PIGMENTED NEUROEPITHELIAL TUMOUR OF INFANCY

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Pigmented neuroepithelial tumour of infancy is an uncommon tumour arising typically in the anterior part of the maxilla in an infant under the age of one year. Since first described by Krompecher in 1918, who termed it "congenital melanocarcinoma", it has been described under a large number of synonyms; Nosicka and Spacek (1978) listed sixteen. Formerly believed to behave always in a benign manner, recent reports affirm multicentric origin (Pontius et al, 1965) and a malignant form akin to neuroblastoma with raised urinary V.M.A. levels (Block et al., 1980). There have been three other cases reported from India (Bhargava et al., 1977).

Case Report

A female child of age five months was brought with a history of a painless swelling of the left maxilla of two months duration. The tumour had apparently appeared spontaneously in the anterior maxillary alveolus. A curettage, followed later by an excision biopsy, was performed in another hospital one month earlier and both tissue samples were reported as chronic inflammatory reaction. The tumour, nevertheless, rapidly increased in size and at admission measured 3×3×4 cm., bulging the left maxillary alveolus, palate and antero—lateral wall. (Fig. 1 and 2). The covering gingival mucosa was coloured bluelish-black. The regional lymph nodes were not enlarged. X-rays showed that the tumour had grown into the antral cavity. (Fig. 3). With a working diagnosis of Sarcoma, a deep biopsy was taken under general anaesthesia. The histological diagnosis was melanotic progonoma. The tumour was completely excised by a partial maxillectomy and the defect covered with a sheet of split-thickness skin graft. At the sixth monthly review the child was well and without any evidence of the tumour.

Disection of the maxillectomy speciment revealed a 2×2 cm. fleshy mass completely filling up the maxillary sinus. (Figs. 4 and 5). It was white with irregular specks of blackish pigmentation. Microscopically, it consisted of a fibrocollagenous background and tumour cells of two distinctive morphology. The larger cells were arranged in poorly formed acini or nests. Fine brown pigment granules were diffusely distributed in the cytoplasm. The pigment was confirmed to be melanin by Schmorl's technique. The other cells were seen in clusters slightly retracted from the stroma. (Fig. 6). The combination of pigmented epithelium and neuroblast like cells in this site in a five month old infant were classical for the diagnosis of pigmented neuroepithelial tumour of infancy.

Discussion

Pigmented neuroepithelial tumour of infancy may present as a pigmented or non-pigmented epulis, or it may be contained within the bone (Pindborg et al, 1971).

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The area of bone dectruction may be traversed by bony septa and the developing teeth are often displaced. The cut surface of the lesional tissue shows pigmentation varying from mottled grey patches to a uniform deep black. At the margins the tumour sometimes extends irregularly into the bone, so that the lesion appears invasive.

In a series of 158 reported cases, five were described as malignant, a rate of 3.2 per cent (Cutler et. al, 1981). The everwhelming number of lesions (95 per cent) occur in children under one year of age. There is no sex prediliction. The most common sites of origin are the maxilla (68.8%), skull (10.8%), mandible (5.8%), and brain (4.3%), 92.8% occuring in the head and neck region. The local recurrence rate is 10 to 15 per cent. Malignancy may arise in a benign lesion or de novo.

Block et. al (1980) reported a case with four recurrences after surgical excisions. At autopsy tumour deposits were found in all major lymph node groups, visceral pleura, bone marrow and liver. Histologically, the tumour cells resembled a neuroblastoma. There was nothing in the clinical presentation or the histologic appearance of the original lesion to indicate the fatal outcome. Palacios (1980) reported another case of malignant transformation with neuroblastic features in a lesion arising in the mandible.

Elevated urinary V.M.A. levels have been found in a limited number of cases. High urinary V.M.A. levels are found in tumours arising from the neural crest as neuroblastoma, ganglioneuroblastoma and pheochromocyloma. The urinary V.M.A. levels return to normal after removal of the tumour (Borello and Gorlin, 1966).

