

# **Large Supratentorial Ectopic Ependymoma With Massive Calcification and Cyst Formation**

## **—Case Report—**

Shigeki ONO, Tomotsugu ICHIKAWA, Yasuhiro ONO, and Isao DATE

*Department of Neurological Surgery, Okayama Graduate School of Medicine and Dentistry, Okayama*

### **Abstract**

**A 6-year-old boy presented with a large supratentorial ependymoma with massive calcification and central cyst formation manifesting as generalized convulsion and right hemiparesis. Computed tomography and magnetic resonance imaging showed a large, poorly enhanced, left frontal mass with massive calcification and a central cyst. Angiography revealed no extracranial blood supply to the tumor, which was supplied by branches of the left middle cerebral artery. 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 reactivity for glial fibrillary acidic protein and epithelial membrane antigen, but not synaptophysin. These findings were compatible with ependymoma, World Health Organization grade 2. Postoperative magnetic resonance imaging clearly showed that the tumor was located in the intradural, intraaxial space with no relationship to the ventricles.**

Key words: ectopic ependymoma, supratentorial ependymoma, calcification

### **Introduction**

Ependymoma is a relatively rare type of glioma, which originates from the ependymal cells of the ventricular system and arises from the ventricular system or spinal cord.<sup>6,10,13,16</sup> However, ependymoma is common in pediatric patients like medulloblastoma or astrocytoma, and diagnosis is generally not difficult. On the other hand, supratentorial ependymoma is less common, cases including up to 30% of posterior fossa ependymoma.<sup>6</sup> The tumor location, presence of calcification, and enhancement patterns are very helpful in understanding the nature, but the variety of neuroimaging appearances hinder the diagnosis. Moreover, ependymoma must be distinguished from intraventricular meningioma, oligodendroglioma, subependymal giant cell astrocytoma, and so on. Supratentorial, extraaxial, intraparenchymal ependymoma isolated from the ventricular system is extremely rare.

Here we report a case of a non-ventricular, large supratentorial ependymoma with massive calcification and central cyst formation, which illustrates the difficulty of preoperative diagnosis.

### **Case Report**

A right-handed, 6-year-old boy had shown gradual deterioration in drawing ability over a period of a few weeks. The patient subsequently experienced a localized seizure on March 5, 2003, and was taken to an affiliated hospital 2 days later. On admission, mild right hemiparesis was evident. Computed tomography (CT) demonstrated a large mass extending from the thalamus to the frontal convexity, with massive calcification and a central low density area (Fig. 1). Magnetic resonance (MR) imaging revealed that this tumor reached the convexity of the dura mater, and the left lateral ventricle was compressed inferomedially (Fig. 2). T<sub>1</sub>-weighted MR imaging with gadolinium showed the tumor as isointense with a cystic central lesion, and slightly heterogeneous interior. T<sub>2</sub>-weighted MR imaging showed the

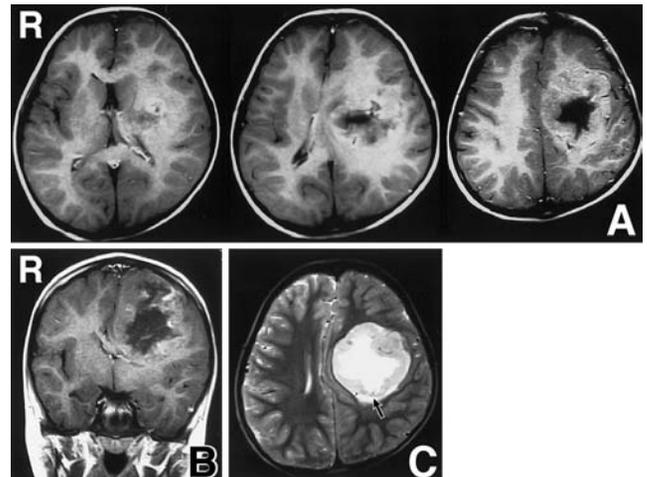


**Fig. 1** Computed tomography scan showing massive calcification of the left frontoparietal tumor with a central low-density area. No peritumoral edema was detected.

peritumoral rim as hyperintense, suggesting the tumor was an extraaxial mass such as meningioma, or ependymoma originating from the ventricles. The patient was transferred to Okayama University Hospital for further investigation and treatment on March 10, 2003.

