

Malignant Fibrous Histiocytoma of the Brain in the Pediatric Patient: A Case Report and Literature Review

Histiocitoma fibroso maligno cerebral en paciente pediátrico: Presentación de un caso y revisión de la literatura



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Summary

Brain tumours are the second cause of malignant neoplasms in children while primary sarcomas in the central nervous system (SNC), are extremely rare. For this reason, we present the results of the study of a case of a girl and revision on the pathophysiology, clinical manifestations, and findings on brain MRI of this pathology. We report the case of a seven-year-old patient admitted to the emergency department with headache and symptoms suggestive of intracranial hypertension, and decreased strength of the right side of the body. In brain MRI, we found a left temporoparietal extra-axial mass, displacing the midline. The patient was examined by the neurosurgeon who performed a left frontoparietal craniotomy, with complete tumor resection. The pathology study showed high-grade spindle cell sarcoma, classified as malignant fibrous histiocytoma.

Resumen

Los tumores cerebrales son la segunda causa de neoplasias malignas en niños y los sarcomas primarios en el sistema nervioso central son extremadamente raros. Por esta razón, presentamos los resultados del estudio de un caso en una menor de edad y la revisión realizada sobre la fisiopatología, manifestaciones clínicas y hallazgos en las imágenes de resonancia magnética cerebral de esta patología. Se trata de una niña de 7 años de edad que ingresa al servicio de urgencias con cefalea, sintomatología sugerente de hipertensión endocraneana y disminución de la fuerza del hemicuerpo derecho, por lo que se le practica resonancia magnética cerebral (RM) con medio de contraste, con hallazgo de masa extraaxial temporoparietal izquierda, que desplaza la línea media. La paciente es valorada por neurocirugía y remitida a craneotomía frontoparietal izquierda, con resección completa del tumor. El informe de patología muestra sarcoma fusocelular de alto grado, compatible con histiocitoma fibroso maligno.

Introduction

Brain tumors account for between 16% and 23% of all tumors in the pediatric age (1) and are the second cause of childhood cancer, with a homogeneous distribution from birth to 14 years of

age (2). Sarcomas represent approximately 6% of all cancers in this age group (1); however, primary brain sarcomas are extremely rare neoplasms, with an estimated incidence of 0.1 - 4% of all intracranial neoplasms (1). The term malignant fibrous histio-

cytoma (MFH) was first coined in 1963 by Ozzello. These tumors compromise usually the limbs and the retroperitoneum, and intracranial presentation is very rare, with very few cases described in the literature (3). Magnetic resonance imaging (MRI) is the study of choice for the diagnosis of intracranial tumors, surgical planning and post-treatment control (4, 5). Histopathologically there are more than fifty soft tissue sarcomas subtypes, according to the classification of the World Health Organization (WHO) (6), for which it is essential an adequate treatment to clearly identify the type of lesion, through the use of biomolecular techniques and the identification of the tumor genome (7).

Clinical history

Seven-year-old girl who entered the emergency department due to multiple episodes of emesis with a week of evolution, frontal headache and loss of strength of the right hemiface, ipsilateral epiphora, previously medicated with antiemetics and analgesics without improvement. Physical examination revealed a deviation of the left corner of the lips, decreased strength of the right hemiface and limitation for ipsilateral palpebral occlusion. We also found a decrease in strength in the right hemibody 4/5, which limited standing and wandering. Negative Romberg sign. MRI of the brain with contrast medium showed a left temporoparietal extra axial mass and a subphalpine hernia that displaced the midline to the right (figures 1, 2, 3 and 4). By endocranial hypertension clinic and Neurosurgery decision, a left frontoparietal craniotomy was performed with complete resection of the lesion, which resulted in partial improvement of the symptomatology in the immediate postoperative period.

The pathology report reveals malignant neoplasm of fusocellular pattern with elongated hyperchromatic nuclei, some rounded, multilobulated, with frequent mitotic figures (up to 50 per 10 CAP) and foci of necrosis (figure 5a). Adjacent gray matter rejected by the lesion is scarce and adequate circumscription between the tumor and the adjacent parenchyma is observed (figure 5b). Cells are arranged in bundles and some foci show cells with large pleomorphic eosinophil cytoplasm (figure 6). Positivity of tumor cells to vimentin (figure 7a), focally to desmin in pleomorphic cells (figure 7c) and negativity for acute myeloid leukemia (AML), epithelial membrane antigen (EMA), myogenin and glial fibrillary acidic protein (GFAP). The cell proliferation index (Ki 67) is of 90% (figure 7b). The INI1 is preserved. This immunohistochemical profile together with the architecture of the lesion in which marked cellular atypia is observed, with necrosis and high mitotic count allows to diagnose high-grade fusocellular neoplasia with pleomorphic sarcoma type compatible with malignant fibrous histiocytoma.

