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Large Supratentorial Cortical Ependymoma in a Child

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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 uncommon nervous system tumors, constituting 1.2% to 7.8% of all intracranial neoplasms or 2% to 6% of all gliomas.[1-8] Extraventricular ependymomas are especially rare, and whereas they have definite ependymal morphology, their histogenesis remains uncertain. Some tumors may extend to the gray matter reaching the pial surface, but pure cortical ependymomas are uncommon. We have encountered a few pediatric cases of supratentorial cortical ependymoma in the English literature.[1-10]

We present a case of an 11-year-old boy with a large supratentorial cortical ependymoma with massive calcification and central cyst formation manifested as generalized convulsion.

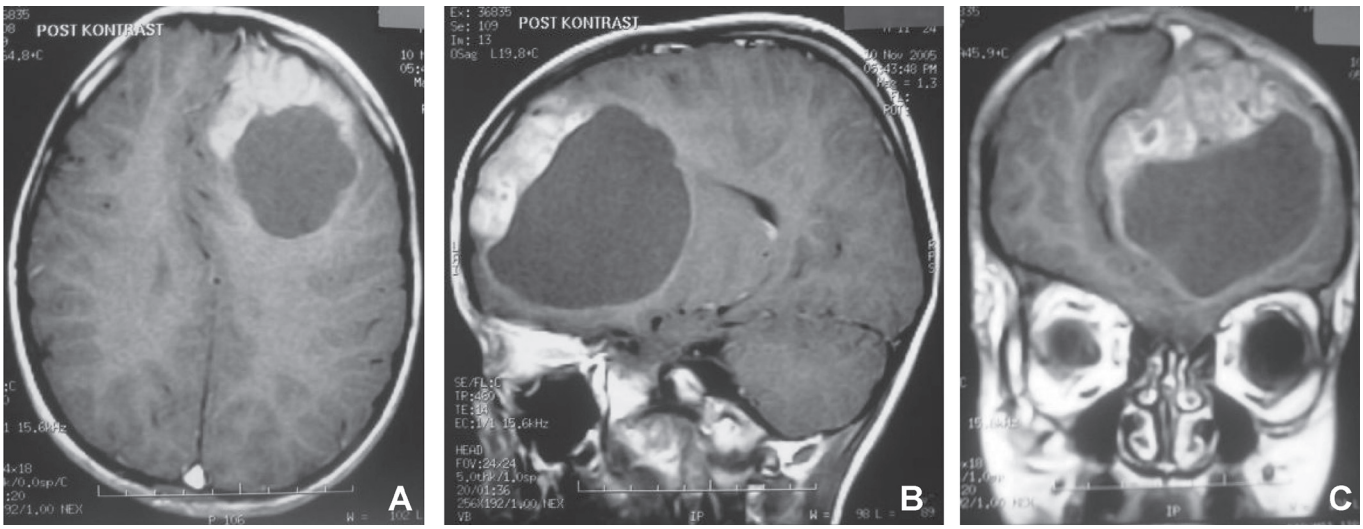

Figure 1. Preoperative axial, sagittal and coronal magnetic resonance images of the brain, showing a large left frontal cortical tumor associated with massive calcification and cyst formation.

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The Aperture, like the opening in the lens of a microscope that allows light to pass through, is a forum for art, humor, and images that provides a portal for new or different views of medicine and research.