Routine laboratory examinations, and hormonal and serological tests showed no abnormality. Right external and internal carotid angiography demonstrated no feeding arteries from the external carotid artery, and a few feeders from the branches of the middle cerebral arteries (not shown). A left frontoparietal craniotomy was performed on March 18, 2003. The inner surface of the frontal bone was slightly depressed, but no dural attachments or abnormalities were evident (Fig. 3A). The red-brown tumor was well demarcated from the normal brain tissue and could be dissected out from the epiarachnoid surface of the brain. The central cyst did not appear to exhibit central necrosis, but consisted of an assembly of microcysts. The tumor was soft, friable, and vascular rich, and calcification was psammomatous rather than bony. After debulking, the tumor was grossly removed en bloc. The attachment of the tumor was periventricular but the left lateral ventricle was not opened during this procedure (Fig. 3B). The tumor with its attachment was totally covered with pia-like membrane.

Histological examination of the surgical specimens revealed a high density of round nucleic cells with some mitoses, small calcifications, and perivascular pseudorosettes. Such psammomatous-type calcifications were widely scattered in the tumor.



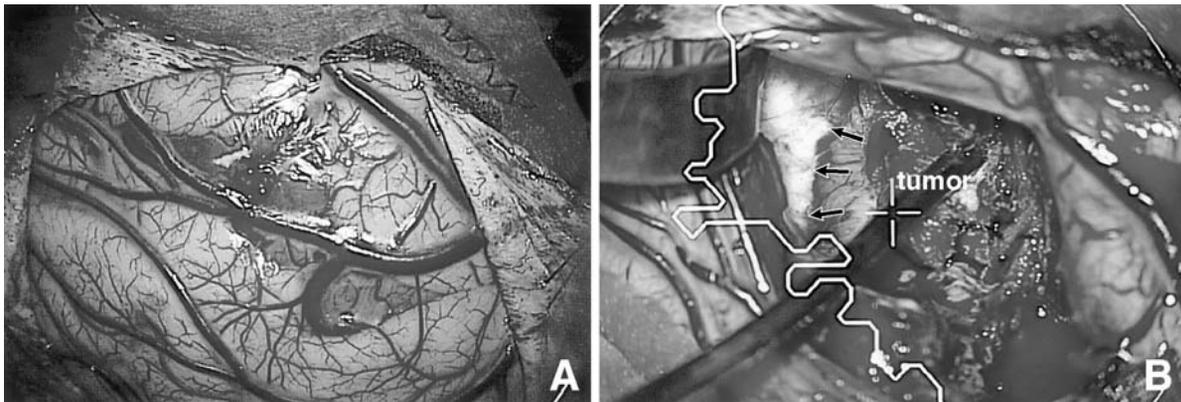
**Fig. 2** A: Axial T<sub>1</sub>-weighted magnetic resonance (MR) images with gadolinium enhancement showing the tumor was enhanced very slightly and appeared to extend to the convexity dura mater, but no dural enhancement. B: Coronal T<sub>1</sub>-weighted MR image with gadolinium enhancement showing the left ventricle was depressed inferomedially. C: Axial T<sub>2</sub>-weighted MR image showing the peritumoral high signal rim (arrow), without parenchymal edema.

Immunohistochemical staining was positive for glial fibrillary acidic protein and epithelial membrane antigen (EMA) (Fig. 4), but negative for synaptophysin. The MIB-1 labeling index was 6%. EMA-positive cells were found in the inner side of the rosettes.

