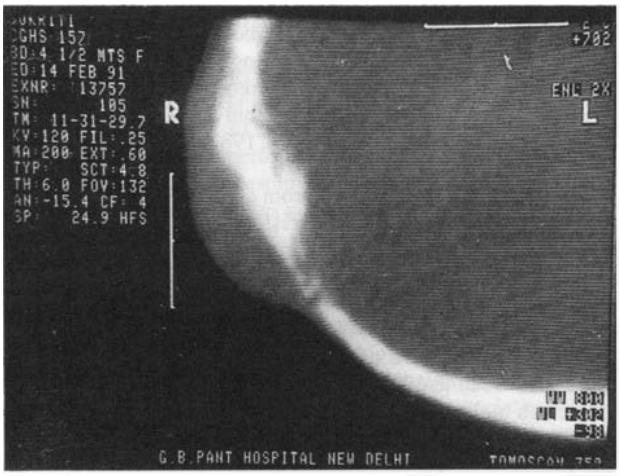

FIG. 2
Axial CT scan showing destruction of the outer table of the occipital bone.

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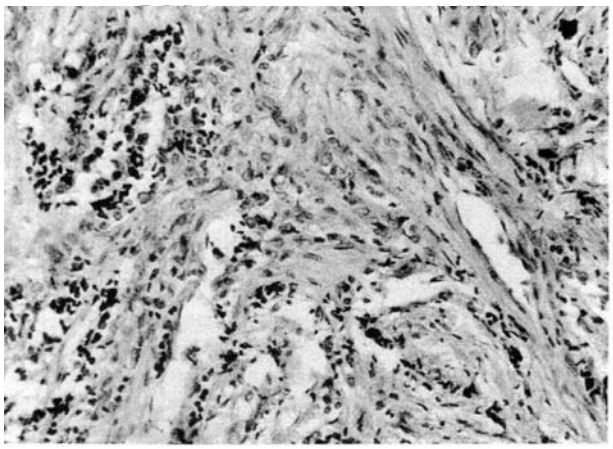

FIG. 3

Low power micrograph showing tumour cell nests in hypocellular connective tissue.

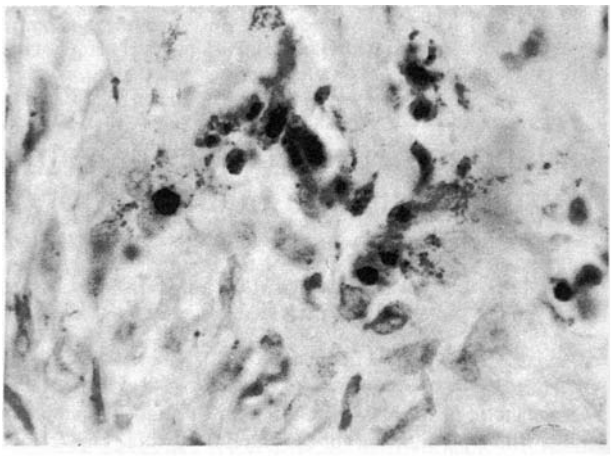

FIG. 4

High power micrograph showing pigmented and non-pigmented cells aligned in clusters.

the postaural region. There was no intracranial extension (Figures 1 and 2).

An excision biopsy under general anaesthesia *via* a transverse skin incision was performed. The mass was found to be encapsulated and was dissected off the squamous and occipital bone *in toto*. The facial nerve was not involved.

The patient had an uneventful post-operative course and was sent home on the tenth day. The patient has been asymptomatic on follow-up for the last six months.

Histological examination revealed a tumour composed of isolated nests of cells contained in a background of dense, hypocellular connective tissue. Pigmented and non-pigmented cells of neuro-ectodermal derivation were aligned in clusters (Figures 3 and 4).

Discussion

The melanotic neuro-ectodermal tumour of infancy is a rare tumour and may become malignant in about three per cent of cases (Batsakis, 1987): nearly 100 cases have been reported in the literature and these were reviewed by this author. The predominant site of origin is the pre-maxilla. Johnson *et al.* (1983) and Hupp *et al.* (1981) have reported that up to 80 per cent of these tumours occur in this region. There is no sex predilection, both are affected equally. The majority of cases manifest themselves in the first year of life. This tumour tends to recur locally in 10 to 15 per cent of patients following excision (Dehner *et al.*, 1979; Cutler *et al.*, 1981). The recurrences occur in a bimodal time pattern in which some occur within 30 days and others are delayed (Block *et al.*, 1980; Batsakis, 1987). The tumour is often multifocal (Batsakis, 1987).

The treatment protocols vary from curettage, with excision of the mass with no margins, to excision of the mass with 5 mm clinically normal surrounding margins. In the presence of rapid and repeated recurrences, the possibility of malignant degeneration should be considered and treated aggressively with surgery, high dose radiation and multi-agent chemotherapeutic agents similar to those used for neuroblastomas. Despite this aggressive treatment the overall prognosis is poor in the rare cases with malignant change (Block *et al.*, 1980).

The treatment of choice for this uncommon benign tumour, with low malignant potential, is conservative local excision with long-term follow-up.

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