

## Pediatric Isolated Cortical (Ectopic) Anaplastic Ependymoma

### Abstract

Ependymomas are tumors derived from ependymal cells lining the ventricles or from the central canal of the spinal cord. It usually arises in the ventricles with extra ventricular extension. Less than 15 cases of purely cortical ependymomas are reported. We report a rare case of purely cortical anaplastic ependymoma in a pediatric patient, which is rarely reported.

**Keywords:** *Anaplastic, ectopic, ependymoma, pediatric, purely cortical*

### Introduction

Ependymomas are tumors derived from ependymal cells lining the ventricles or from the central canal of the spinal cord. It represents 3–9% of all neuroepithelial neoplasms, 6–12% of all pediatric brain tumors. Forty percent of ependymomas are supratentorial whereas 60% are infratentorial in location. Ependymomas may manifest at any age, posterior fossa ependymoma arises most often in children (mean age, 6 years) while supratentorial ependymoma generally manifests in an older age group (mean age, 18–24 years). It usually arises in the ventricles with extra ventricular extension, but they may occur outside the ventricular structures, without any relationship of the ventricular system, representing the rare group of ectopic ependymoma. Less than thirty cases of ectopic ependymomas were reported, and almost fifteen were purely cortical, and only five cases were anaplastic lesions.<sup>[1]</sup> We report a rare case of pure cortical ectopic anaplastic ependymoma [Table 1].

### Case Report

An 11-year-old girl presented to us with progressively increasing headache and vomiting for 1 month duration. She also had a left-side motor weakness. Neurological examination showed left hemiparesis. Contrast computerized tomography (CT) brain scan showed a right frontoparietal superficially located mixed density lesion with heterogeneous enhancement [Figure 1a and b]. Magnetic resonance imaging (MRI) demonstrated

a solid/cystic cortical lesion in the right frontoparietal region with significant edema and peripheral enhancement on contrast, without any relationship of lateral or third ventricle [Figure 2a-c]. A right-sided craniotomy and gross total excision of the tumor was done. The tumor had no connection to the ventricular ependymal lining. Tumor was moderately vascular, grayish, and suckable. Margins were well defined. The postoperative period was uneventful. Postoperative CT scan showed complete excision of the lesion [Figure 3]. She had postoperative weakness on the left-side which improved with hyperbaric oxygen therapy. Histopathology of the excised tumor showed microvascular proliferation and pseudopalisading. True rosettes and perivascular pseudorosettes were also seen. Mitosis was seen in 8F/10 HPF with necrosis was seen. It was positive for glial fibrillary acidic protein and epithelial membrane antigen. Ki67 proliferation index was >15% suggestive of anaplastic cortical ependymoma Gr 3 [Figure 4a and b].

### Discussion

Fifty percent of the supratentorial ependymomas arise from the wall of third or lateral ventricles and are purely intraventricular; the remaining has an extension through adjacent cerebral tissue, representing extra ventricular forms of ependymoma. Only a few cases occur in distant places of the ventricular system, representing rare cases of ectopic lesions. It is speculated that ectopic ependymomas

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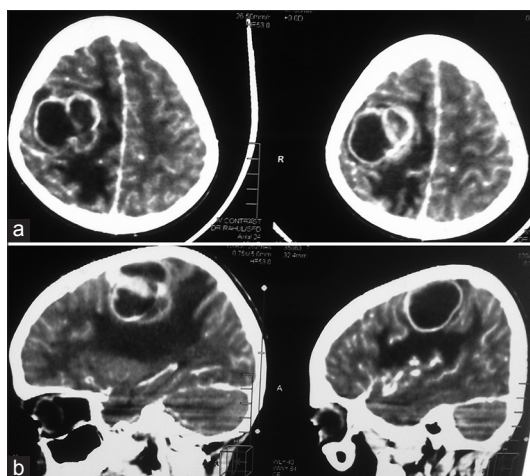


Figure 1: (a and b) Computerized tomography brain with contrast images. Axial and sagittal cuts

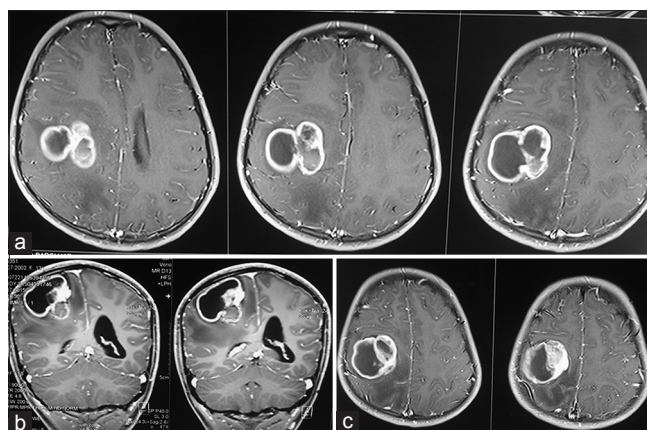


Figure 2: (a-c) Magnetic resonance imaging brain with contrast, T1- and T2-weighted images showing the lesion. No communication with the ventricle. Right sided ventricle is compressed