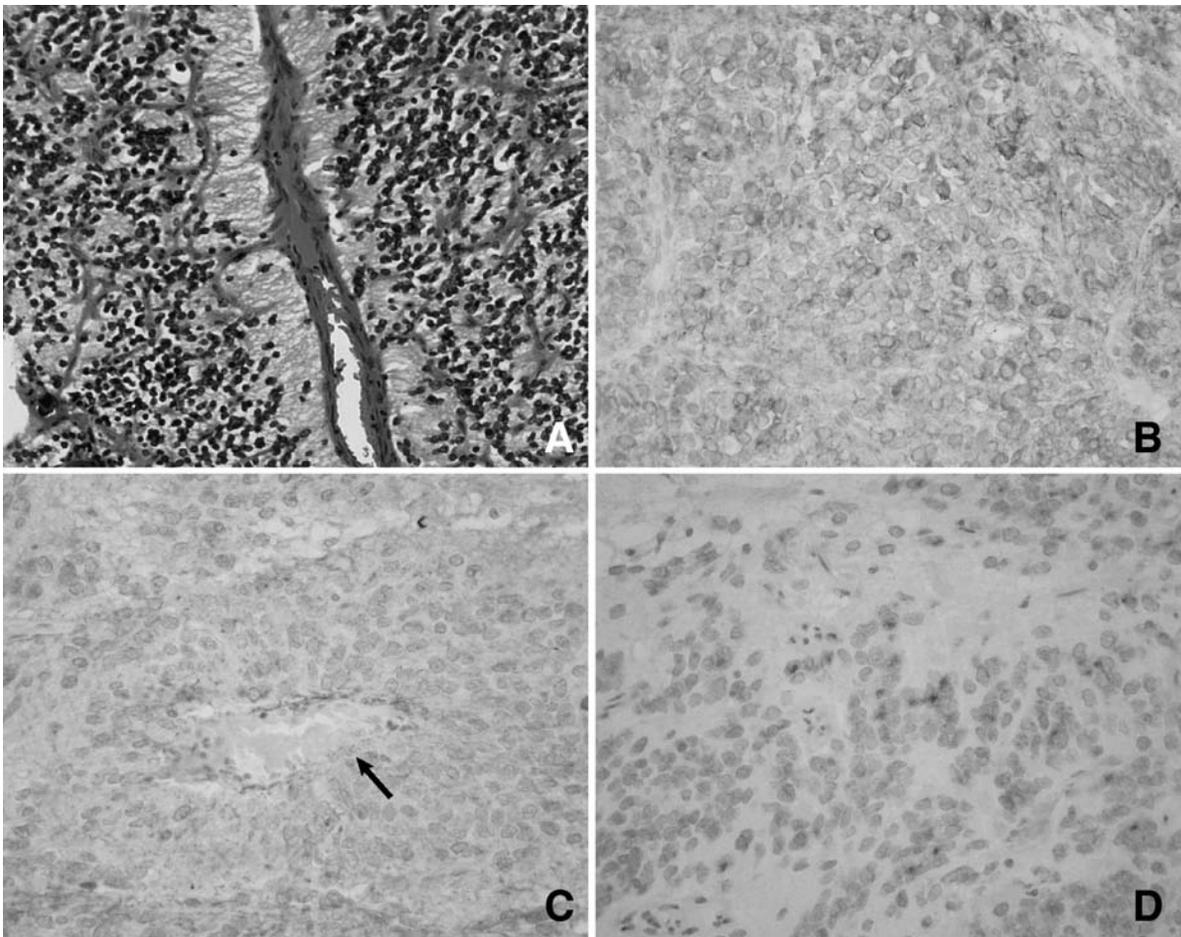
His hemiparesis resolved within 1 week of surgery, and no symptoms including headache or seizure have occurred to date. Postoperative MR imaging demonstrated total removal of the tumor and the lateral ventricle was intact (Fig. 5).

## Discussion

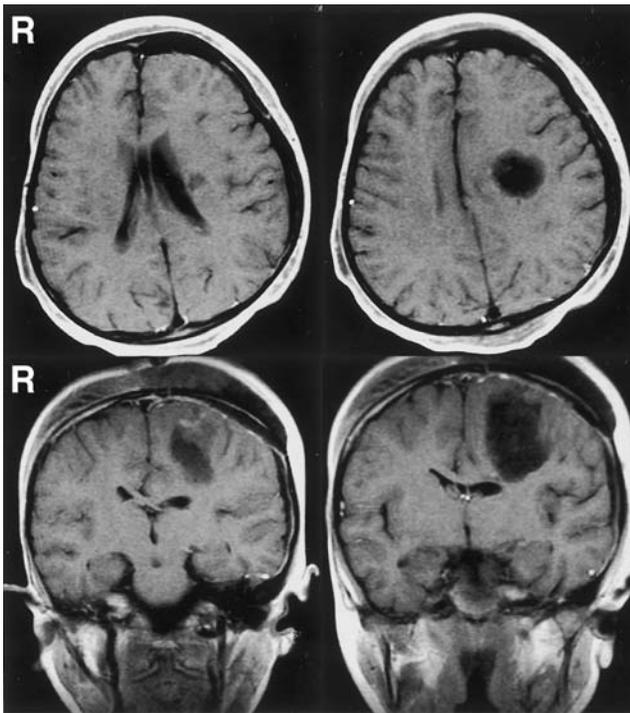
Our patient presented with a very rare large supratentorial extraaxial calcified ependymoma with no relationship to the ventricular system. Supratentorial ependymoma is not common in the general population,<sup>1-5,7-9,11,12,14,15</sup> but sometimes occurs in this location, especially in pediatric patients.<sup>11,16</sup> Eleven patients with supratentorial ependymoma had a mean age of 18.4 years, and CT generally showed a moderately calcified, moderately enhanced, mixed-cystic lesion, with peritumoral edema, but the locations were not described.<sup>1</sup> Another 20 patients with supratentorial ependymo-



