

MELANOTIC NEUROECTODERMAL TUMOR OF INFANCY

Case report

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ABSTRACT - *Introduction* Melanotic neuroectodermal tumor of infancy (MNTI) is a rare tumor, locally aggressive, usually originated from maxilla and mandible and rarely from the skull. A case of a 4 month-old child presenting a bulging lesion in the midline of the occipitoparietal region with progressive growth is reported. *Case report* The neurologic examination had normal developmental milestones. Computerized tomography scan and magnetic resonance Image showed a highly enhancing tumor, dislocating anteriorly and inferiorly the superior sinuses. In order to prevent excessive bleeding, surgical resection was performed in three stages, with complete removal. *Conclusion*: Based on the absence of tumor recurrence, we believe in a favorable neurological prognosis and in a possible of cure, although the patient was not submitted to any adjuvant treatment.

KEY WORDS: neuroectodermic melanotic tumor, cranial tumor, tumor of infancy.

Tumor melanótico neuro-ectodérmico da Infância: relato de caso

RESUMO - O tumor melanótico neuro - ectodérmico da infância é raro, localmente agressivo, originado da maxila ou da mandíbula, raramente originando-se da calota craniana. Relatamos o caso de um menino de 4 meses de idade que apresentava lesão abaulada com crescimento progressivo no couro cabeludo. O exame neurológico apresentava desenvolvimento neuro-psicomotor adequado. A tomografia computadorizada e a ressonância magnética do encéfalo mostravam lesão que impregnava pelo contraste, deslocando anterior e inferiormente o seio sagital superior. Com o objetivo de prevenir o risco do sangramento excessivo, o procedimento foi realizado em três tempos, com remoção completa da lesão. Baseados na ausência de recidiva do tumor, acreditamos no bom prognóstico neurológico e na possibilidade de cura, embora o paciente não tenha sido submetido a nenhum tratamento adjuvante.

PALAVRAS-CHAVE: tumor melanótico neuro-ectodérmico, tumor craniano, tumor da infância.

Melanotic neuroectodermal tumor of infancy (MNTI) is a rare lesion. Approximately 200 cases have been reported since Krombecher first described it in 1918¹. Most of the tumors are located in the maxilla and become evident in the first year of life. Only a few cases occur in the neurocranium. Most patients were taken to a hospital because of progressive growth of a craniofacial mass, without associated neurological symptoms.

We report a 4-month-old-male patient with a MNTI in the neurocranium.

CASE

A 4 month-old boy was brought to our hospital with a bulging lesion in the midline of the occipitoparietal

region. There were no remarkable antecedents. His mother noticed progressive growth of "a tumour in his head" beginning on the 20th day of life. On admission, the patient had no relevant neurological findings, and had normal developmental milestones. There was a bulging lesion, firm, non-mobile, painless, and apparently adherent to deeper structures. CT scan revealed a huge median occipitoparietal mass lesion with periosteal sclerosis, and growing intracranially as well as extracranially. MRI showed a highly enhancing tumor, dislocating the superior, transverse and straight sinuses anteriorly and inferiorly (Fig 1).

Operation – In order to prevent excessive bleeding in a patient 4 months-old, the resection was performed in three stages, this meaning that removal of the tumor

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was staged in three procedures. The patient was in the prone position, and a semilunar incision was used. The tumor was extremely adherent to the dura-mater and the superior sagittal sinus, with petrous consistency and a rocky appearance (Fig 2). With the aid of the ultrasonic aspirator, the tumor was almost completely resected, except for a small layer covering the posterior portion of the superior sagittal sinus. We were concerned that sinus thrombosis might occur after excessive surgical manipulation.

Postoperative course – The pathological findings and immunohistochemical studies were suggestive of a primitive benign lesion with a final diagnosis of neuroectodermic melanotic tumor (Fig 3). Immunohistochemical studies showed positivity for sinaptofisin, epithelial membrane antigen (EMA1+EMA3) and monoclonal anti-melanin antibody (HMB-45), and negativity for S-100 and glial fibrillary acidic protein. A CSF fistula developed after the second operation, and was followed by meningitis: a lumbar drain was left in place for 5 days, and vancomycin was given for 14 days, with complete resolution of symptoms. Currently, two years after presentation, the patient is neurologically normal with no evidence of tumor recurrence.

The presentation of this case was authorized by the baby's mother and also the Ethics Committee of the Hospital was previously notified.

DISCUSSION

First described by Krompecher in 1918¹, the MNTI is a rare tumor, and approximately 200 cases have been reported²⁻⁸. It is usually benign, though locally aggressive and fast growing. Most of the patients are infants, although a few adult patients have been reported. Male to female ratio is almost equal (6:7) and the mean age of the patients is 4.3 months. Of the reported tumors, 68.6% are located in the maxilla, followed by the skull (10.6%), mandible (7.3%), intracranial structures (5.3%), epididymis (4%), soft tissues, uterus and mediastinum⁹.

