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Neuroblastoma Craniocerebral Metastases (A Case Report)

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Introduction

Neuroblastoma is a solid embryonal tumor that is very common in pediatrics. It originates from sympathetic tissue derived from neural crest cells. Imaging plays an important role in the management of patients with neuroblastoma and is essential to assess the metastatic extension. Neuroblastoma Metastases are very frequent, secondary craniocerebral neuroblastoma being among the most dreadful metastases. We report the case of a 7-year-old child with metastatic abdominal neuroblastoma who presented secondary craniocerebral neuroblastoma with cranial vault, orbital and leptomeningeal metastases.

Observation

A 7-year-old male child followed for metastatic abdominal neuroblastoma treated with chemotherapy (Figures 1-3). A cerebral CT scan without and with injection carried out as part of an extension workup revealed bilateral fronto-parietal and right occipital epidural tissue deposits (Figure 2A), giving rise to a diastasis of the sutures with an irregular appearance of its margins (Figure 2C, 3), heterogeneously enhanced after injection of the iodinated contrast product, associated with a grass fire periosteal reaction (Figure 1C), more marked at the frontal bone, invading bilaterally, more marked on the right, the jugal and prepalpebral soft tissues, the lateral rectus muscles and the lacrimal glands (Figure 2B), with thickening and diffuse contrast of the leptomeninges without any parenchymal lesion at the supratentorial and subtentorial levels, nor of the cisterno ventricular system. This CT scan also revealed bi-parietal bone thickening (Figure 1A) diffuse secondary osteolytic bone lesions at the level of the cranial vault and the base of the skull (Figure 1B).

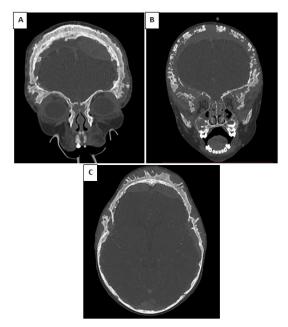


Figure 1: Cerebral CT scan without contrast injection (Bone window): **(A)** coronal section: bi-parietal bone thickening. **(B)** Coronal section: diffuse osteolytic bone lesions at the level of the cranial vault and the base of the skull. **(C)** Axial section: Bifrontal grass fire periosteal reaction.

Discussion

Neuroblastoma is the 3rd most frequent malignant tumor in pediatrics [1,2] and affects mostly children under 5 years of age [3]. It is a malignant embryonal tumor of the sympathetic nervous system derived from neural crest cells [4] and is metastatic in 70% of cases. MRI and CT are the tests of choice for detecting craniocerebral metastases of primary extra cerebral neuroblastoma [5]. In our case, the patient only had a cerebral scan as part of the extension workup. Secondary craniocerebral neuroblastoma is most often manifested as bone metastasis, which represents 25% of neuroblastoma metastases [1]. They mainly involve the cranial vault, which can be the site of bone thickening, hair-on-end periosteal reaction and lytic lesions with extension to the epidural space in the form of epidural deposits [1,6].

Our patient presents with epidural deposits, bi-parietal bone

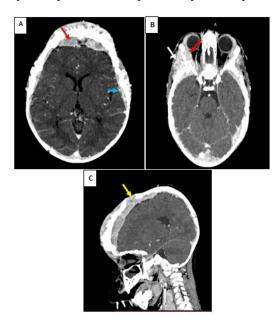


Figure 2: Cerebral CT scan with injection (Parenchymal window): (**A**) Axial section: frontal epidural tissue deposits (red arrow) with leptomeningeal enhancement (blue arrow). (**B**) Axial section: Infiltration of the jugal soft tissues (white arrow) and lateral rectus muscles (red arrow). (**C**) Sagittal section: Epidural deposits reaching the vertex and insinuating between the margins of the coronal suture (yellow arrow).

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Figure 3: 3D section: Diastasis with irregular appearance of the margins of the cranial vault sutures.

thickening, and diffuse secondary osteolytic bone lesions at the level of the vault bones and the base of the skull with a hair-on-end periosteal reaction.

The epidural deposits are inserted between the sutures and lead to a diastasis of the latter with an eroded and irregular aspect of its margins [1] which is the case for our patient.

Orbital metastases of neuroblastoma are present in 10 to 20% of cases [6] often in the form of an extra conical orbital mass [6] which manifests as exophthalmos, periorbital ecchymosis [6] with risk of blindness due to optic nerve compression [7].

Central nervous system metastases in children with neuroblastoma are rare (8% at 3 years of age) [1], and are often detected during recurrence [1,8], Involvement of the brain parenchyma is rare, representing 2 to 16%, and is mainly located above the tentorial region [1]. Cerebral metastases are often of the hemorrhagic type (70% of cases) without perilesional edema, sometimes associated with calcifications [8,9], they may also be of the cystic type, in which case they are secondary to hemorrhagic necrosis or to a rupture of the bloodbrain barrier with exudation of plasma proteins [8] Intra ventricular metastases are exceptional, they are often of hemorrhagic type [8]. In our patient, there was bilateral invasion of the prepalbebral soft tissues, lateral rectus muscles and lacrimal glands, with no extra conical orbital mass, nor parenchymal or intra ventricular metastases.

Leptomeningeal metastases are rare except in patients with disseminated involvement [1,2], they have a poor prognosis with a survival rate of less than 6 months [8], metastases in the dura are

often associated with bone metastases [1]. The radiological appearance of leptomeningeal metastases is nonspecific; they are manifested by diffuse or localized linear or nodular enhancement [8], with diffusion restriction of non-hemorrhagic leptomeningeal nodular lesions [9]. Epidural and leptomeningeal extension is best characterized by MRI. Our patient presents with thickening and diffuse contrast of the leptomeninges. The differential diagnosis of craniocerebral metastases arises primarily in the face of lytic bone lesions secondary to langerhans cell histiocytosis, leukemia, lymphoma, and sarcoma metastases [1]. For parenchymal metastases, the main differential diagnoses are hematoma [10], oligodendroglioma, astrocytoma, ependymoma, and other primitive neuroectodermal tumors [1]. Most leptomeningeal metastases are associated with medulloblastoma, ependymoma and pineal gland tumors [1]. Diastasis of the sutures may also be secondary to intracranial hypertension, but in this case the margins of the sutures are intact [1].

Conclusion

Neuroblastoma is a frequent malignant tumor in pediatrics. Craniocerebral metastases of neuroblastoma are dreadful and most often occur during recurrence. CT and MRI are the key examinations for detecting most intracranial and craniofacial metastases.

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