**Fig. 3** A: Intraoperative macroscopic view of the brain surface showing the tumor was not attached to the dura mater, with an apparent thin capsule. B: Intraoperative microscopic view showing the border between the tumor and intact brain surface with some vasculatures (arrows). The tumor was easily dissected from the brain tissue. The white zigzag line shows the border of the tumor according to the magnetic resonance imaging-guided navigation system.



**Fig. 4** Photomicrographs of the resected tumor demonstrating perivascular pseudorosettes (A, C), and positive immunostaining for glial fibrillary acidic protein (GFAP) in the tumor cells (B) including the cells around the perivascular pseudorosettes (C: arrow), and for epithelial membrane antigen (EMA) in the tumor cells (D). A: hematoxylin-eosin stain,  $\times 200$ ; B, C: GFAP stain,  $\times 400$ ; D: EMA stain,  $\times 400$ .



**Fig. 5** Postoperative T<sub>1</sub>-weighted magnetic resonance images with gadolinium enhancement showing the tumor was totally removed. The coronal images clearly demonstrated that the tumor was not attached to the left lateral ventricle.

ma had a mean age of 13.2 years, but the origins were not reported.<sup>7)</sup> Only four cases of supratentorial ependymoma have definitely originated from intracranial extraaxial regions with no relationship to the ventricular system.<sup>3,5,9,14)</sup> Patient characteristics varied greatly, such as age 6–63 years old, partial or massive calcification, and location in the frontal, parietal, and occipital lobes. Classification of the present tumor as an extraaxial mass without relationship to the ventricular system was based on both the intraoperative findings (Fig. 3) and the postoperative MR imaging findings (Fig. 5).

Preoperative diagnosis of a supratentorial mass located in the paraventricular area is very difficult in pediatric patients. The tumors are usually large in young children, and sometimes appear to be attached to the dura mater or the ventricular system. The MR imaging characteristics of these tumors are also not uniform. For instance, ependymoma can exhibit various degrees of calcification, or have heterogeneous enhancement. Moreover, the differential diagnosis includes many rare tumors, including intraventricular meningioma, oligodendroglioma, central neurocytoma, subependymal

giant cell astrocytoma, choroid plexus papilloma, subependymoma, germ cell tumors, desmoplastic infantile ganglioglioma, and primitive neuroectodermal tumor.

Our patient presented with a confusing picture of an extraaxial mass with no feeding arteries from the middle meningeal artery. The final diagnosis of ependymoma could not be confirmed until the postoperative histological examination was performed. We considered that this tumor was ectopic with no relationship to the ventricular system because of the existence of the pia-like membrane covering the tumor. The reason for the extramedullary parenchymal location of the ependymoma in this case remains unclear. A well-known theory advocates that bundles of subependymal neural glia enter the white matter in the embryonal period to form cell nests separate from the ventricles,<sup>9,11,14)</sup> but we cannot explain the ectopic growth of this tumor, or the presence of the pia-like membrane on the surface of the tumor as seen in meningiomas or vestibular schwannomas. More cases may help us to understand the nature of this tumor.

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- Address reprint requests to: S. Ono, M.D., Department of Neurological Surgery, Okayama Graduate School of Medicine and Dentistry, 2-5-1 Shikata-cho, Okayama, Okayama 700-8558, Japan.  
e-mail: sono@cc.okayama-u.ac.jp