Ectopic Solitary Cortical Anaplastic Ependymoma in a Child: A Rare Entity with Short Illustrative Review

Sachin Guthe 1  Pravin Survashe 1  Vernon Velho 1  Laxmikant Bhopale 1  Poonam Darade 2

1Department of Neurosurgery, Grant Medical College and Sir J.J. Group of Hospitals, Mumbai, Maharashtra, India  
2Department of Radiology, Grant Medical College and Sir J.J. Group of Hospitals Mumbai, Maharashtra, India

Ependymomas are usually infratentorial and intraventricular. They originate from the ependymal cell lining of the ventricles. Cortical extraventricular supratentorial ependymomas are rare and fewer than 15 cases are reported worldwide. In pediatric age group, seven cases are reported. We report a rare case of 4-year-old boy with right frontoparietal anaplastic ependymoma who underwent gross total excision of lesion.

Abstract

Keywords
► anaplastic  
► cortical  
► pediatric ependymoma

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Introduction

Ependymomas are tumors derived from ependymal cells lining the ventricles or from the central canal of the spinal cord. It represents 3 to 9% of all neuroepithelial neoplasms, 6 to 12% of all pediatric brain tumors, and almost one-third of all brain tumors in patients younger than 3 years. Approximately 40% of ependymomas are supratentorial, whereas 60% are infratentorial in location. Ependymomas may manifest at any age (documented age ranges from 1 month to 81 years) with no sex predilection. Posterior fossa ependymomas are common in children (mean age: 6 years) and supratentorial ependymoma generally manifests in an older age group (mean age: 18–24 years). Although the lesion arises from the ventricular system, it can grow beyond the ventricles, through the cerebral tissue, representing the extraventricular form. It can exist in extraventricular structures, without any connection to ventricular system, representing the rare group of ectopic ependymoma. In the literature not more than 30 cases have been reported, of which 15 were diagnosed purely cortical and only 7 cases were anaplastic (grade III) lesions → Table 1.  

We report a rare case of solitary cortical pediatric anaplastic ependymoma.

Illustrative Case

A 4-year-old boy presented with progressively increasing headache and vomiting of 1-month duration. On examination he was drowsy and had left hemiparesis. A contrast-enhanced computed tomographic (CT) scan of the brain showed a right frontal superficially located mixed-density lesion with heterogeneous enhancement (→Fig. 1). Intraoperatively gross total excision of the tumor was done via

Table 1 All reported cases of pure cortical supratentorial grade III ependymomas

<table>
<thead>
<tr>
<th>Series</th>
<th>Cases</th>
<th>Grade</th>
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<tbody>
<tr>
<td>Davis et al 1</td>
<td>1</td>
<td>III</td>
</tr>
<tr>
<td>Alexiou et al 2</td>
<td>1</td>
<td>III</td>
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<td>Hamano et al 3</td>
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<td>III</td>
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<td>Akyuz et al 4</td>
<td>1</td>
<td>III</td>
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<tr>
<td>Romero et al 5</td>
<td>1</td>
<td>III</td>
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<tr>
<td>Kharosekar et al 6</td>
<td>1</td>
<td>III</td>
</tr>
<tr>
<td>Present study</td>
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<td>III</td>
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</tbody>
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1Department of Neurosurgery, Grant Medical College and Sir J.J. Group of Hospitals, Mumbai, Maharashtra, India
2Department of Radiology, Grant Medical College and Sir J.J. Group of Hospitals Mumbai, Maharashtra, India

Address for correspondence Dr. Sachin Guthe, MCh Neurosurgery, Department of Neurosurgery, 4th Floor, Main Building, J.J. Hospital Campus, Byculla, Mumbai, Maharashtra 400 008, India (e-mail: spguthe@gmail.com).
right frontoparietal craniectomy (►Fig. 2). No connection of tumor to the ventricular ependymal lining was noted. Tumor was moderately vascular and suckable. Margins were well defined. The postoperative period was uneventful. He made a good recovery and there was no fresh neurologic deficit. Histopathology examination showed plump spindle-shaped ependymal cells. The cells showed a tendency to form rosettes. There were pseudorosettes around blood vessels. Frequent mitosis, foci of microvascular proliferation, and large confluent areas of necrosis were seen. There were foci of dystrophic calcification (►Fig. 3). Immunohistochemistry was positive for glial fibrillary acidic protein and epithelial membrane antigen. Ki67 proliferation index was greater than 15% suggestive of anaplastic cortical ependymoma grade III.

**Discussion**

Although approximately half of the supratentorial ependymomas arise from the wall of third or lateral ventricles,
and are purely intraventricular, few may extend through adjacent cerebral tissue, representing extraventricular forms of ependymoma. Only few cases occur in distant places of the ventricular system, representing rare cases of ectopic lesions. It is speculated that ectopic ependymomas may arise from embryonic rests of ependymal tissue trapped in the developing cerebral hemispheres. Supratentorial ependymoma grows into third or lateral ventricle; it is predominant involving the brain parenchyma at the diagnosis. Hamano et al reported that 83% of supratentorial ependymomas are in the cerebral parenchyma. Owing to its parenchymal location, the supratentorial ependymoma tends to be larger in size at the diagnosis.

Roncaroli et al found that 94% of supratentorial tumors manifest with a size larger than 4 cm and often contain a cystic component. Despite their large size in the cerebral hemispheres, symptoms are relatively mild until a later stage of presentation. Symptoms of raised intracranial pressure such as headache and vomiting are common, whereas focal signs as limb weakness and seizures are less prevalent. The principal differential diagnosis of extraventricular supratentorial ependymoma must include astrocytoma (both low-grade and
Prognostic factors of ependymomas that positively contribute to progression-free survival and longer survival are still elusive, even in histologic characteristics. The 5-year progression-free survival rate for children overall is approximately 50% and 10-year survival rates for adults are 57.1% and 45%, respectively. Only total tumor resection is considered as a reliable prognostic factor for predicting longer survival time. Of patients with no radiologic evidence of residual tumor, 75% ± 15% will remain tumor free after 5 years as opposed to the group of patients with residual disease in which progression cannot be stopped.

An increased risk of recurrence was reported with a high histologic grade, incomplete resection, and a Karnofsky performance status that is 80 or less.\(^7\)\(^8\)

Our patient was treated with radical surgery and subjected to postoperative radiotherapy, due to anaplastic grade of tumor. There was no evidence of gross residual tumor at postoperative imaging. The patient had a good recovery of neurologic symptoms. His left hemiparesis improved. Now he is regularly being followed in our outpatient clinic.

**Consent**

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

**Funding**

None.

**Conflict of Interest**

None.

**References**


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Fig. 3 Histopathology image showing perivascular pseudorosettes.