The histogenesis of this tumour has attracted much speculation, the main hypotheses being :

1. Aberration of dental epithelium—"malanotic epithelial odontome" (Mummery and Pitts, 1926).
2. Entrapment of retinal anlage in embryonic fusion lines (Halpert and Patzer, 1947).
3. Atavism of sensory neuroectodermal origin based on similarities with the vomeronasal organ of Jacobson—melanotic progonoma (Stowens, 1957),
4. High urinary V.M.A. levels suggestive of neural crest origin (Borello and Gorlin, 1966).
5. Ultrastructural studies demonstrate the prescence of neurogenic cells (Dehner et al., 1979).
6. Tissue culture studies show that late in the course only the smaller neuroblast-like cells were present and this metamorphoses in the tumour paralleled the rise in urinary V.M.A. levels (Block et al., 1980).

Except for melanoma, there are few interosseous pigmented lesions. The similarity between the pigmented pineal gland of foetuses and infants and this tumour led Dooling et al., (1977) to suggest that differentiating neuroepithelial cells may normally be capable of melanin production. The electron microscopic demonstration of neural structures in addition to melanoblasts supports a neuroepithelial hypothesis (Misugi, 1965 ; Hayward, 1969, and Nikai, 1977).

It is presumed that the neural crest gives rise to the spinal and cervical ganglia, the peripheral autonomic system, the chromaffin cell system, Schwann cells, leptomeninges,

the melanoblasts of the skin, choroid and iris, and that its cranial part also gives rise to skeletal element and odontoblasts (Bolande, 1974). According to Nozue and Kayano (1978) undifferentiated cells of the dental papillae may represent undifferentiated neural crest cells, in addition to the dental follicle, cementoplasts and fibroblasts, which may also be of neural crest origin. Nozicka and Specek (1978) hypothesized that undifferentiated cells are stem cells that may develop into both nerve cells and melamocytes.

Treatment

Recent case reports have made it clear that this tumour can no longer be considered to be entirely benign in its course. Curettage and enucleation are inadequate for its extirpation. A careful, complete oxcision followed by periodical reviews is essential. On the two premises of the correct recognition of the pathologic entity and then the prediction of its biologic behavior rests the proper therapeutic approach to the tumour.

Summary

A case of pigmented neuroepithelial tumour of infancy involving the maxilla is presented. Recent knowledge of its multicentric origin and capacity to metamorphose into a neuroblastoma necessitate a reappraisal in the policy of its management. A correct histopathological diagnosis is essential for proper surgery which is curative in large majority of cases. Rarity of the lesion leading to lack of awarenoess and mistaken diagnosis of malignent lesion may in turn create problems in the management and prognosticetion.

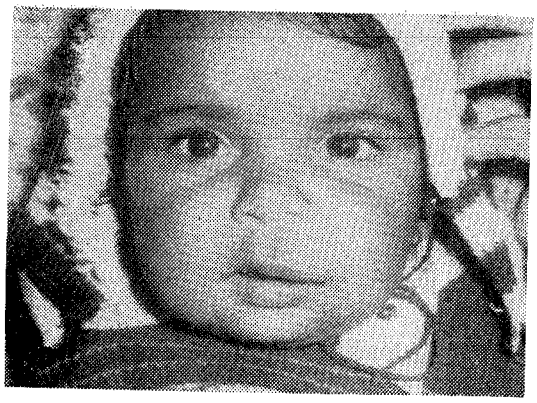

Fig. 1. Swelling of left check with lifting of the alar base.

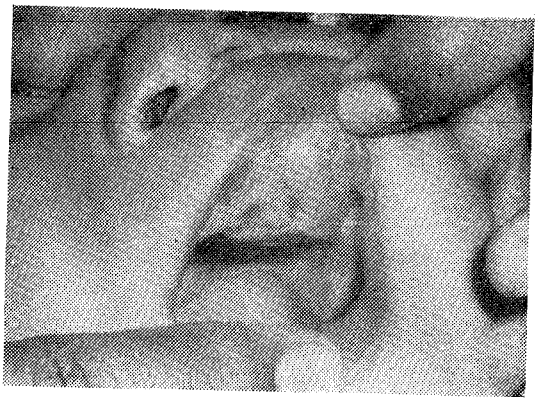

Fig. 2. Tumour bulging the alveolus, palate and antero-lateral wall of maxilla.

