

Melanotic neuroectodermal tumor of infancy in the skull: a case report

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Melanotic neuroectodermal tumor of infancy (MNTI) is a rarely occurring neoplasm of neural crest origin, appearing in children during the first year of life. MNTI is a benign tumor but is nonetheless locally aggressive; it usually originates from the maxilla and the mandible. Recognition of the diagnostic features of MNTI is important, so that it may be differentiated from benign lesions (dermoid or epidermoid cysts, vascular lesions) as well as from malignant lesions such as small round cell tumors. We present a case of a 2-month-old infant who developed right retroauricular swelling with extension through the calvarium and into the central nervous system. The tumor was totally resected. Aggressive benign or malignant lesions of the calvarium may entail dural infiltration and brain involvement, as in our case. Early diagnosis and surgery are thus mandatory for benign or malignant tumors of the calvarium.

Key words: Melanotic neuroectodermal tumor, skull tumor, tumor of infancy, diagnosis treatment.

Melanotic neuroectodermal tumor of infancy (MNTI), first described by Krompecher in 1918, is a rare tumor of neural crest origin¹⁻³. It is classically a benign but locally aggressive tumor that predominantly affects children⁴. It was earlier presumed to originate from odontogenic or retinal rests, and this led to a variety of terminologies being assigned to the tumor, including melanotic epithelial odontoma, retinal anlage tumor, pigmented ameloblastoma, pigmented teratoma, atypical melanoblastoma, retinal choristoma, melanotic adamantinoma, melanotic progonoma, retinoblastic teratoma and pigmented epulis^{1-3,5}. It usually occurs in the head and neck region and predominantly affects the maxilla and mandible in the first year of life⁶. Borello and Gorlin⁵ described melanocytic neuroectodermal tumor of infancy in 1966.

It comprises locoregional aggressive tumors with a more rapid and severe spread. We report an additional case of MNTI of the skull

and a present a review of the literature on its diagnosis and treatment.

Case Report

A 2-month-old boy was admitted to the head and neck department with a right retroauricular swelling. This rapidly growing lesion had been noticed in the last 2 weeks. There was no history of trauma or congenital anomaly. Examination revealed a well-defined, nontender, nonpulsatile, nonulcerated right retroauricular swelling. There was no palpable lymphadenopathy. Laboratory tests, most notably VMA, were normal. No significant neurological findings were noted on examination. Computed tomography (CT) and magnetic resonance imaging (MRI) showed an extracranial and intracranial lesion with (temporal squama) petrosal ridge bone invasion and involvement of the underlying dura mater and the tentorium (Fig. 1). Whole-body screening showed no evidence of distant tumor metastasis.

The tumor was firstly dissected in the subcutaneous area to the healthy area, cut by cautery to the bone and then dissected by periosteal elevator to the bone defect. The infiltrated bone, which included melanocytic pigment, was destroyed, being drilled away to the border of the normal bone. The intracranial part of the tumor, which had invaded the underlying dura mater and the upper part of the tentorium, was totally excised. There was a good dissection plane between the tumor and the brain. The entire tumor was thus excised.

Histopathological examination revealed a melanocytic tumor (Fig. 2). A hematoxylin-eosin-stained specimen showed that the tumor was composed of a heterogeneous cell population, consisting of larger epithelioid cells, mesenchymal cells and small rounded neuroblast-like cells. The mesenchymal component of the tumor cells had fusiform nuclei. The melanin-containing large epithelioid cells were diffuse positive with pan-CK and

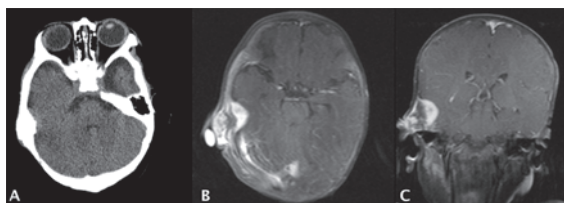


Fig. 1. (A) A preoperative axial CT demonstrating a well-defined, hyperdense lesion indenting the right side of temporal bone. An axial (B) and a coronal (C), contrast-enhanced, T1-weighted images showing solid, intensely enhancing, extracranial and intracranial lesion.

