

MELANOTIC NEUROECTODERMAL TUMOR OF INFANCY*

A Case Report

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Melanotic neuroectodermal tumor of infancy (MNTI) is a rare, usually benign tumor occurring in children under one year of age. Since first described in 1918, 168 cases have been reported in the literature¹⁻⁵. Four cases had previously been reported in Turkey⁶⁻⁸. This tumor has also been referred to as congenital melanocarcinoma, pigmented ameloblastoma, melanotic ameloblastic odontoma, melanotic progonoma, retinal anlage tumor, retinoblastic teratoma, pigmented congenital epulis, each implying a proposed origin of the tumor. However, in recent years ultrastructural, biochemical, and histochemical studies have strongly suggested a neural crest origin^{1,2,5,9}. In this article, a case of MNTI is presented with a brief review of the literature.

Case Report

A six-month-old male infant was admitted to the Ear-Nose-Throat Department of Çukurova University Hospital because of a swelling on his maxilla noticed by his parents for a month. Physical examination revealed a semi-solid mass covered with normal mucosa which was palpated on the anterior maxilla. The bluish mass extended onto the hard palate posteriorly, left canine fossa laterally, and pushed the upper lip outwards (Fig. 1). No lymph nodes were palpated in the head and neck region. Radiologic examination revealed a soft tissue mass without any bone destruction. Other laboratory findings were within normal limits.

During the excision of the mass, it was noted that the tumor was 4 x 4 x 2 cm in diameter with no apparent capsule. Two teeth germs attached to the tumor also came out.

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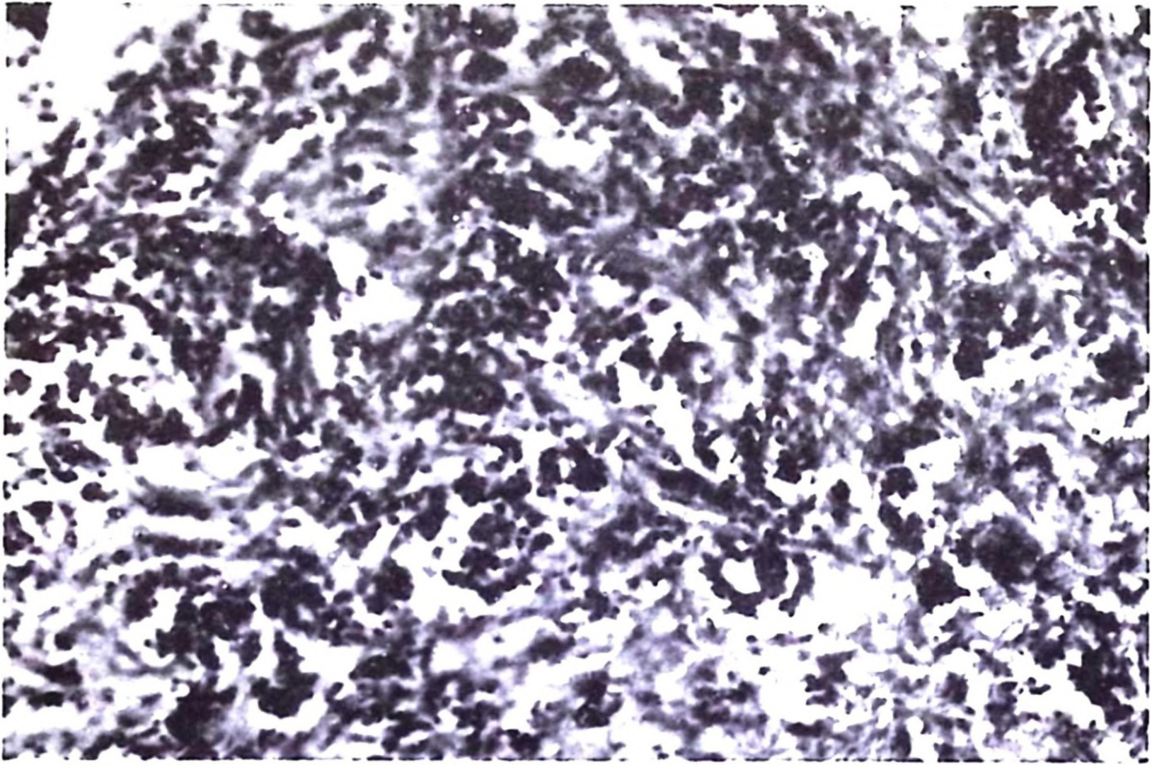


Fig. 1: Melanotic neuroectodermal tumor, composed of epithelial-like cells and small immature cells, forming tubules, columns, and groups are seen in the fibrous stroma. Melanin pigment is noticed in the cytoplasm of some epithelial cells. (H and E x 150).