Computerized tomography scan with bone windows reveals usually an expansive mass with periosteal sclerotic, well-demarcated, hyperdense mass with contrast enhancement. Magnetic resonance image T1-weighted images reveal an isointense mass homogeneously enhanced by gadolinium and a low-intensity lesion on T2-weighted images. The adherence of the lesion to the dural sinuses is best shown in magnetic resonance angiography¹⁰.

The tumor is usually moderately vascularized, highly pigmented, solid, with fibrous connective

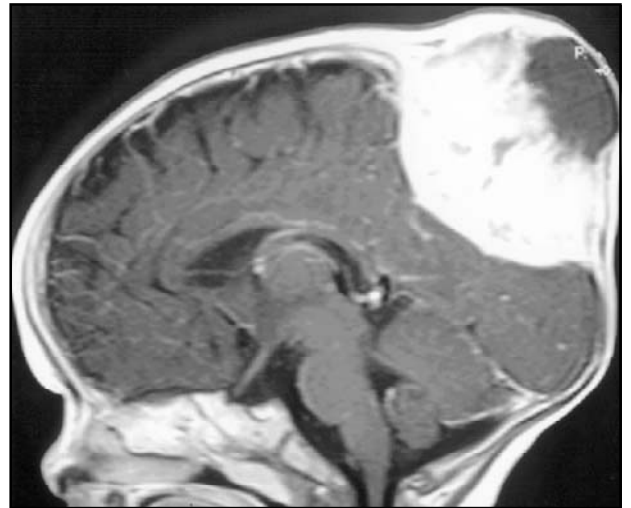


Fig 1. MRI shows a highly enhancing tumor displacing the superior, transverse and straight sinus anteriorly and inferiorly.

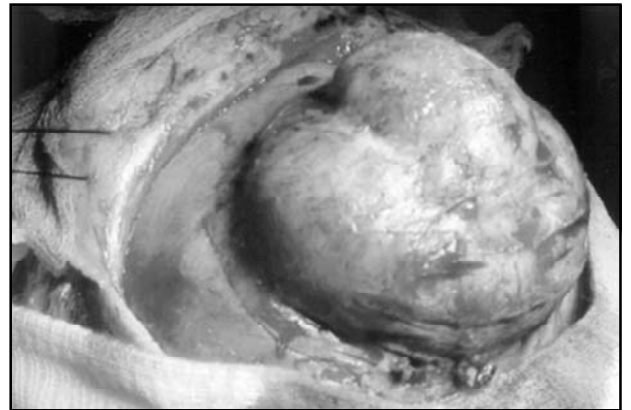


Fig 2. Transoperative photograph, first operation.



Fig 3. Pathological examination showing hyperchromatic nucleus, slit-like alveoli and hyperchromatic nuclei. Photomicrography, hematoxylin and eosin; X100.

tissue, and some fine calcifications inside it. There are usually three different types of cells: a large, cuboidal, epithelial-like cell with abundant, vacuolated cytoplasm and a large ovoid sharply margined nucleus; the second one is small and immature with hyperchromatic nucleus and nonpigmented cytoplasm; the third is also small, but stellate, resembling a fibroblast¹¹. The tumor cells are divided into islands with slit-like alveoli, which are irregular in shape and size¹². Mitoses, pleomorphism, and other findings of anaplasia are not seen in this kind of tumor.

Cytologic diagnosis of MNTI can be obtained. Aspirating from the lesion are usually cellular, with a bimodal population mainly of small neuroblast-like cells admixed with a few large epithelioid cells with melanin granules¹³. Immunohistochemical studies are of great help in the diagnosis of MNTI: they are usually positive for cytokeratin, vimentin, epithelial membrane antigen, S-100, glial fibrillary acidic protein and NSE.

Electron microscopic and histochemical studies have now established that these tumors probably originate from neural crest cells because of morphological similarities between tumor cells and melanocytes and between tumor cells and neuroblasts¹⁴. There are reports of high serum levels of adrenaline, noradrenaline and vanillylmandelic acid that further corroborate this hypothesis.

Because MNTI is locally aggressive, the treatment of choice is primary surgical resection. If the tumor occurs in a favorable location, surgery can be curative. Surgical treatment is more difficult for tumors in unfavorable locations, particularly those along the midline and at the cranial base: some of these lesions are particularly problematic, whe-

re there is the possibility of involvement of dural venous sinuses, and also those with significant intracranial extension¹⁵. In such patients, surgery carries a higher risk. When surgery is not a consideration, chemotherapy may be helpful^{11,16}.

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