

## Case Report

# Anaplastic supratentorial cortical ependymoma presenting as a butterfly lesion

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Received: 16 May 12

Accepted: 07 August 12

Published: 13 September 12

**This article may be cited as:**Ng DW, King NK, Foo AS, Sitoh Y, Lee HY, Ng WH. Anaplastic supratentorial cortical ependymoma presenting as a butterfly lesion. *Surg Neurol Int* 2012;3:107.Available FREE in open access from: <http://www.surgicalneurologyint.com/text.asp?2012/3/1/107/101001>

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## Abstract

**Background:** Anaplastic cortical ependymomas are rare lesions with few cases reported in the literature.**Case Description:** We present a unique case of an anaplastic cortical ependymoma in a 51-year-old female presenting as a butterfly lesion with involvement of both frontal lobes. The patient underwent gross total resection of her tumor with further adjuvant treatment. We present the findings in our case and review the literature surrounding supratentorial ependymomas and their treatment outcomes.**Conclusion:** Rarely, cortical ependymoma can present as a butterfly lesion and should be considered in the differential diagnosis of such lesions in adults.**Key Words:** Adult, Anaplastic, Cortical, Ependymoma, Supratentorial**Access this article online****Website:**[www.surgicalneurologyint.com](http://www.surgicalneurologyint.com)**DOI:**

10.4103/2152-7806.101001

**Quick Response Code:**

## INTRODUCTION

Ependymomas are tumors arising from the cells lining the ventricular system and the central canal of the spinal cord. In adults, ependymomas are uncommon, have a propensity to occur supratentorially and are associated with a higher incidence of anaplasticity.<sup>[18]</sup> Supratentorial ependymomas (STE) can be further divided into intraventricular and extraventricular types [Table 1]. Intraventricular ependymomas can be in either the third ventricle or lateral ventricles. Extraventricular ependymomas arise distant to the ventricles within the cerebral parenchyma and can even be purely cortical, arising purely within the cerebral cortex. The origin of the extraventricular STE is thought to be from fetal ependymal cell rests located in the periventricular region or at the angles of the ventricles.<sup>[28]</sup> Pure cortical ependymomas are the rarest, and we present, to our

knowledge, a previously unreported case of an anaplastic ependymoma presenting as a butterfly lesion crossing the midline.

## CASE REPORT

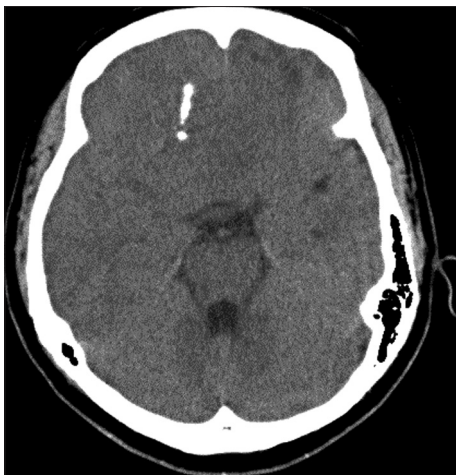
A 51-year-old Chinese female presented with the incidental finding of a large heterogeneously enhancing mass in the frontal lobes after a computer tomography (CT) scan for minor head injury following a fall [Figure 1]. Her past medical history included only adenomyosis and endometrial polyps. No neurological abnormalities were observed.

Magnetic resonance imaging (MRI) revealed a 76 × 70 × 54 mm mass in both frontal lobes with extension across the midline that was hypointense on T1-weighted, hyperintense on T2-weighted images, and

demonstrated avid postgadolinium contrast enhancement [Figure 2]. The lesion showed foci of calcification and peripheral cystic components, with mild perilesional edema and significant mass effect. The corpus callosum was displaced posteriorly. Inferiorly, the lesion extended up to the olfactory bulb.