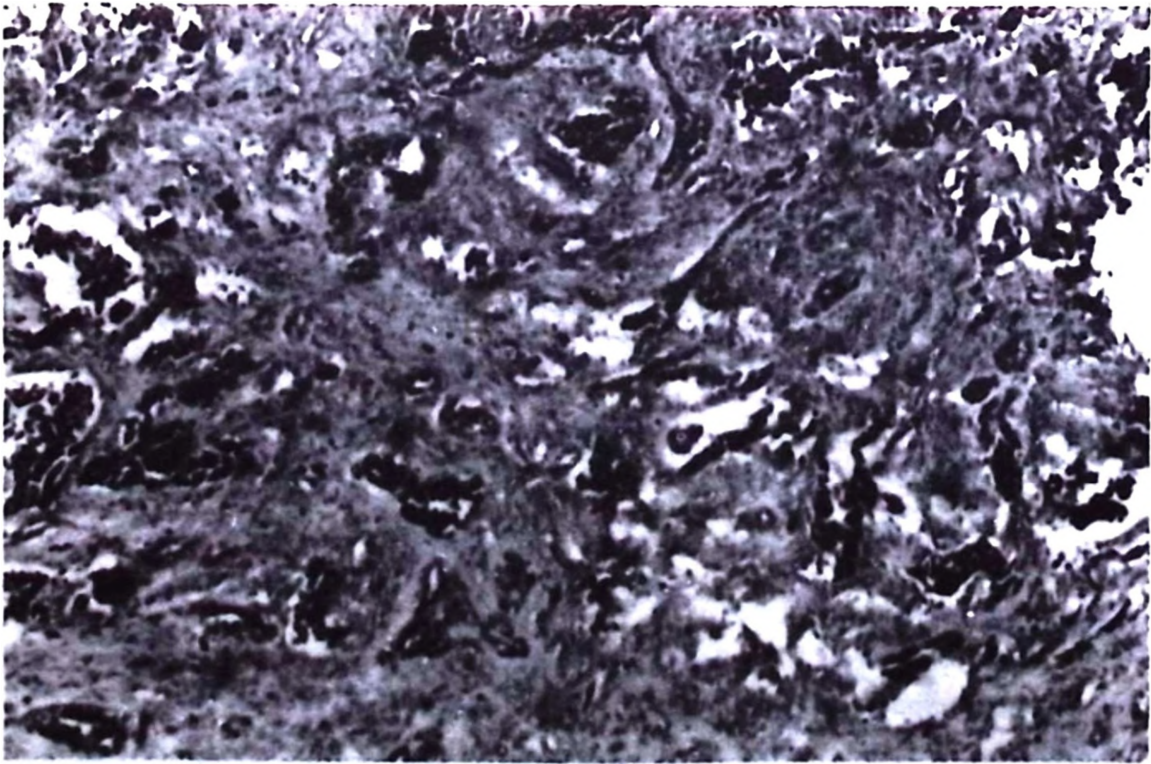


Fig. 2: An admixture of immature small cells and epithelial-like cells are seen.

Microscopic examination revealed a tumor composed of atypical epithelial-like cells and scattered lymphocyte-like immature cells in a dense fibrous stroma. The epithelial-like cells with oval and round hyperchromatic nuclei and pale cytoplasm were arranged in strands, solid sheets, and tubules. Some of the epithelial-like cells contained melanin pigment. These morphological findings are typical of melanotic neuroectodermal tumor (Figs. 1 and 2). The infant has been symptom-free for sixteen months.

Discussion

Two of the characteristics of MNTI are its site of predilection and age of onset. The head and neck region sites account for 92.8 percent of MNTI with 80 percent arising on the maxilla. The vast majority (95%) of MNTI present in children under one year of age, and there is no sex predilection^{1,5,9}. As Batsakis⁹ has described, the classical histologic appearance of MNTI has two cell types in the company of a fibrous connective tissue stroma; one form consists of small, lymphocyte-like or neuroblast-like cells while the other is larger, epithelium-like, with melanin-containing cells arranged in strands or cords. In the case presented, the tumor was located in the maxilla of a six-month-old infant with typical histopathologic findings of MNTI.

Of the 168 cases reported, only eight have been designated malignant. If MNTI of the head and neck region is only considered, the malignancy rate is 2.3 percent^{1,9}. As Cutler et al¹ have stressed, even though MNTI is usually benign, this rate of malignancy is striking especially when comparing the rate of malignant melanomas to benign nevi in adults (0.0006 per 100 nevi). Furthermore, the incidence of malignant melanoma is 40 times less in children than in adults. The local recurrence rate has been reported to be 10 to 15 percent. Recurrences may be explained partially by insufficient resection and partially by the multifocal pattern often seen in these tumors. In some cases, recurrence may occur within 30 days after primary treatment, while in others it may be delayed for longer periods. Thus, MNTI cases require a long term follow-up⁹. Simple surgical excision of the tumor is the treatment of choice^{1,5,9,10}.

Summary

A typical case of melanotic neuroectodermal tumor of infancy, located on the maxilla of a six-month-old boy is presented and the literature is discussed.

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