AJNR

Melanotic neuroectodermal tumor of infancy.

J C George, M K Edwards, R I Jakacki and J Kho-Duffin

AJNR Am J Neuroradiol 1995, 16 (6) 1273-1275
http://www.ajnr.org/content/16/6/1273

Melanotic Neuroectodermal Tumor of Infancy

Joseph C. George, Mary K. Edwards, Regina I. Jakacki, and Jennie Kho-Duffin

Summary: We present a case of malignant melanotic neuroectodermal tumor of infancy arising in the skull and secondarily invading brain. The central tumor was hyperintense to brain on T1-weighted images and hypointense to brain on T2-weighted images. This appearance corresponded to the surgical and histologic findings of melanin-containing tumor.

Index terms: Neuroectodermal tumor; Skull, neoplasms; Brain, neoplasms; Children, neoplasms

Melanotic neuroectodermal tumor of infancy (melanotic progonoma) is a rare tumor of neural crest origin that is commonly found in the maxilla of infants (1). Brain involvement is rare (2–4). One report cites malignancy in six cases (1). We describe a case of melanotic neuroectodermal tumor that presented as an aggressive skull-based tumor that was resected and later recurred in the posterior fossa. One year later, the tumor metastasized to the subarachnoid spaces. The imaging findings of this tumor are described.

Case Report

A 19-month-old patient presented to an outside institution with acute obtundation and eventual unresponsiveness. Plain radiographs showed bone expansion and hyperostosis involving the left temporoparietal calvarium (Fig 1A). Noncontrast computed tomography (CT) scans showed a hyperdense mass and hydrocephalus (Fig 1B). Magnetic resonance (MR) images showed involvement of both infratentorial and supratentorial brain compartments (Fig 1C). The central tumor was hyperintense to brain on T1-weighted images and hypointense on T2-weighted images. Portions of the tumor in close apposition to bone were hypointense on both T1- and T2-weighted images. The tumor showed intense enhancement after intravenous gadolinium administration (Fig 1D). The patient underwent radical debulking surgery. A pathologic diagnosis of ganglioglioma with stromal melanosis was made.

The patient received no further treatment until the age of 3½ years, when she presented to our institution with progressive headaches, weakness, and lethargy. Follow-up MR showed tumor recurrence in the posterior fossa. The patient underwent surgery again. The tumor was difficult to resect because of extensive tumoral calcification and firm adherence to underlying bone. Foci of black pigmentation were seen. Histologic sections of the tumor showed abundant pigmented cells forming glandlike structures suspended in a collagen-vascular stroma (Fig 1E). The cells stained positive for melanin. Neoplastic ganglion cells suspended in a delicate fibrillary network also were seen and stained positive for neuroectoderm. A large portion of the tumor contained small, tightly packed undifferentiated cells. A pathologic diagnosis of partly malignant melanotic neuroectodermal tumor of infancy was made. The diagnosis was confirmed with a review of tissue sections from the original surgery.

Discussion

Melanotic neuroectodermal tumor of infancy has been termed *melanotic ameloblastoma* and *retinal anlage tumor* because of prior controversy regarding its origin from odontogenic epithelium versus displaced retinal anlage, respectively (3). The tumor now is widely accepted as being of neuroectodermal origin on the basis of ultrastructural, immunocytochemical, and electron microscopic studies (2). Melanotic neuroectodermal tumor is an uncommon lesion. Our survey indicates 179 cases in the literature (1–3). The majority of lesions arise in the maxilla (68.8%), followed by the skull (10.8%), mandible (5.8%), and brain (4.3%) (2). Although the majority of tumors are benign, a local recurrence rate of 10% to 15% and a malignancy rate of 3.2% is reported (2).