The patient underwent gross total resection (GTR) of the tumor. Intraoperatively the tumor was not found to breach ventricles and the tumor was deemed to be entirely extraventricular. Intraoperative frozen section suggested the diagnosis of a high grade glioma. Histology showed a well demarcated cellular tumor [Figure 3] with prominent perivascular pseudo-rosettes and very occasional true (ependymal) rosettes. The tumor cells showed enlarged, hyperchromatic, pleomorphic nuclei, a granular 'salt and pepper' chromatin pattern, and fibrillary cytoplasm. Mitotic figures, including atypical forms were readily identified. There were areas of palisading necrosis and microvascular proliferation. The Ki-67 proliferative index was about 20%. These features were diagnostic of an anaplastic ependymoma (WHO grade III).

MRI taken on postoperative day 1 [Figure 4] showed no evidence of residual tumor. The patient's postoperative recovery was uneventful. Cerebrospinal fluid (CSF) studies were negative for malignant cells and no drop metastases were detected on neuroaxis MRI. The patient underwent adjuvant intensity-modulated radiation therapy of 60 Grays in 30 fractions to a region encompassing the tumor bed and a 2 cm margin around it over a period of 2 months. At follow up, the patient developed local recurrence in the frontal lobes within 2 months of completing radiotherapy. At 8 months postsurgery, progression of disease locally had caused her to become increasingly drowsy and by then she was wheelchair bound.

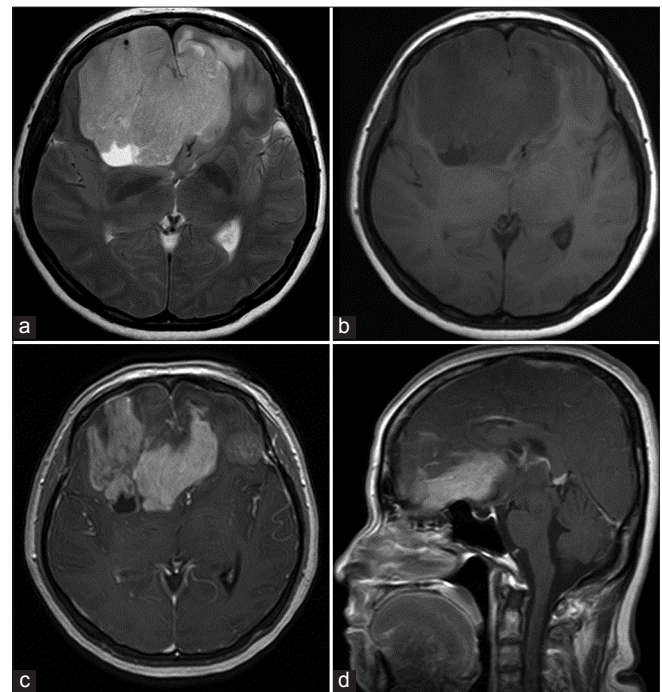


**Figure 1:** Noncontrast CT brain showing bifrontal lesion with area of focal calcification

## DISCUSSION

The term 'butterfly lesion' describes lesions that extend across the midline, often via involvement of the corpus callosum. Lesions are typically aggressively infiltrative tumors such as glioblastoma multiforme (GBM) and B-cell lymphoma, and, given that the corpus callosum comprises primarily of myelinated axons, demyelinating disease such as multiple sclerosis.<sup>[5,6]</sup> In an immunocompromised patient, primary central nervous system (PCNS) lymphoma, progressive multifocal leukoencephalopathy, and rarely, toxoplasmosis have been reported.<sup>[5,7,8]</sup> In our patient the corpus callosum was found to have been displaced posteriorly rather than infiltrated, a feature that helped to differentiate her tumor from an infiltrative process. As cortical STEs are rare and have not been reported to present as butterfly lesions, they were not considered in our pre-operative differential diagnosis.

