

Cortical Ependymoma with Extension to Thalamus

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ABSTRACT

Ependymomas are glial tumors originate from ependymal cells lining the ventricles and the central canal of the spinal cord. Two thirds of ependymomas arise in the infratentorial or intraventricles and one-third are detected in supratentorial area. However supratentorial “cortical” ependymomas are very uncommon. Ependymomas are usually infratentorial, intraventricular tumor, including 2–9% of all central nervous system tumors. We present a unique case of an anaplastic cortical ependymoma in a 48-year-old man. The patient presented with transient amnesia and right weakness and seizure. This tumor was located in the left parieto-occipital region with extension to corpus callosum and thalamus. Microscopy and immunohistochemistry showed grade III differentiation ependymoma.

Keywords: Brain tumor; Ependymoma; Supratentorial

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INTRODUCTION

Supratentorial cortical ependymomas (CE) are rare tumors with very few cases described up to now in the literature. They occur in the superficial cortical strip and have no relation with the ventricular lining¹. These tumor have partially benign course and shows close connection with the current explained item i.e., angiocentricglioma (AG)².

CASE PRESENTATION

A 48-year-old right-handed man was admitted to our unit with complaints of repetitive transient amnesia from 2 years ago and generalized tonic clonic seizure. The patient had history of parietal headache. His systemic examination was normal. Sensory and motor examinations revealed right hemiparesis about 4/5 in upper and lower limbs.

Magnetic resonance imaging (MRI) showed a large mass in the left parieto-occipital region with extension to corpus callosum and thalamus with no enhancement and cortical presentation (Figure 1). Brain CT showed hyperdense mass lesion in left parieto-occipital area

(Figure 2). Surgical biopsy of the mass was planned and the patient underwent biopsy of the tumor. Sections show brain tissue infiltrated by a glial neoplasm composed of atypical glial cells some with occasionally clear cell features and minigemistocytes cells. In some foci perivascular pseudo-rosettes architecture was detected. Immunohistochemistry reveal diffuse positive reactivity for glial fibrillary acidic protein. The Ki-67 index was focally up to 30% and EMA was positive; World Health Organization (WHO) Grade III (Figure 3).

DISCUSSION

Ependymomas are central nervous system (CNS) tumor originating from ependymal cells covering the inner surface of brain ventricle and across the central spinal cord³. They are usually detected in the cervicothoracic area of the spinal central canal and fourth ventricle^{3,4}. Supratentorial cortical ependymoma is an uncommon item where the tumor is seen in cortical area with no relation to the ventricular lining². The recent WHO classification of CNS tumor categorized ependymoma into low grade (Grade II) and high grade

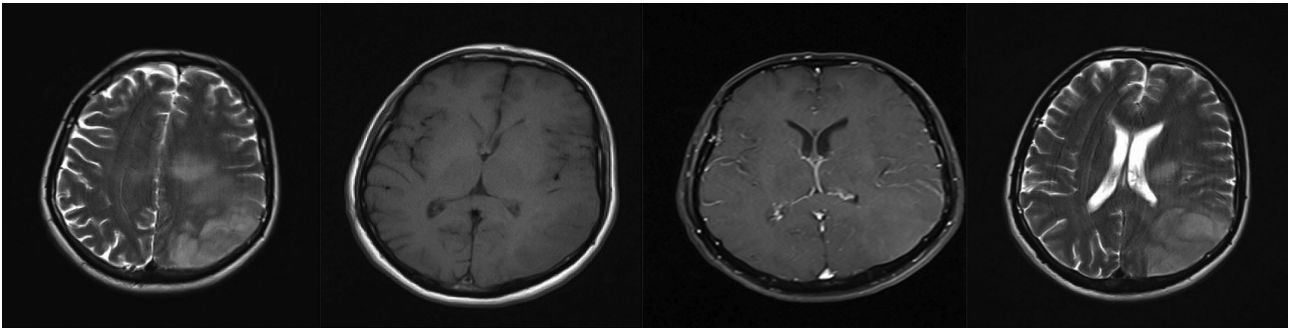


Figure 1. Cortical left parietooccipital mass lesion with extension to thalamus and corpus callosum with no enhancement.