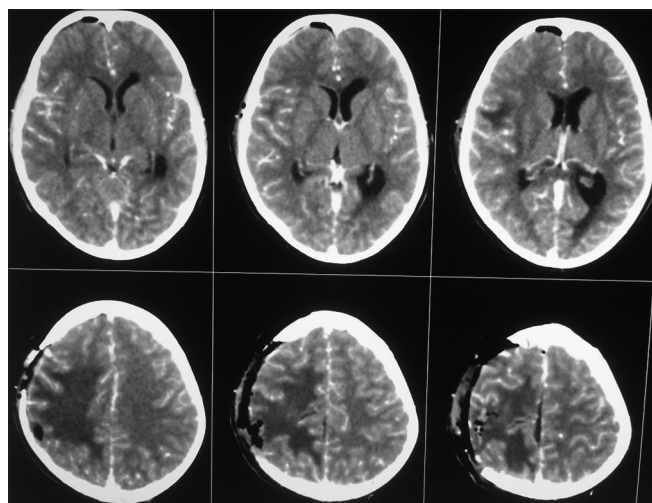


Figure 3: Postoperative computerized tomography brain with contrast images showing complete excision

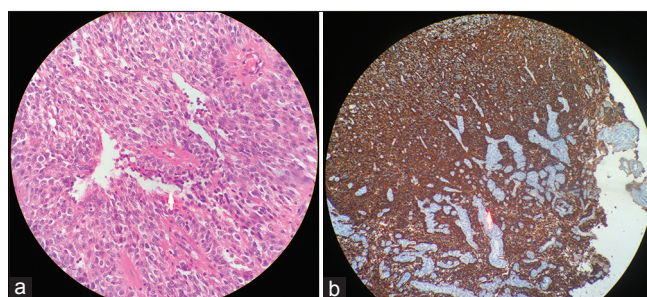


Figure 4: (a) Histopathology images. Perivascular pseudorosettes can be seen. (b) Immunohistochemistry

**Table 1: Cases of pure cortical anaplastic ependymoma till 2012**

Series	Cases	Grade
Davis <i>et al.</i>	1	III (anaplastic)
Alexiou <i>et al.</i>	1	III
Hamano <i>et al.</i>	1	III
Akyuz <i>et al.</i>	1	III
Romero <i>et al.</i>	1	III

may arise from embryonic rests of ependymal tissue trapped in the developing cerebral hemispheres. Supratentorial ependymoma grows up of the third or lateral ventricle, it is predominant involving the brain parenchyma at the diagnosis. Owing to its parenchymal location, the supratentorial ependymoma tends to be larger in size at the diagnosis. These tumors usually present with symptoms of raised intracranial pressure such as headache and vomiting are common, whereas focal signs as limb weakness and seizures are less prevalent. Differential diagnosis

of these tumors includes glioblastoma multiforme, supratentorial primitive neuroectodermal tumor (PNET), and oligodendroglioma. Despite the malignant designation of anaplastic ependymomas, they tend to be solid and well demarcated with limited infiltration to the edges of the lesion. They do not have typical images findings and extension to the ventricular system is suspicious. They are iso- to slightly hypo-attenuating to surrounding normal brain tissue at unenhanced CT. They are iso- to hypo-intense relative to normal white matter on unenhanced T1-weighted MRIs and hyperintense on T2-weighted MRIs. Foci of signal heterogeneity within a solid neoplasm represent methemoglobin, hemosiderin, necrosis, or calcification that is very common in this tumor.<sup>[1]</sup>

Histologically, the tumor cells are characteristically organized in perivascular pseudorosettes. Although ependymomas are moderately cellular tumors with rare mitotic figures (WHO 2), our patient had a more aggressive tumor, classified as the WHO Grade III. Less than five ectopic anaplastic ependymomas are reported previously in literature. It is also reported that approximately 15% of anaplastic ependymoma have a prior diagnosis other than ependymoma, and these tumors were subsequently reclassified as ependymoma, as in our case, which was thought to be PNET initially. The lack of clinicohistopathological co-relation highlights the need for

establishing criteria for classifying these tumors according to their degree of anaplasticity.<sup>[2]</sup>

The 5 years progression-free survival rate for children overall is about 50%. Age at presentation is a significant prognostic factor. In the young children, 5 years survival is 22–40% as compared to 60–75% in older children. Furthermore, patients with symptoms before diagnosis for <1 month have a worse outcome than those with a more protracted course. Successful gross total resection appears to be the best prognostic indicator of long-term survival. Pure cortical (ectopic) tumors can be approached easier than lesions with ventricular extension, having better outcome. The need for postoperative adjuvant therapy has been controversial for supratentorial ependymomas. Postoperative radiation therapy must be administered in every case of partially resected ependymomas or anaplastic tumors. An increased risk of recurrence was reported with a high histological grade, incomplete resection, and a Karnofsky performance status that is  $\leq 80$ . In our patient complete excision was achieved, but postoperative radiation therapy was advised because of the anaplastic grade of tumor.<sup>[1,3]</sup>

## Conclusion

As these are very rare tumors, there is increased need to increase the awareness of these lesions during preoperative studies.

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## Conflicts of interest

There are no conflicts of interest.

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