Supratentorial Cortical Ependymoma in a 21-Month-Old Boy

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Two-thirds of ependymomas arise in the infratentorial or intraventricles, whereas one-third are located supratentorially. But supratentorial “cortical” ependymomas are very rare. We report a case of a cortical ependymoma in a 21-month-old boy. The patient presented with simple partial seizures. This tumor was located in the postcentral gyrus and he had gross total excision. Microscopy and immunohistochemistry showed grade II differentiation ependymoma.

Key Words: Ependymoma · Pediatrics · Supratentorial neoplasm · Immunohistochemistry.

INTRODUCTION

Ependymomas are rare neuroectodermal tumors arising from ependymal cells of the ventricular system, choroid plexus, filum terminale, or central canal of the spinal cord. Ependymomas are relatively rare nervous system tumors, constituting 1.2% to 7.8% of all intracranial neoplasms or 2% to 6% of all gliomas49. Ependymomas are frequently infratentorial, and a third of ependymomas are supratentorial. Supratentorial ependymomas outside the ventricular system as a distinct location are infrequent. As reported in some articles, ectopic ependymoma can arise in the supratentorial parenchyma with no attachment to the ventricular system9. However, these are very rare and reported in only a few cases in the literature9. Our patient was only 21 months old, which to our knowledge, is the youngest pediatric case of supratentorial ectopic cortical ependymoma in published papers. Herein, the clinical features and pathologic findings of supratentorial ectopic cortical ependymoma in are described.

CASE REPORT

History and examination

A 21-month-old boy presented with repetitive left leg rigidity lasting two weeks. At birth he was delivered vaginally without complications, and did not have any remarkable history, including no history of headache or body weakness. The child did not show abnormalities in the neurologic examinations and the sensori-motor and gait assessments were also normal. He did present with simple partial seizure activity with clonic movement at the left lower extremity. The seizure episodes had been occurring an average of eight times per day and lasting 30-60 seconds. He did not lose consciousness during seizure attacks.

Radiological examination

Brain magnetic resonance (MR) revealed a 1.5×1.5 cm diffuse enhancing mass in the right postcentral gyrus cortex with some focal calcifications. Magnetic resonance imaging (MRI) revealed hyperintensity on T1-weighted images and mixed hypo- to hypertensity on T2-weighted images. The mass had relatively smooth margins and only minimal brain swelling was noted around the peri-mass lesion. On the sagittal image the mass was closely contacted with dura of the sagittal sinus and falx. There was low signal line between the mass and dura, showing the presence of cerebro-spinal fluid. Imaging showed no evidence that the mass was connected to the ventricular ependymal lining (Fig. 1).

The neuro-radiologic differential diagnosis included dysembryoplastic neuroepithelial tumor or low grade glioma, and meningioma less likely, due to the parasagittal tumor location.

Operation and pathological findings

A right parietal craniotomy was performed and complete excision of the tumor was done. The tumor was firm, gray, and contained calcifications. It had well defined margins, allowing
Post-operative course and follow-up

The patient made a good recovery and had no neurologic deficiency. He did not have any seizure episodes after the surgery. Follow-up brain computed tomography one day post-operation and brain MR images 10 days after the surgery revealed no evidence of residual lesions, and MR imaging of the whole spine showed no evidence of leptomeningeal seeding. Adjuvant chemotherapy or radiotherapy was not considered because a gross total resection was done and the tumor did not have anaplastic clear planes of dissection. Because location of the mass was an eloquent area, we carefully dissected the mass wall from the adjacent gyri and resected the mass en bloc.

Histopathology showed spindle shaped ependymal cells. These cells formed perivascular rosettes and were positive for glial fibrillary acidic protein (GFAP) (Fig. 2). The proliferation index was 5-10% by Ki-67 immuno-staining. The pathologic diagnosis was confirmed as ependymoma, World Health Organization (WHO) grade II.

Fig. 1. Pre-operative brain MR image. Brain MR revealed a 1.5×1.5 cm diffuse enhancing mass in the right postcentral gyrus cortex with some focal calcifications. MRI reveals hyperintensity on T1-weighted images and mixed hypo- to hypointensity on T2-weighted images. The mass shows relatively smooth margins and only minimal brain swelling is noted around peri-mass lesion. On the sagittal image the mass is closely contacted with dura of the sagittal sinus and falx. There is low signal line between the mass and dura, showing the presence of cerebro-spinal fluid. Imaging shows no evidence the mass is connected to the ventricular ependymal lining. (A : Sagittal T1 weighted image, B : sagittal T1 weighted image with enhancement, C : coronal T1 weighted image with enhancement, D : axial T1 weighted image with enhancement). CT : computed tomography, MR : magnetic resonance.

Fig. 2. Histologic examination. A : Hematoxylin-Eosin (×400); an ependymal rosette (a canal with irregular or ciliated lumen surrounded by nuclei arranging like rosette) is seen at the center. This feature is characteristic to ependymoma. At the right side of the rosette, some calcifications (purple round or irregular figured bodies) are seen. Among the nuclei (purple round), many fine fibers are seen. B : Hematoxylin-Eosin (×200); large purple bodies are calcifications. The white spaces without cells are an artifact. Calcified region may become vacant space through the cutting process from the paraffin blocs. C : Glial Fibrillary Acidic Protein (GFAP) (×200); the fine fibers are positive for GFAP, which means they are of neuronal origin (astrocyte).