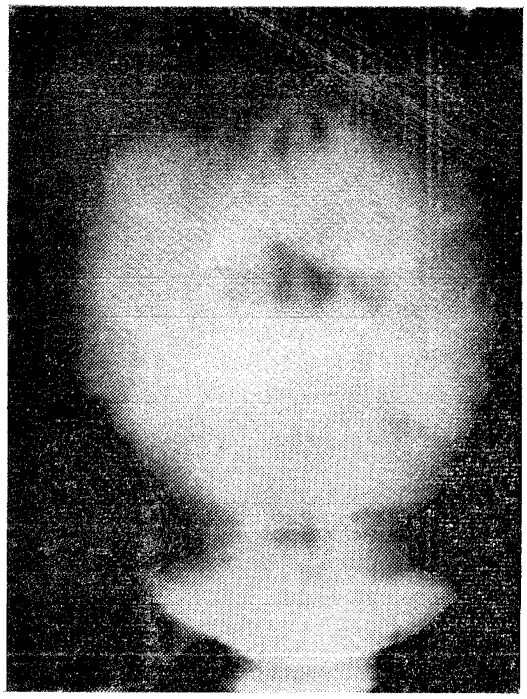

Fig. 3. Radiograph showing extent of involvement of the left maxillary antrum.

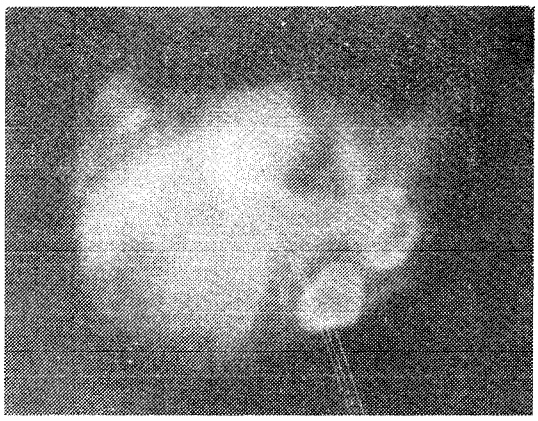

Fig. 4. Radiograph of resected specimen showing displaced tooth germs.

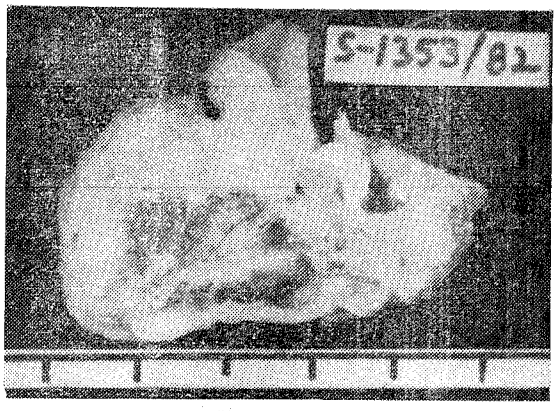

Fig. 5. Cut section of maxilla showing tumour occupying the sinus cavity.

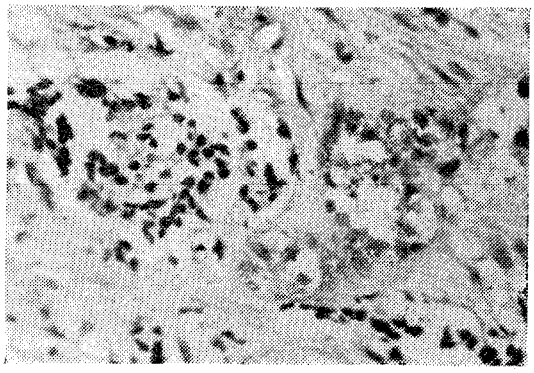

Fig. 6. Microphotograph showing two types of cells. Fine granularity of the larger cells is due to molamin. Haematoxylin and Eosin 430 .

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