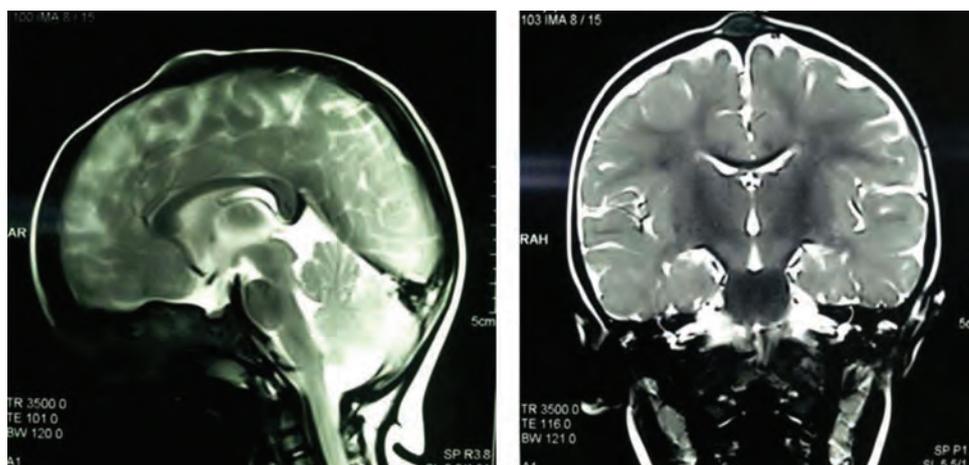


Melanotic Neuroectodermal Tumor of Infancy: A Neuropathologic Image

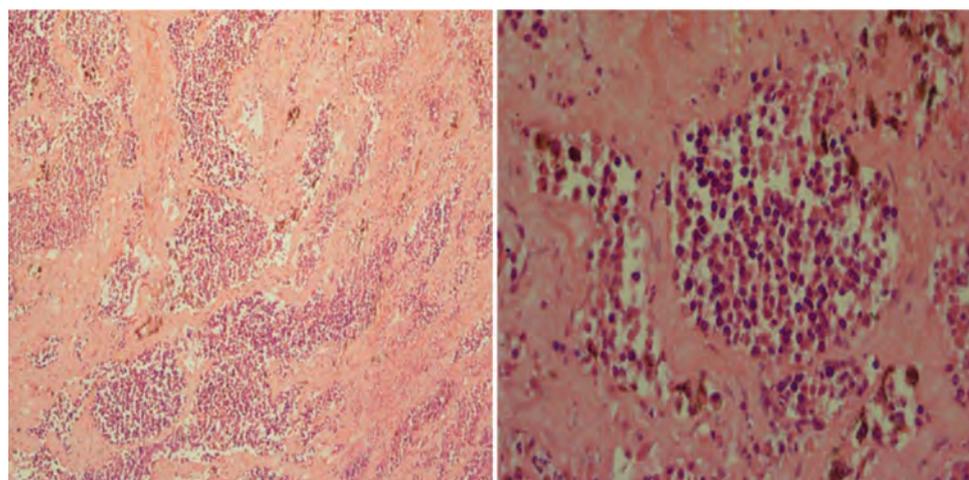
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Figures 1 and 2. Magnetic resonance image (MRI) of melanotic neuroectodermal tumor of infancy (MNTI).



Figures 3 and 4. Sections from the skull mass show a biphasic tumor with some epithelioid cells resembling melanocytes and smaller neuroblast-like cells with diffuse melanin deposition. Hematoxylin and eosin staining. Magnification: Figure 3 (100 \times); Figure 4 (250 \times).

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A seven-month-old infant referred to the Neurosurgery Department of our center due to presence of a bulge in the midline of the skull. This bulge was noticed about three months earlier and had gradually increased in size. On clinical examination there was a firm, non-tender mass that measured 4×3cm at the midline of the skull, above the superior sagittal sinus with no overlying skin changes. There was no evidence of any neurologic deficit. Plain radiograph image of the skull showed bone expansion and calvarial hyperostosis. Magnetic resonance images (MRI) revealed a well-defined oval shaped subcutaneous lesion that measured 4×3×2.5cm in high skull in midline that extended to the dipole region with no signs of extension to the dura mater and brain parenchyma or scalp. The lesion had an iso- to hyposignal intensity compared to the cortex in T2 weighted images and a hypersignal intensity in T1 weighted sequences (Figures 1 and 2).

The patient underwent surgical excision of the tumor with total tumor resection and clear margins. Histopathology study showed a biphasic pigmented neoplasm composed of clusters of tumor cells in nest and cord-like patterns set in a dense collagenous stroma. Tumor cells had a biphasic proliferation pattern with a number of small neuroblastic cells that had dense nuclei and scant cytoplasm and large melanin pigmented epithelial cells with large vesicular nuclei and prominent nucleoli (Figures 3 and 4). Immunohistochemistry (IHC) findings were positive for synaptophysin (SY38), cytokeratin (AE1/AE3), HMB45, chromogranin (DAK-A3) and NSE (BBS/NC/Vi-H14). The tumor was negative for LCA, S100, desmin and MIC2 (CD99). Findings were consistent with a diagnosis of melanotic neuroectodermal tumor of infancy (MNTI). The post-operative course was uneventful.

Melanotic neuroectodermal tumor of infancy (MNTI) is a rare neoplasm of neural crest origin which usually affects infants during the first year of life. It most commonly occurs in the maxilla however the mandible, skull, brain and epididymis, mediastinum, ovaries and uterus can also be involved.¹ It usually presents as a pigmented,

non-tender, non-ulcerated single mass however multiple lesions have also been reported.²⁻⁴

Typical CT radiological findings are of an isohypodense soft tissue component which demonstrates contrast enhancement. Bony involvement is variable and includes a spiculated appearance, indentation of underlying bone and complete bone destruction. Magnetic resonance image findings usually show T1 shortening that is expected with melanin deposition. Its radiologic differential diagnosis consists of Langerhans cell histiocytosis, Ewing sarcoma, lymphoma, neuroblastoma, metastasis and fibromatosis.⁵ Biopsy can be very helpful for surgical planning. Histologically the differential diagnosis of MNTI involves other pediatric small round cell neoplasms that include neuroblastoma, Ewing sarcoma, rhabdomyosarcoma, peripheral neuroepithelioma, desmoplastic small round tumor, malignant melanoma and lymphoma.²

Despite rapid local growth, MNTI is considered to be a benign neoplasm. Adequate surgical excision is the treatment of choice. A high recurrence rate and malignant transformation potential has been reported, therefore early diagnosis by learning its distinctive clinicopathological and imaging features is mandatory as more and more cases are being reported.^{1,3-7}

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