Fig. 3. Post-operative follow-up brain MR image. Follow-up brain MR was taken at 12 months post-operation. There is no evidence of tumor recurrence. Only a small, clean, empty cavity is noted at the tumor removal site (arrow, image A). A : Sagittal T1 weighted image. B : sagittal T1 weighted image with enhancement. C : coronal T1 weighted image with enhancement. D : axial T1 weighted image with enhancement.
features in the histopathologic examination.

Follow-up brain MR and whole spine MRI were taken at 6 and 12 months post-operation and showed no evidence of tumor recurrence (Fig. 3).

DISCUSSION

Ependymomas are usually known to arise from ependymal cells of the ventricular system, choroid plexus, filum terminale, or central canal of the spinal cord. The clinical and pathological characteristics of supratentorial ependymomas in children are not well identified in the literature, because most series deal with ependymomas regardless of their location or age of the patient. Extraventricular ependymomas are especially rare, and while they have definite ependymal morphology, their histogenesis remains uncertain. However, in some articles the pathogenesis of supratentorial, extraventricular, cortical, and ectopic ependymoma were explained in other ways. Vernet et al. hypothesized the following pathogenesis for supratentorial ectopic ependymomas: 1) The tumors develop from intraparenchymal or subarachnoid ependymal cysts resulting from disorders of migration from the germinal matrix; 2) The tumors represent primitive neuroectodermal tumors that have differentiated extensively along the ependymal lineage; 3) The tumors might be the result of neoplastic growth within an ectopic ependymal cell and are therefore, at least in part, the consequence of a migrational error.

Pathologic findings

In our case, we identified several pathologic findings such as round to oval nuclei with evenly dispersed stippled chromatin, perivascular pseudorosettes, true ependymal rosettes, and frequent immunoreactivity for GFAP—the latter often in a perinuclear dot and/or ring pattern. These are typical and classic cellular features of ependymoma.

Ependymal tumors do not characteristically infiltrate surrounding normal parenchymal tissue, but Norman reported pathologic findings of six cases of ependymomas infiltrating along axonal tracts, perineuronal satellitosis, and subpial molding. Additionally, all six cases showed infiltrating individual tumor cells in the parenchyma and non-rosetting angiocentric spread along small cerebral blood vessels. However, unlike in “diffusely infiltrative” gliomas, the infiltration demonstrated by supratentorial ependymal tumors appeared to be mostly locally infiltrative, confined to areas at the periphery of the main tumor masses.

In our case, the proliferation index was 5-10% by Ki-67 immunostaining with no evidence of an anaplastic component. The pathologic diagnosis was confirmed as ependymoma, WHO grade II. In adult ependymoma patients, the majority of supratentorial lesions are classified as WHO grade III. However, roughly 70% of all ependymomas diagnosed in the pediatric population are histologically benign and are classified as WHO grade II; less than 2% are considered WHO grade I or subependymoma, and the remainder are classified as WHO grade III or the anaplastic variant.

Treatment and prognosis

There has been much debate about the usage of post-operative radiotherapy for treatment of supratentorial extraventricular ependymomas. Roncaroli et al. concluded that cortically based low-grade supratentorial ependymomas should be treated with surgery alone, given that they are amenable to gross total resection and are not prone to local recurrence or leptomeningeal spread. The researchers concluded that post-operative radiotherapy did not play a role in treatment. However, in another recent study of supratentorial ependymomas, it has been suggested that radiotherapy should be given to patients with anaplastic ependymomas and in cases where only partial resection of either benign or malignant tumors has been achieved.

As mentioned previously, ependymomas can infiltrate surrounding normal tissue, so incomplete removal of the tumor during surgery may lead to a bad prognosis. As a result, infiltrating supratentorial ependymal tumors have sometimes previously been generically labeled as “infiltrating gliomas”. Because the most important prognostic factor for ependymal tumors is extent of resection, it is essential to alert clinicians to the ependymal nature of these neoplasms so attempts at gross total resection can be made whenever possible. Lastly, recognition of the potential infiltrative nature of supratentorial ependymal tumors may have important implications for future tumor classification and prediction of its biological behavior and response to adjunctive treatment.

In some articles, not only the extent of tumor resection, but also proliferative index have been statistically shown to be important independent prognostic indicators in intracranial ependymomas. But in the case of supratentorial ependymal tumors, proliferative index may not be as important as an independent prognostic factor, particularly in tumors limited to the cortex and immediately underlying white matter.

In the pediatric population of supratentorial extraventricular cortical ependymoma, gross total resection appears to be the best prognostic indicator of long-term survival in both WHO grade II and WHO grade III lesions.

CONCLUSION

In this article, we report a case of very young patient with supratentorial cortical ependymoma and its clinical manifestation. To make a proper differential diagnosis on supratentorial cortical mass lesions in a pediatric patient, even one regarded as too young, ependymoma should be considered as a differential diagnosis. The total removal of ependymoma in children is especially important because if the surgeon is successful in completely removing the tumor, adjuvant therapy such as radiotherapy or chemotherapy are not required.
References