In the control MRI, postoperative changes are evidenced with tumor residue, which is why the chemotherapy cycle begins with vincristine, phosphamide, cisplatin, etoposide, and radiation therapy. Clinically, the patient manifests complete recovery of the neurological state. In the perfusion control and MRI spectroscopy at 2, 9, 12 and 19 months post-treatment there is no tumor recurrence observed.

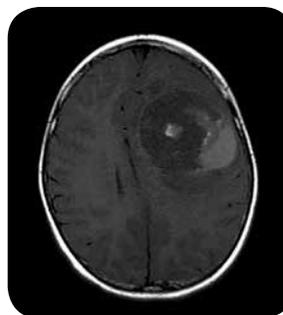


Figure 1. MRI with T1, axial information. In the left frontoparietal region the extra-axial mass of defined contours is observed, predominantly low signal and some high signal foci that correspond to intratumoral bleeding, displacing the adjacent parenchyma, it is associated with subphalpine herniation to the right with obliteration of the frontal horns of the lateral ventricles.



Figure 2. a) Axial; b) coronal and c) axial FLAIR sequence. Mass of heterogeneous signal with multiple cystic foci and important associated vasogenic edema.

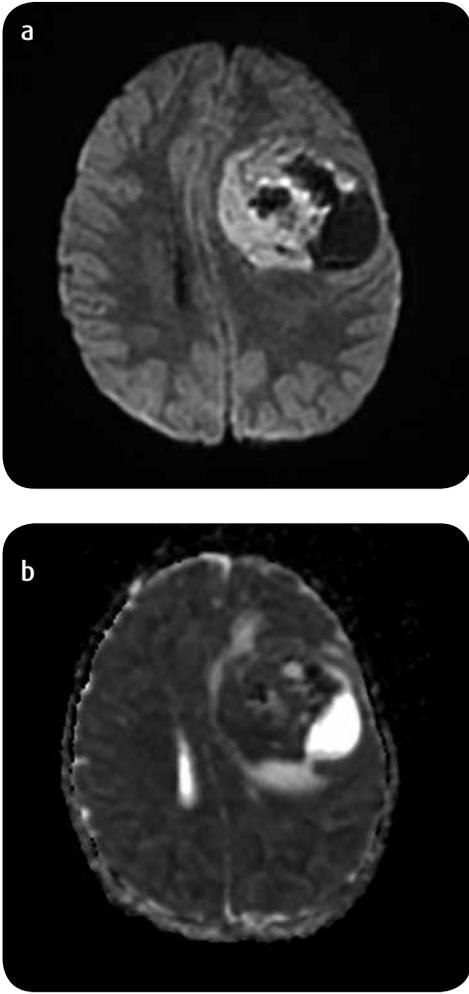


Figure 3. a) Diffusion sequence and b) ADC Map. High signal of the diffusion lesion, with low signal of the same in the ADC map, which corresponds to restriction by high cellularity.

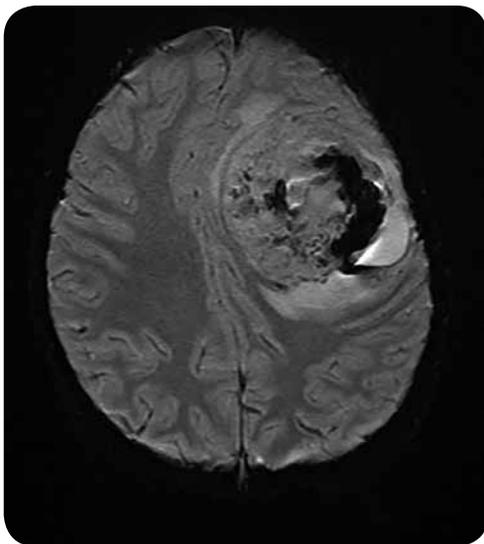


Figure 4. Echo gradient sequence (magnetic susceptibility). Areas of low intratumoral signal, which correspond to areas of hemorrhage.