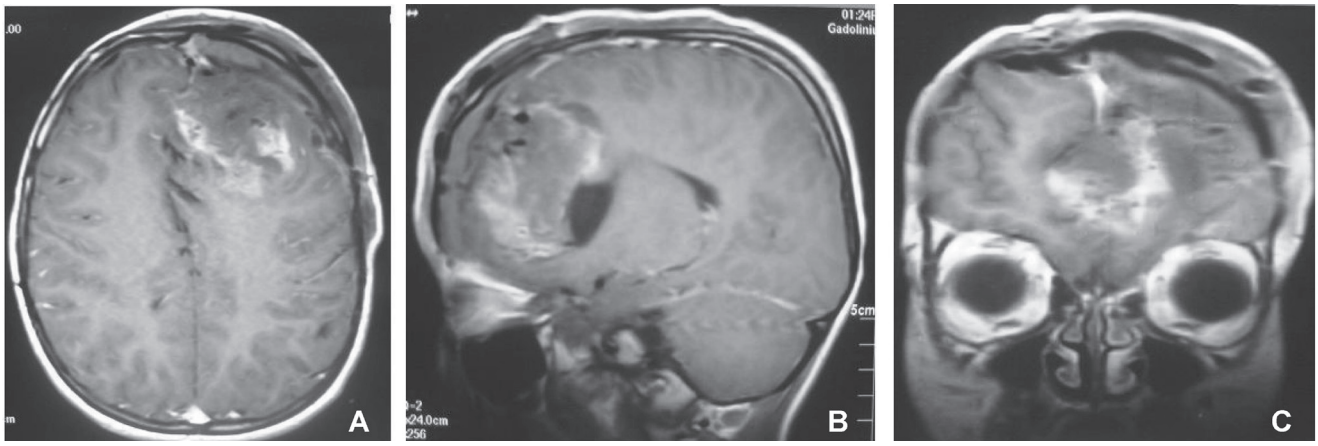

Figure 2. Postoperative axial, sagittal and coronal magnetic resonance images of the brain, showing the resection of the mass.

Case Presentation

An 11-year-old right-handed boy was admitted to our unit with complaints of headache, vomiting and generalized seizures. His systemic examination was normal. Neurological examination revealed bilateral papilledema. Routine laboratory investigations were normal. Computed tomography (CT) and magnetic resonance imaging (MRI) showed a large, poorly enhanced, left frontal mass with massive calcification and a central cyst (figure 1). Surgical resection of the mass was planned, and the patient underwent total resection of the tumor, which was located in the parenchyma with no dural attachment. The tumor was clearly demarcated and dissected subpially from the surrounding brain parenchyma. The surgical findings suggested no relationship with the lateral ventricular system. Histological examination of the tumor demonstrated perivascular pseudorosette formation and mitosis with massive calcification, and immunocytochemical positive reactivity for glial fibrillary acidic protein (GFAP) and epithelial membrane antigen, but not synaptophysin. These findings were compatible with ependymoma, World Health Organization grade 2. The patient's postoperative course was uneventful. He reported no complaints at his 3-month follow-up, and CT and MRI revealed no recurrence (figure 2). The patient was observed to be in good health 1 year after the operation.

Discussion

Ependymomas account for 3% to 9% of all neuroepithelial tumors and, although occurring most often within the ventricular system, they may arise from the extraventricular parenchyma as well.[1-5,7-10] Large supratentorial cortical ependymomas in children are a rarely seen pathological diagnosis.[1-10] They represent the most frequent primary brain tumors after pilocytic astrocytomas and medulloblastomas.[1-3,5,8] In patients under 20 years of age, approximately 60% of ependymomas arise in the infratentorium, or more specifically, in the fourth ventricle.[1-5,8,10]

The peak incidence occurs in the fourth and fifth decades, and 10% to 15% of the cases occurred in the first and second decades of life.[1-6,8-10] A few pediatric cases of supratentorial cortical ependymoma have been reported in the English literature,[1-10] but 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 whereas they have definite ependymal morphology, their histogenesis remains uncertain. Our patient shared several clinical features with previously published cases.[3-6,8,9]

Postoperative irradiation plays a role in the management of ependymomas, but indications for this adjuvant therapy remains a subject of debate. In recent studies of supratentorial ependymomas, it has been suggested that radiotherapy should be given to patients with anaplastic ependymomas,[1-6,8,9] and in cases where only partial resection of either benign or malignant tumors has been achieved.[1-5,7,8,10] A postoperative MRI with contrast has also been recommended for further evaluation of the extent of resection.[3-5,7-10] Early second-look surgery may be proposed to achieve total excision in selected patients with accessible residual tumor detected on postoperative MRI.[1-8] Therefore, reoperation to attempt complete tumor resection before initiation of radiotherapy should be considered.[1,2,6,7,9,10] Chemotherapeutic agents have been tried, particularly in children for whom radiotherapy is contraindicated or in previously irradiated subjects with an inoperable tumor, but their benefit is still questionable.[1,2-5,7,9] The 5-year overall survival and recurrence-free survival rates were 55% and 37%, respectively, and were highly affected by the extent of resection.[1-4,7,9,10] Gender, histopathologic type, localization of the tumor, extent of surgery, postoperative radiotherapy, and chemotherapy influenced the prognosis.[1-6,8,10]

Conclusion

The importance of total removal is emphasized as ependymomas do not require adjuvant therapy such as radiotherapy or chemotherapy if there is total removal of the tumor.

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