

Melanotic Neuroectodermal Tumour of Infancy

Pages with reference to book, From 75 To 76

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Introduction

Melanotic neuroectodermal tumour of infancy is a rare neoplasm arising from cells derived from the neural crest. It is usually benign but may be malignant in a few cases. Its usual site is the maxilla but may arise in other sites. The treatment is surgical excision and requires long-term follow-up to watch for any local recurrence or distant metastasis.

Case Report

A six months old infant presented with a localised swelling of the upper gum. Past history disclosed admission to the hospital after birth for fever. X-ray of the maxilla showed a cystic lesion resembling a dentigerous cyst. Physical examination, routine urine examination and complete blood counts did not reveal any significant findings. The specimen was surgically removed by a dental surgeon and sent to the Department of Pathology, for pathological examination. It consisted of a rounded, soft black mass, measuring 3.0 cm in diameter. Hematoxylin and eosin stained sections revealed two types of tumour cells embedded in abundant fibrous stroma (Figure).



Figure. Melanotic neuroectodermal tumour of infancy showing two types of neoplastic cells embedded in fibrous stroma. One type of cell is large epithelial-like melanin producing cell and the second is small, non-pigmented lymphocyte-like cells called neuroblasts (H&E stain x 200).

One type of cell was cuboidal to columnar in shape with an epithelial appearance, having abundant cytoplasm. The cytoplasm of these cells showed scanty to heavy accumulation of brown pigment. The pigment was stained as black granules with Masson-Fontana technique and bleached with 0.25% potassium permanganate and with 10% hydrogen peroxide, thus being consistent with melanin pigment. These cells were aligned along cleft-like spaces or arranged in small ductal structures. The second type of cells were small, round, lymphocyte-like cells, which had darkly staining nuclei and scanty cytoplasm - these were neuroblasts. The histological diagnosis of melanotic neuroectodermal tumour of infancy was made. The patient is on follow-up for the last one year and so far there is no evidence of local recurrence or distant metastases.

Discussion

Melanotic neuroectodermal tumour of infancy is a rare tumour^{1,2}. Approximately 164 cases have been reported in the literature from 1983 to 1994. The tumour most commonly occurs in the maxilla, but some cases have also occurred in the mandible, skull, long bones, epididymis, mediastinum and soft tissues of the limbs^{3,4}. It was previously thought to arise from odontogenic epithelium but it is now confirmed that it originates from neural crest cells. The confirmation has been made by immunohistochemical demonstration of neuron-specific antigens, electron microscopic demonstration

of melanosomes and neurosecretory granules and in few cases, differentiation of neuroblasts into mature neurons and secretion of Vanillyl Mandelic Acid (VMA) by the tumour cells⁵⁻⁸. The tumour is usually benign, but some cases have behaved in a locally aggressive fashion, while few others have resulted in distant metastasis^{9,10}. Few cases have revealed presence of heterologous tissue elements like rhabdomyoblasts and bone in the tumour¹¹. There are many synonyms of melanotic neuroectodermal tumour of infancy. These include pigmented neuroectodermal tumour of infancy, melanotic progonoma and retinal anlage tumours¹². The last name is probably based on the fact that the histological structure of the tumour recapitulates that of the retina at 5 weeks of pregnancy. The tumour has also a striking similarity to the pineal glands of foetus and infants¹³. This histological similarity further supports the idea that the tumour is of neural origin, as both the retina and pineal gland are neural structures.

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