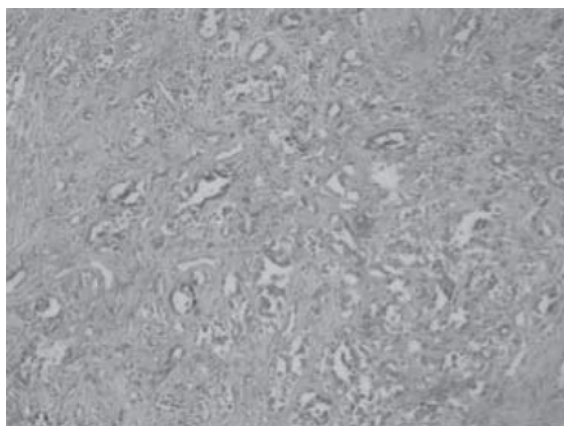


Fig. 2. Histopathological image (Hematoxylin and eosin stain; X100) demonstrated larger epithelioid cells, mesenchymal cells and small rounded neuroblast-like cells.

synaptophysin and focal positive with melen-A, but S-100 protein was negative.

No complications occurred after the surgery. The patient did not receive chemotherapy. Clinical examination and MRI showed no recurrence at the one-year follow-up.

Discussion

MTNI is a locally aggressive and fast-growing but usually benign tumor^{2,5,7}. MNTI clinically presents as a painless, expansile, partly pigmented mass, typically in the maxillary region, which is usually unencapsulated⁴. It may be locally aggressive in 15-36% of cases; 3-7% of tumors have an overtly malignant behavior with distant metastases⁴. The average recurrence rate after surgery is between 10% and 60%^{2,5}.

Approximately 360 cases of MNTI have been reported in the literature⁵. The clinical presentation is a typically a nontender, nonulcerated, progressively enlarging craniofacial mass without associated neurological symptoms^{1,2,5}. The tumor causes compression rather than infiltration of adjacent structures, with local invasion causing bone destruction, tooth displacement and feeding difficulties¹. Most patients are infants less than one year of age, with incidence peaking between two and six months without sex predilection, although a few adult patients have been reported^{1,2,7}. The tumor usually occurs in the head and neck region because of its origin from the neuroectoderm. Of the reported tumors, 68.6% were located in the maxilla, followed by the skull (10.6%), mandible (7.3%), brain (5.3%), epididymis (4%), soft tissues, uterus and mediastinum^{4,7}.

Imaging aids in the diagnosis of MNTI as well as in pre-operative planning; it is imperative in surveillance for recurrence⁵. Conventional radiographs of bony lesions usually show a central lucency with adjacent bony sclerosis and hyperostosis^{4,5}. It is typical of the CT scans to reveal hyperdense masses, but hypodense variants have been reported as well⁴. CT imaging defines the extent of the lesion and osseous involvement for surgical planning^{4,5}. Magnetic resonance imaging shows a hypodense mass, with focal areas of hyperdensity in T1-weighted images and an isointense mass on T2-weighted images⁴.

Recognition of the diagnostic features of MNTI is important, because they may be difficult to differentiate from benign or malignant tumors of the calvarium. Benign lesions include cephal hematoma, infection, dermoid and epidermoid cysts, congenital epulis of the newborn, central giant-cell granuloma, ossifying fibroma, fibrous dysplasia, craniopharyngioma, fibromatosis and vascular malformations^{3,5}. The differential diagnosis of malignant lesions is related to other pediatric “small round cell” neoplasms^{4,5}. These include neuroblastoma, Ewing’s sarcoma, rhabdomyosarcoma, peripheral neuroepithelioma, desmoplastic small round cell tumor, malignant melanoma, peripheral primitive neuroectodermal tumor, Langerhans cell histiocytosis, leukemia and lymphoma³⁻⁵. Reported malignant cases have the same histologic description, with increased mitosis, hypercellularity and focal necrosis³. Metastatic lesions have been described in the lymph nodes, liver, adrenal gland, spinal cord and a variety of other sites³.

Total excision of MNTI is important to prevent a recurrence; however, extension of the intracranial tumor or its location along the cranial midline or base may be problematic for the surgeon⁸. Individuals with MNTI that is not amenable to surgical management alone may receive other modes of treatment⁴. In general, this may be chemotherapy alone; chemotherapy with radiotherapy; chemotherapy before or after surgical treatment; radiotherapy and surgical treatment; or a combination of excision, chemotherapy and radiotherapy⁴.

In conclusion, even though MNTI is a rare cause of calvarial tumors in infancy, it should be considered in the differential diagnosis of all patients presenting with head and neck masses. MRI should be performed for a local and aggressive head and neck tumor before surgery. Although MNTI is considered a benign tumor, early diagnosis and aggressive surgery is essential in dealing with tumors that extend into the CNS, because of their association with morbidity and mortality.

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