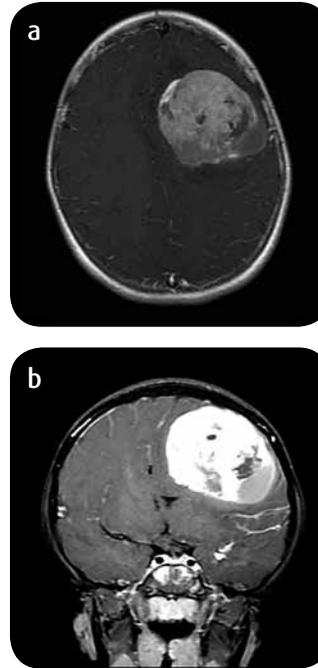


Figure 5. a) Axial sequences with T1 information with contrast medium, and b) coronal. Enhancement intensity of the lesion with the contrast medium.

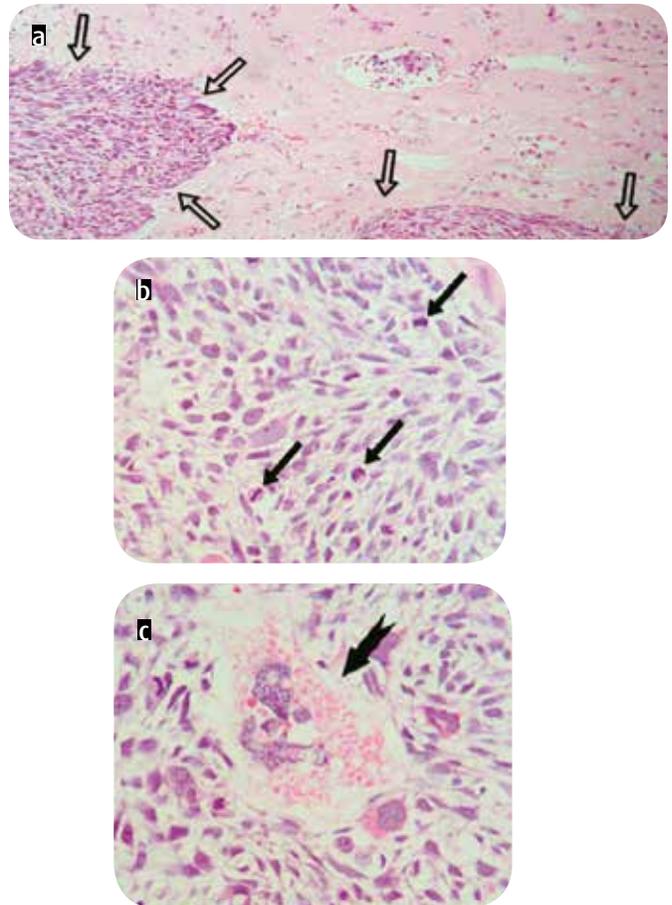


Figure 6. a) Transition between tumor and adjacent normal cerebral parenchyma (circumscribed edge, hollow arrows). b) Frequent mitotic figures (arrows). c) Zoom (40X), cell is observed with pleomorphic nucleus and atypical (arrow with notch).