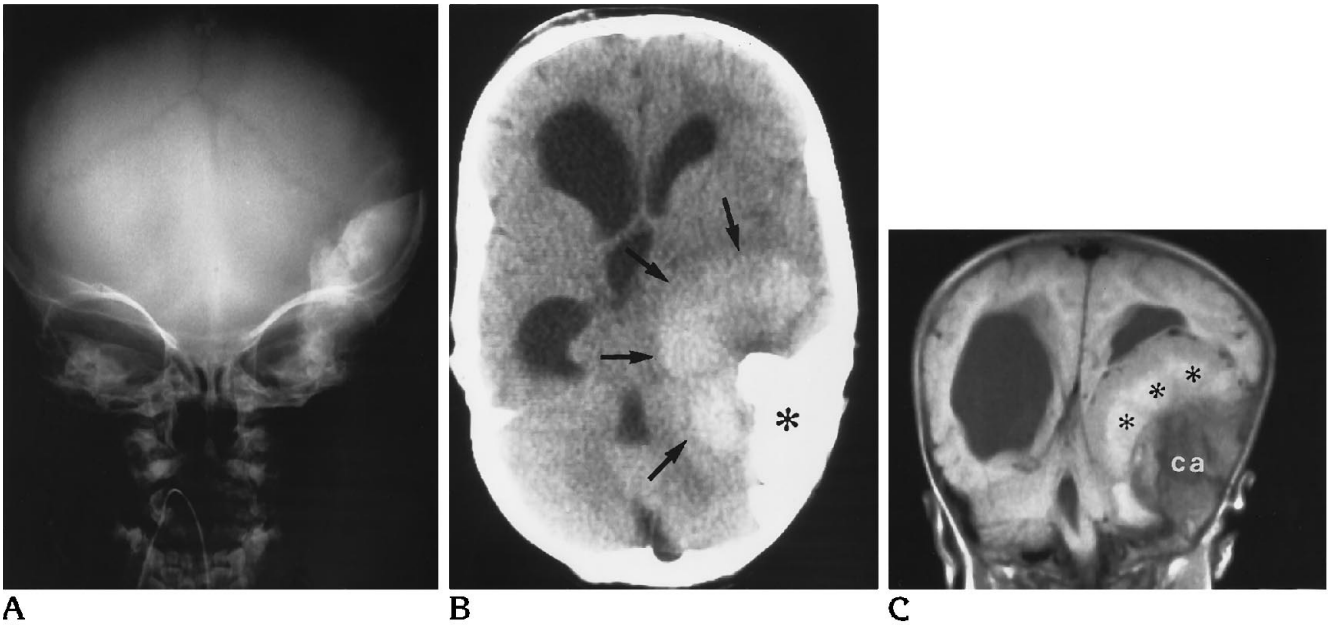
There are several prior reports of neuroectodermal tumor of infancy arising from the skull, in particular, the anterior fontanelle region (1, 2,

Received December 27, 1993; accepted after revision June 2, 1994.

From the Departments of Radiology (J.C.G., M.K.E.), Oncology (R.I.J.), and Pathology (J.K.-D.), Indiana University School of Medicine, James Whitcomb Riley Hospital for Children, Indianapolis.

Address reprint requests to Joseph C. George, MD, Department of Radiology, Indiana University School of Medicine, James Whitcomb Riley Hospital for Children, 702 Barnhill Dr, Indianapolis, IN 46202-5200.

AJNR 16:1273–1275, Jun 1995 0195-6108/95/1606–1273



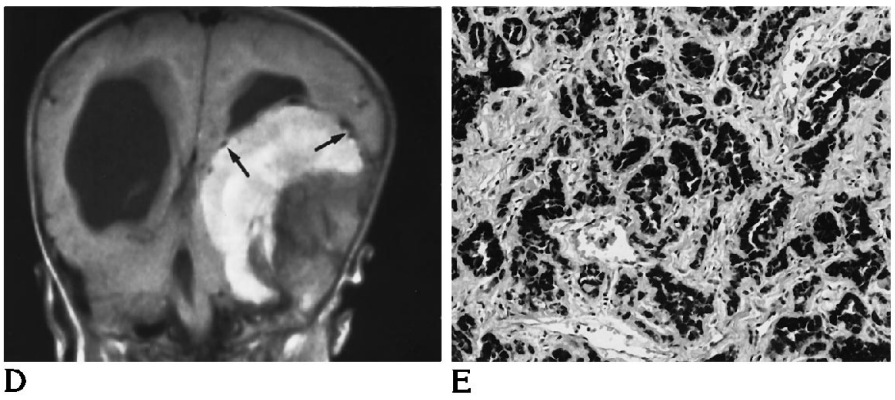


Fig 1. *Melanotic neuroectodermal tumor of infancy.*

A, Plain radiograph shows calcified mass arising from left temporoparietal calvarium.

B, Axial noncontrast CT scan shows a hyperdense mass (*arrows*) arising from the left parietotemporal calvarium. Note associated hyperostosis (*asterisk*) and moderate hydrocephalus.

C, Coronal spin-echo (600/30/2 [repetition time/echo time/excitations]) noncontrast MR scan shows three areas of signal intensity within the tumor. The tumor in apposition to bone is hypointense to brain, corresponding to areas of tumor calcification (*ca*). Centrally, the tumor is hyperintense and corresponds to areas of melanosis (*asterisks*). The peripheral tumor is isointense to brain.

D, Coronal spin-echo (600/30/2) MR scan after intravenous gadolinium shows intense enhancement of central and peripheral tumor. Note flow voids on tumor surface (*arrows*) from displaced cortical vessels.

E, Photomicrograph shows pigmented (dark) cells forming slits and glandlike structures surrounded by lighter collagenous stroma. (hematoxylin and eosin, magnification ×100).

4). Primary tumors of brain are uncommon and reported cases have arisen in the cerebellar vermis and third ventricle (4–8). Brain parenchymal involvement more commonly results from intraaxial extension of skull lesions (2–4). In one case of melanotic neuroectodermal tumor reported by Mirich et al, the tumor arose in the occipital calvarium and showed hyperostosis and bone spicules within the tumor (3). We believe that the tumor in our patient arose in the temporoparietal skull and secondarily invaded brain. In a reported case of melanotic neuroectodermal tumor with leptomeningeal metastases (8), a 3-year-old boy presented with a cerebellar tumor and underwent subtotal resection. He died 2 months after surgery, and at autopsy, melanotic deposits were found in virtually every part of the subarachnoid space.