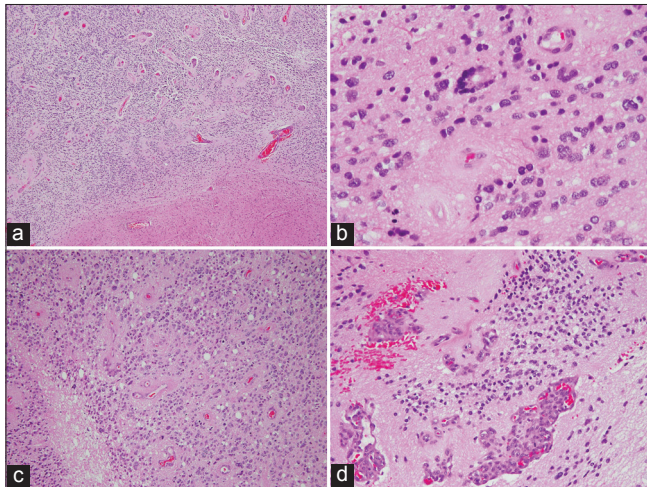
STEs are usually large ( $\geq 4$  cm) and cystic with their location classified as shown in Table 1. On CT they demonstrate moderate to intense contrast enhancement with homogeneous or ring-enhancement patterns. On MRI, they appear hypointense on T1-weighted and hyperintense on T2-weighted imaging, respectively. Peritumoral edema and hydrocephalus may be present and about one-third of tumors may harbor calcification.<sup>[2,29]</sup> Extraventricular tumors tended to present with seizures, focal neurological deficits, and behavioral disturbances



**Figure 2:** MRI showing bifrontal lesion crossing the midline and displacing the corpus callosum posteriorly (a) axial T2-weighted (b) axial T1-weighted (c) postcontrast axial T1-weighted, and (d) postcontrast sagittal T1-weighted images

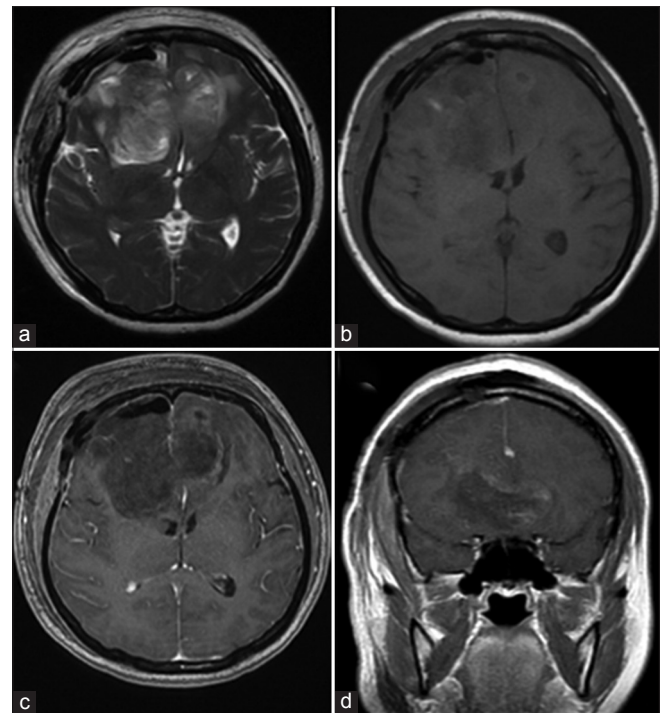
while intraventricular lesions present with hydrocephalus and raised intracranial pressure.<sup>[15,30,31]</sup>

Although the clinical features in this case were highly suggestive of a GBM, this was later disproved by the classic histopathological features of an anaplastic ependymoma. There are also no known cases of GBM with ependymal differentiation reported in the literature.



