

Melanotic neuroectodermal tumor of infancy

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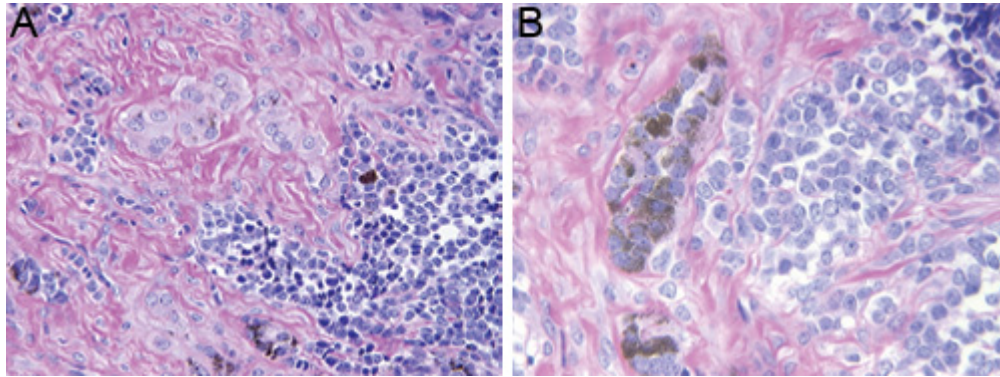


Figure. A: An intermediate-power view shows the biphasic tumor cell population, with large and small cells. B: High-power view shows a fibrous connective tissue stroma separating a nest of large, heavily melanin-pigmented cells. The large cells are separated from small cells that have high nuclear-to-cytoplasmic ratios and hyperchromatic nuclei.

Melanotic neuroectodermal tumor of infancy is a rare, neural-crest–derived neoplasm that is believed to be congenital. The tumor has a marked predilection for the head and neck—particularly the maxilla, where approximately 70% of these tumors are located. The anterior maxilla is most commonly affected. There is no predilection for either sex. Nearly all patients present with an enlarging mass, usually within a few years of birth. Intraoral lesions may appear “blue,” suggesting the presence of pigment. Radiographic images will often show a destructive lesion with tooth displacement, but they are nonspecific. Laboratory studies show high urinary levels of vanillylmandelic acid.

Histologically, the tumor is characterized by a biphasic tumor cell population in a trabecular, tubular, or alveolar arrangement (figure, A). Larger pigmented cells surround groups of the smaller, round-to-oval, blue neuroectodermal cells (figure, B). The trabeculae are separated by a dense collagenous stroma. The periphery of the tumor is usually infiltrative into the surrounding bone or soft tissue. The large cells are epithelioid, cuboidal cells that line the exterior of the alveolar spaces or trabeculae. They demonstrate relatively large, vesicular nuclei with prominent nucleoli that are surrounded by eosinophilic cytoplasm with ill-defined cell borders. The cytoplasm contains variable amounts of medium- to dark-brown melanin granules (figure, B). The small, round-to-oval cells contain a hyperchromatic nucleus and scant cytoplasm. Immunohistochemically, both types of tumor cells are reactive to vimentin and neuron-specific enolase; the larger cells may also be reactive to keratin and HMB-45. The tumor must be differentiated from melanoma, olfactory neuroblastoma, lymphoma, and rhabdomyosarcoma. Rare examples of pigmented primitive neuroectodermal tumors may be included in the differential diagnosis.

Complete surgical excision with histologically clear margins (0.5 cm) is the treatment of choice. However, complete extirpation is difficult to achieve, and postoperative bleeding and infection can complicate surgery. Recurrences develop quite frequently when the initial surgery is inadequate. The tumor’s histologic appearance does not reliably predict its clinical course. Metastasis is rare.

Suggested reading

- Barrett AW, Morgan M, Ramsay AD, et al. A clinicopathologic and immunohistochemical analysis of melanotic neuroectodermal tumor of infancy. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2002;93:688-98.
- Gaiger de Oliveira M, Thompson LDR, Chaves AC, et al. Management of melanotic neuroectodermal tumor of infancy. *Ann Diagn Pathol* 2004;8:207-12.
- Kapadia SB, Frisman DM, Hitchcock CL, et al. Melanotic neuroectodermal tumor of infancy. Clinicopathological, immunohistochemical, and flow cytometric study. *Am J Surg Pathol* 1993;17:566-73.