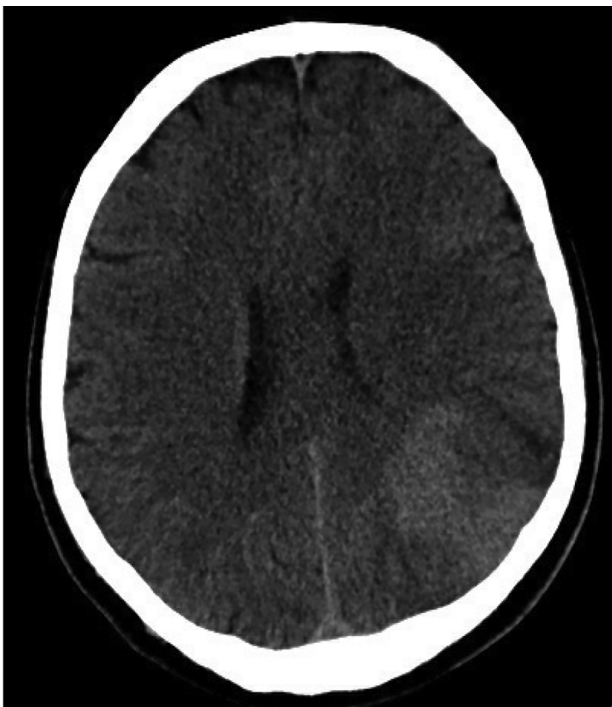


Figure 2. Hyperdense left parietooccipital mass lesion in brain CT.

(Grade III) or anaplastic ependymoma, reserving Grade I for myxopapillary ependymoma⁵. The mean age of presentation is 27 ± 19 years³. They generally, but not always present with seizures, but even though their high relation to epilepsy they are scarcely seen in temporal lobe⁶.

The pathogenesis of ependymoma is undetermined. Some explain that originate from cell type other than terminally differentiated ependyma since they are detected at locations where ependymal cells are absent⁶. Hegyi et al. in their reported case of ectopic retinal ependymoma considered their origin from Muller cells, concluding that glial cells with progenitor features are the cell of origin for these tumor in place of terminally differentiated ependyma⁷. Progenitor cell hypothesis

could be proposed to describe the origin of CE as their occurrence in teratomas seem to support this prospect⁸⁻¹⁰. Roentgenographically, it shows a well-defined solid to cystic mass detected in peripheral cortical rim with least associated vasogenic edema³.

On squash cytology it indicates that cellular smear consisted of tumor cells in papillary pattern opposite to fibrillary background with local region exhibiting perivascular pseudorosettes. The cells exhibit round nucleus with dot chromatin and 1–2 small nucleoli¹¹. Histopathology exhibits true ependymal rosettes and perivascular pseudorosettes which are the typical properties determining these tumors. Less classic morphological properties that can be detected consisted of tancytic, epithelioid and clear cell properties³. Uncommonly it can exhibit additional spindle cell element and schwannian like nodules properties that are classic for AG as explained by Lehman¹². AG are seizure associated tumor characterized by an angiocentric pattern of growth, monomorphous bipolar cells and properties of ependymal differentiation¹³. The close differential that can also be proposed for this tumor is astroblastoma, which also shows rosettes which have shorter and stouter cytoplasmic processes when compared to rosettes of ependymoma¹⁴. Immunohistochemistry plays a key role in discriminating these two items as EMA show dot like positivity in ependymomas, which is representative for this tumor². Anaplastic Grade III ependymomas are known by hypercellularity, increased mitotic activity, microvascular proliferation and pseudopalisading necrosis. It is important to determine anaplastic ependymoma as it indicates increased chances of recurrences and needs radiation therapy following survival resection¹. Metastatic seeding across the CSF pathway is usually detected and is extensive in anaplastic form¹. Therefore, preoperative CSF study for excluding metastatic seeding along with postoperative MRI to examine the size of excision is required. Surgical excision is the essential modality of