**Figure 3:** (a) Low power view showing a cellular tumor with perivascular pseudorosettes and a well demarcated border with the adjacent brain parenchyma (H and E, × 40). (b) An occasional true (ependymal) rosette is identified (H and E, × 400). (c) The tumor is mitotically active and shows areas of necrosis (H and E, × 200). (d) Microvascular proliferation is also present (H and E, × 200)

Several prognostic factors have been identified with regard to survival outcome and tumor recurrence. Compared with infratentorial or spinal ependymomas, patients with STE tend to have poorer overall survival (OS) and progression-



**Figure 4:** MRI on postoperative day 1 showing complete tumor resection (a) axial T2-weighted (b) axial T1-weighted (c) postcontrast axial T1-weighted, and (d) postcontrast coronal T1-weighted images

**Table 1: Classification of adult supratentorial ependymomas by tumor location**

Classification of STE	Definition	No. of cases	Studies
Intraventricular	Arising from ventricles	a. 1 b. 1 c. 1 d. 24	a. Danyemez (1999) <sup>[8]</sup> b. Hayashi (2000) <sup>[9]</sup> c. Ragel (2005) <sup>[23]</sup> d. Metellus (2008) <sup>[15]</sup>
Extraventricular	Parenchymal/Subcortical	a. 2 (thalamic) 23 (parenchymal)	a. Molina (1999) <sup>[17]</sup>
		b. 22	b. Metellus (2008) <sup>[15]</sup>
	Pure Cortical	a. 1 b. 2 c. 3	a. Saito (1999) <sup>[26]</sup> b. Roncaroli (2005) <sup>[25]</sup> c. Niazi (2008) <sup>[19]</sup>

**Table 2: Case series reporting on outcome in adult with supratentorial ependymomas following treatment**

Study	N	WHO Grade (%)	Surgery GTR (%)	Adjuvant RT (%)	Chemo	Outcome
Saito (1999) <sup>[26]</sup>	1	II (100)	100	100	None	NED at 14 months
Molina (1999) <sup>[17]</sup>	25	III (100)	96	80	12%	5-year OS: 16.0%
Roncaroli (2005) <sup>[25]</sup>	2	II (100)	100	50	None	5-year OS: 100%
Metellus (2007) <sup>[14]</sup>	46	II (46) III (54)	50	48	None	5-year OS: 63% ± 9%
Niazi (2008) <sup>[19]</sup>	3	II (33) III (67)	100	67	None	1 death after 14 months, and NED at 29 months and at 42 months
Metellus (2010) <sup>[16]</sup>	22	II (100)	55	32	None	5-year OS: 75% ± 11%

GTR: gross total resection, NED: No evidence of disease, OS: overall-survival, RT: radiotherapy



free survival (PFS) in patients.<sup>[1,13,14]</sup> Other predictors factors of poor prognosis include older age,<sup>[13,15]</sup> presence of microvascular proliferation, presence of necrosis, number of mitoses, degree of hypercellularity,<sup>[20]</sup> and greater Ki-67/MIB-1 proliferation index.<sup>[3,15]</sup> Extent of tumor resection was a positive predictor of OS and PFS,<sup>[1,14,15]</sup> and subtotal resection was correlated with subsequent CSF dissemination and metastasis. There is controversy about the prognostic value of the current histological grading system,<sup>[11,12,14]</sup> and a lack of consensus in establishing the criteria for classifying tumors according to the degree of anaplasticity.<sup>[12,14,15]</sup> However, overall histological grade and incompleteness of resection have been identified as significant predictors of tumor recurrence.<sup>[3,27]</sup>

Maximal safe resection with adjuvant radiotherapy (RT) is the current mainstay of treatment.<sup>[13,19,20,25]</sup> There has been interest in identifying patients in whom RT can be avoided without worsening treatment outcome with several reports showing treatment success with GTR without adjuvant RT for low grade ependymomas<sup>[4,21,24]</sup> and in some anaplastic STE in pediatric cases.<sup>[10,22]</sup> Studies have shown that prophylactic spinal irradiation has little role in the treatment of STE as the cortical lesions have a negligible risk of drop metastases and leptomeningeal seeding due to their distance from CSF pathways.<sup>[20]</sup> The overall outcome after surgery for STE is summarized in 2.

Our case is a unique case of an adult anaplastic supratentorial cortical ependymoma presenting as a butterfly lesion. This particular diagnosis should be considered in the differential diagnosis of such butterfly lesions in an adult [Table 2].

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