The imaging appearance of melanotic neuroectodermal tumor of infancy involving brain is not well described. On plain skull radiographs, adjacent bone may be sclerotic and hyperostotic (3, 4). In one case of an occipital melanotic neuroectodermal tumor reported by Mirich et al, bone spicules within the tumor produced increased density (3).

There are limited reports of the CT and MR appearances of melanotic neuroectodermal tu-

mor (3, 4). The CT scans in our patient showed calvarial hyperostosis, expansion, osteogenesis, and extensive tumoral calcification. The tumor enhanced with intravenous contrast material. These CT findings are in agreement with two prior reports of melanotic neuroectodermal tumor involving the skull and secondarily invading brain (3, 4).

On MR scans, the tumor in apposition to bone was hypointense to brain on both T1- and T2-weighted images and corresponded to histologic areas of calcification. This appearance is related to a lack of hydrogen nuclei and a very short T2 relaxation time attributable to local magnetic field inhomogeneity. Other causes of this MR appearance include chronic hemorrhage (hemosiderin), flow void, and physiologic iron. The central tumor was hyperintense relative to brain on T1-weighted images and correlated with histologic areas of tumor containing large amounts of melanin. These areas were relatively hypointense to brain on T2-weighted images. The tumor periphery was slightly hypointense to gray matter on T1-weighted images and isointense on T2-weighted images. In a previously described case of melanotic neuroectodermal tumor arising from the anterior fontanelle, the tumor was slightly hypointense to brain with a focus of marked hyperintensity on T1-weighted images and isointense on T2-weighted images (4). It was thought that the focus of hyperintensity represented an area of extensive melanosis, similar to the central portion of the tumor in our report. In two MR descriptions of melanotic neuroectodermal tumor arising from the maxilla, the tumor was isointense to brain on T1-weighted images and slightly hyperintense on T2-weighted images (3). This discrepancy suggests the possibility of a variable histology and/or amount of melanin in tumors arising from different locations.

In most cases of melanotic tumor, the paramagnetic effect of melanin produces increased signal on T1-weighted images. This is because melanin acts as a free-radical trap that chelates paramagnetic metal ions, enhancing proton relaxation (4). On T2-weighted images, melanotic tumors show decreased signal. T2 relaxation time is related to magnetic field inhomogeneity in the proton environment that leads to precessional dephasing. Melanin has heterogeneous magnetic susceptibility, which results in a rapid T2 decay and low signal intensity. A similar MR appearance may be seen with subacute hemorrhage containing paramagnetic methemoglobin and in tumors with a high degree of cellularity.

Histologically, melanin may result from cellular differentiation in neuroectodermal tumors (melanotic neuroectodermal tumor, melanotic meningioma, melanotic schwannoma, and primary meningeal melanoma) and from neuroepithelial tumors (melanotic ependymoma, melanotic medulloblastoma, and malignant epithelial choroid plexus tumor) (1). In melanotic neuroectodermal tumor of infancy, clusters of small round cells, primitive glandlike structures, and a collagenous background stroma are diagnostic features.

In summary, we report a case of malignant melanotic neuroectodermal tumor of infancy and describe the CT and MR appearances. MR images corresponded to the histologic findings of tumoral calcification and extensive melanin deposition.

References

1. Russell DS, Rubinstein LJ. *Pathology of Tumours of the Nervous System*. 5th ed. Baltimore: Williams and Wilkins, 1989:720–721
2. Cutler LS, Chaudhry AP, Topazian R. Melanotic neuroectodermal tumor of infancy: an ultrastructural study, literature review, and reevaluation. *Cancer* 1981;48:257–270
3. Mirich DR, Blaser SI, Harwood-Nash DC, Armstrong DC, Becker LE, Posnick JC. Melanotic neuroectodermal tumor of infancy: clinical, radiologic, and pathologic findings in five cases. *AJNR Am J Neuroradiol* 1991;12:689–697
4. Atkinson GO, Davis PC, Patrick LE, Winn KJ, Ball TI, Wyly JB. Melanotic neuroectodermal tumor of infancy. *Pediatr Radiol* 1989; 20:20–22.
5. Stowens D, Lin T. Melanotic prognoma of the brain. *Cancer* 1974; 5:105–113
6. Hahn JF, Sperber EE, Netsky MG. Melanotic neuroectodermal tumors of the brain and skull. *J Neuropathol Exper Neurol* 1976; 35:508–519
7. Fowler M, Simpson DA. A malignant melanin forming tumor of the cerebellum. *J Pathol Bacteriol* 1962;84:307–311
8. Sung JH, Mastri AR, Segal EL. Melanotic medulloblastoma of the cerebellum. *J Neuropathol Exper Neurol* 1973;32:437–445