MELANOTIC PROGNOMA

(A Case Report)

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Introduction

The melanotic neuroectodermal tumor of infancy has distinct clinical and pathologic features. It is rare, usually benign, asymptomatic pigmented lesion that arises in the maxilla of children below 1 year of age [1,2]. However, the tumor has also been reported in adults [3], in sites other than maxilla [4] and occasionally, it is characterized by a malignant behaviour [5]. We present a case of melanotic prognoma of maxilla in infancy with review of literature.

Case Report

A 4-month-old male infant was admitted with history of gradually increasing painless swelling in the maxilla since birth. On examination, the upper lip was stretched over an apparently globular, sessile swelling 2.5 x 2.0 x 1.5 cm, in the anterior alveolus obliterating the left anterior maxillary gingival vestibule. The external surface of tumor was smooth, hard in consistency, fixed and nontender. The auricular and cervical group of lymph nodes were not enlarged. Routine investigations were within normal limits. Radiograph of the maxilla showed a soft tissue mass arising from the canine region of the left alveolus. Patient was operated and local excision was carried out. Recovery was uneventful. The patient is under close follow-up. The appearance of the face has distinctly improved. Histology revealed round and cleft like spaces lined with polyhedral cells with hyperchromatic nuclei, cytoplasm contains coarse melanin pigment (confirmed by Masson - Fontana stain). In the background loose connective tissue is seen (Fig 1).

Discussion

Melanotic neuroectodermal tumor of infancy is an unusual tumour known by a variety of names, including melanotic epithelial odontome, odontogenic hamartoma, melanotic adamantinoma, melanoameloblastoma, retinal anlage tumour, pigmented epulis of neuroepithelial origin, melanotic prognoma, pigmented ameloblastoma, melanotic ameloblastic odontoma, congenital melanocarcinoma, pigmented congenital epulis, retinoblastic teratoma and neuroectodermal prognoma. This reflects the predominant histogenetic theories of an odontogenic, neural crest

and primordial germ origin for this tumour. Ninety two percent of tumours were in the head and neck area of which 2/3rd were present in maxilla. Ninety two percent of cases occurred in children less than 1 year of age [4]. The nature of these lesions is uncertain and had the features of both a hamartoma and a true neoplasm with debatable histogenesis. The hypothesis that these pigmented neoplasm was derived from the neural crest is supported by ultrastructural studies and by histochemical and biochemical evidence for the secretion of hormones related to the catecholamines [5]. In one small series, immunophenotypic profile of the tumour revealed the larger pigmented epithelioid cells stained with cytokeratin and the melanoma associated antigen HMB-45, whereas the small cells (in 2 of 3 tumours) reacted with neuron specific enolase and synaptophysin [6]. Both cell types reacted with vimentin antibody. Fairly similar results were obtained in another study [7]. Cutler et al in their review of 159 cases of pigmented neuroectodermal tumour, found 6 tumours that behaved in a malignant fashion. In 3 of the 6 cases the tumor was in maxilla and 1 each in

Fig. 1 : Acinar and cleft like spaces lined with melanin containing polyhedral cells.

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uterus, ovary and brain. The recurrence rate is reported to be 15 per cent, which showed its malignant potential. Generally this tumour pursues a benign course and vast majority of children are cured by local resection, without the need of radiotherapy or chemotherapy.

REFERENCES

1. Lopez J. Melanotic neuroectodermal tumour of infancy: Review of the literature and report of a case. *J Am Dent Assoc* 1976; 93: 1159-65.
2. Kukreja HK, Chhangani DL, Duggal N, Joshi KC. Melanotic progonoma of the maxilla. *J Laryngol Otol* 1977; 91: 981-4.
3. Lurie HI. Congenital melanocarcinoma, melanotic adamantinoma, retinal anlage tumor, progonoma and pigmented epulis of infancy: summary & review of literature and report of the first case in an adult. *Cancer* 1961; 14: 1090-108.
4. Cutler LS, Chaudhury AP, Topazian R. Melanotic neuroectodermal tumor of infancy: an ultrastructural study, literature review and reevaluation. *Cancer* 1981; 48: 257-70.
5. Dehner LP, Sibley RK, Sauk JJ Jr, et al. Malignant melanotic neuroectodermal tumor of infancy: a clinical, pathological, ultra-structural and tissue culture study. *Cancer* 1979; 43: 1389-410.
6. Raju U, Zarbo RJ, Regezi JA et al. Melanotic neuroectodermal tumours of infancy: intermediate filament, neuroendocrine, and melanoma associated antigen profiles, *Appl Immunohistochem* 1993; 1: 69-75.
7. Kapadia SB, Frisman DM, Hitchcock CL, Popek EJ. Melanotic neuroectodermal tumour of infancy: clinico-pathological, immuno-histological, and flowcytometric study. *Am J Surg Pathol* 1993; 17: 566-74.