PEDIATRIC RHABDOID MENINGIOMA

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CLINICAL HISTORY

A 3-year-old boy, with a recent history of seizures, was admitted to the Neurosurgery Unit of the Meyer Children’s Hospital. He was the full-term result of a normal pregnancy. His weight was 17 kilograms and his cranial circumference was 50 cm. There were no neurological focal deficits or signs and symptoms of cranial hypertension. Neurocutaneous stigmata were absent. Computed tomography (CT) scan and magnetic resonance imaging (MRI) showed a right fronto-temporal inhomogeneous, hypervascularized and calcified mass measuring about 4 cm in diameter. It involved the orbital fissure and the lesser sphenoidal wing and entirely occupied the temporal fossa incorporating the ipsilateral middle cerebral artery. The lesion determined mass effect with consequent midline shift and was accompanied by basal hyperostosis. A right pterional craniotomy was performed. During the surgery, the mass, which arose from the meninges, appeared not well demarcated from the perilesional parenchyma particularly in the sylvian area. Post surgical radiological studies documented a sub-total excision. Due to the young age of the patient he was treated with chemotherapy. The lesion remained stable after 18 months of follow-up.

PATHOLOGICAL FINDINGS

Surgical specimen was fixed in 10% buffered neutral formalin and embedded in paraffin. Some 5 μm sections were stained with hematoxylin-eosin (H&E) for the morphological evaluation whereas further 5 μm sections of the most representative specimen were mounted on electrostatic slides and used for the immunohistochemical study. Immunohistochemical stains were performed by standard avidin-biotin complex immunoperoxidase method. The antibodies used were directed against epithelial membrane antigen (EMA), vimentin (VIM), glial fibrillary acidic protein (GFAP), S-100 protein, synaptophysin (SP), neurofilaments (NF), neuron specific enolase (NSE), actin (1A4,
HHF35), desmin, myogenin, myosin, cytokeratins (AE1/AE3, CAM5.2), estrogen and progesterone receptors (ER, PgR), BAF47 (INI-1), p53 protein, and Ki-67 (Mib-1).

The lesion was mainly composed of a proliferation of globoid cells showing an eosinophilic ample hyaline cytoplasm with peripheral nuclei, prominent nucleoli and occasional intranuclear cytoplasmic pseudoinclusion. The cells appeared in many areas rather loosely arranged and sometimes disclose a papillar architecture. In some areas, the lesion was constituted of spindle-shaped cells. Focally, an organoid architectural pattern was also observed (isles of spindle-shaped or globoid cells delimited by smaller cuboidal cells). There were no whorls or psammoma bodies. Interstitial space contained collagen bundles. Mitoses were rare. Hemorrhages and vascular hyperplasia were evident. Focal areas of necrosis were also present.

By immunohistochemical staining, the tumor cells were EMA, VIM and p53 positive and GFAP, SP, NF, NSE, 1A4, desmin, myogenin, myosin, AE1/AE3 and CAM5.2 negative. S-100, HHF35 and progesterone receptors were focally positive while estrogen receptors were negative. Furthermore, the tumor cells exhibited diffuse INI-1 nuclear staining. The proliferation index, as determined estimating the percentage of the Ki-67 positive neoplastic cells in the total tumoral cells in the most positive areas, was 15%.

**FINAL DIAGNOSIS**

Morphologic features along with immunohistochemical results were consistent with the diagnosis of rhabdoid meningioma.

**DISCUSSION**

Meningiomas are frequent primary intracranial neoplasms (about the 25% of all primary tumors in this site) arising from the leptomeningeal covering of the central nervous system and preferentially affecting middle aged and elderly women. Radiation exposure, hormonal and genetic factors have been implicated in their development and growth. Meningiomas are usually sporadic, but may be
also a manifestation of the hereditary syndrome neurofibromatosis type 2 (NF2), which is characterized, at the nervous system level, by the development of bilateral vestibular schwannomas, meningiomas, ependymomas, and, occasionally, gliomas and neurofibromas. The World Health Organization (WHO) recognizes 3 histological grades (WHO I or benign, WHO II or atypical and WHO III or anaplastic) with an increasing risk of recurrence and of unfavorable outcome. Recurrence rates of 7%-20%, 29%-40% and 50%-70% are reported respectively for benign WHO I, atypical WHO II and anaplastic WHO III meningiomas. Meningiomas exhibit a wide range of histological patterns with numerous classified subtype and several uncategorized subtypes. In most cases histological variant do not have prognostic significance. However, clear cell, chordoid, papillary, and rhabdoid meningiomas are clinically aggressive. ¹

Meningiomas are rare in pediatric population. Indeed, although tumors of the CNS are the second most common neoplasms in children, intracranial meningiomas account for only 1% to 4.2% of all brain tumors in this age group. Affected children often have NF2 syndrome or a history of prior skull irradiation. ²

Rhabdoid meningioma is an uncommon meningioma variant. It is categorized as WHO grade III. The majority of the cases occurred in adulthood. In fact only about 10 juvenile cases have been reported in the international literature. The outcome of the reported juvenile rhabdoid meningiomas seems to be better than their adult counterparts. However, adjuvant therapy is essential in the management of the pediatric as well as of the adult cases. ³, ⁴, ⁵, ⁶

The observation in the pediatric age of a central nervous system tumor showing rhabdoid features mainly evocate the possible diagnosis of atypical teratoid/rhabdoid tumor. It is a highly malignant embryonal tumor mostly affecting infants. Atypical teratoid/rhabdoid tumor typically has INI-1 gene mutations which are considered its molecular stigmata. ¹
In our case, considering the young age of the patient the possibility of an atypical teratoid/rhabdoid tumor was taken in consideration. Nevertheless, the immunohistochemical diffuse INI-1 nuclear positivity of the neoplastic cells excluded this possibility. On the other hand, the tumor was completely negative to the glial and neuronal markers which are variably positive in atypical teratoid/rhabdoid tumor.¹

In conclusion, the diagnosis of this tumor subtype, given its prognostic implications, must also be considered in pediatric patients.

REFERENCES