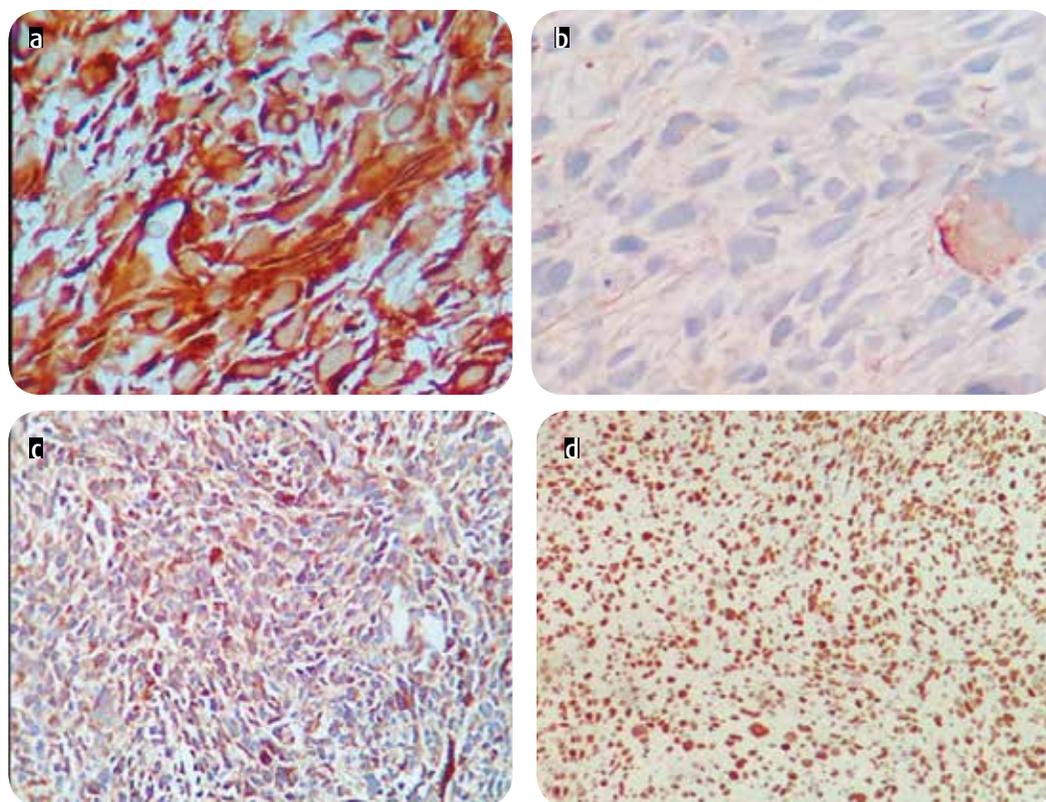


Figure 7. a) Vimentin, b) BCL2, c) desmin, d) Ki67.

Discussion

Soft tissue sarcomas are mesenchymal tumors, originating from mesoderm cells (5). Its primary origin in the central nervous system is extremely rare, with a few case studies over the past 50 years, but with varying histology and derivatives of the meninges mainly, hence it appears as an extra-axial lesion (8). Descriptions have been found of MFH as: pleomorphic sarcoma or sarcoma without other specification (NOS), rhabdomyosarcoma, chondrosarcoma, myxoid liposarcoma and malignant mesenchymal tumor (1). Supratentorial tumors are more common neonates and infants, while in children of more than 2 years of age infratentorial involvement is more common (5). According to the histological type, the tumor of this patient is classified as pleomorphic sarcoma.

Initial descriptions of this tumor were made in the 1960s and 1970s: lesions that compromised the Turkish saddle and the temporal bone with intracranial extension as a result of radiation therapy in pituitary tumors (9, 10). This type of tumor has two peaks of incidence, below 10 and at 40 years of age, with predominance in men (11). Some predisposing factors have been proposed for its development, such as trauma, radiotherapy or intracranial surgery for other causes (11).

Computed axial tomography (CT) may show an extra-axial lesion rejecting the adjacent cerebral parenchyma, with heterogeneous density, defined contours, with some areas of high signal due to intratumoral hemorrhage and low cystic areas and necrosis, as well as peritumoral vasogenic edema and enhancement

after administration of contrast medium (12). In MRI, the lesion shows regular contours, broad dural base and low signal in the white matter in T1, and intermediate signal to slightly high in T2 with some cystic areas and surrounding vasogenic edema. It reflects a high diffusion signal with low ADC values, intense contrast enhancement and loss of intratumoral signal in the sequence of susceptibility due to hemorrhage. Due to its mass effect, it may obliterate the ventricular system and be associated with subphalcaline herniation (8,12). CT-fused positron emission tomography (PET-CT) shows a high uptake of fluorodeoxyglucose (FDG) solely by mass; however, this finding is not specific and may occur in other types of central nervous system (CNS) malignant tumors (12). The treatment is mainly aimed at the surgical resection of the tumor with removal of wide margins, to avoid local recurrence of the tumor, and radium and chemo as adjuvant therapies. Chemotherapy, on its own, has no definite effectiveness. The prognosis depends on the affected area, tumor extension and cellular atypicity (3,12).

In the case studied, a solid left frontoparietal mass was observed, with cystic areas, homogeneous enhancement and surrounding vasogenic edema, which by its characteristics and histological study was compatible with high-grade malignant fibrohistiocytoma (9). The differential diagnosis should be done with meningioma, gliosarcoma, Schwannoma, pleomorphic xanthoastrocitoma, malignant gliomas, lymphoma, and metastasis (3,10,12), hence the importance of the histological correlation and biomarkers for classification.

Conclusion

Brain sarcomas are a rare entity in the general population and more in the pediatric age, for which there are few series of cases published in this age group. Malignant fibrous histiocytoma or primary pleomorphic sarcoma in children is even rarer, so there are few descriptions in the literature and, although their long term prognosis is bad, in the patient in this case there has been no observed tumor recurrence so far.

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