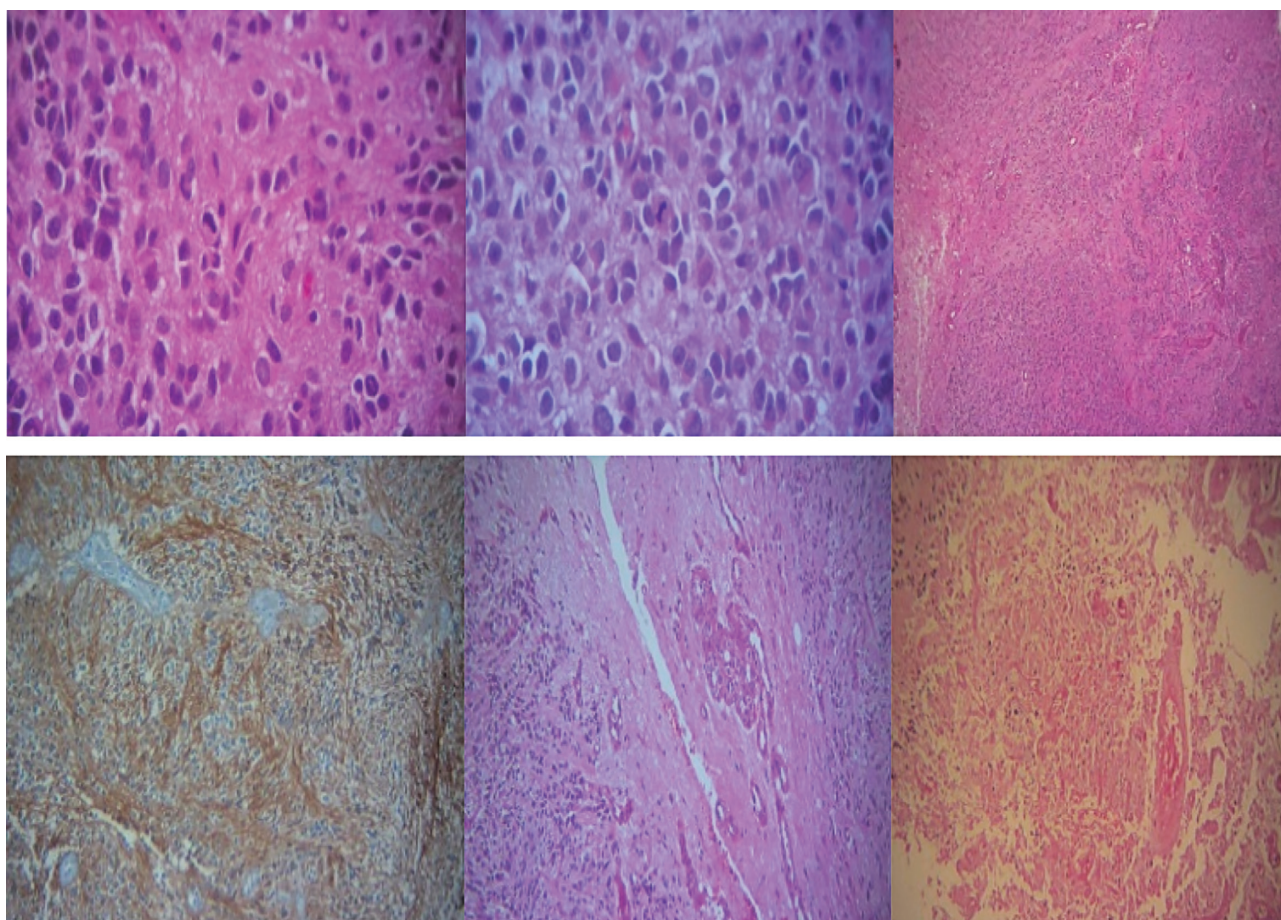


Figure 3. Pathological findings suggestive for anaplastic ependymoma.

treatment in Grade II ependymomas, whereas anaplastic ependymomas need surgical resection accompanied with radiation therapy^{12,15-19}.

REFERENCES

1. Dilli UD, Yıldırım M, Suren D, Turk CC, Goktaş S, Parlak E, et al. Frontal lobe ependymoma: A case report. *Erciyes Med J.* 2013;35:75–8.
2. Hiniker A, Lee HS, Chang S, Berger M, Perry A. Cortical ependymoma with unusual histologic features. *Clin Neuropathol.* 2013;32:318–23.
3. Van Gompel JJ, Koeller KK, Meyer FB, Marsh WR, Burger PC, Roncaroli F, et al. Cortical ependymoma: An unusual epileptogenic lesion. *J Neurosurg.* 2011;114:1187–94.
4. McKeever PE, Boyer PJ. The brain spinal cord and meninges. In: Mills ES, editor. *Sternberg's Diagnostic Surgical Pathology.* 4th ed. New Delhi: Jaypee Brothers Medical Publishers; 2006. pp. 446–51.
5. Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, Burger PC, Jouvet A, et al. The 2007 WHO classification of tumours of the central nervous system. *Acta Neuropathol.* 2007;114:97–109.
6. Roncaroli F, Consales A, Fioravanti A, Cenacchi G. Supratentorial cortical ependymoma: Report of three cases. *Neurosurgery.* 2005;57:E192.
7. Hegyi L, Peston D, Theodorou M, Moss J, Olver J, Roncaroli F. Primary glial tumor of the retina with features of myxopapillary ependymoma. *Am J Surg Pathol.* 2005;29:1404–10.
8. Busse C, Nazeer T, Kanwar VS, Wolden S, LaQuaglia MP, Rosenblum M. Sacrococcygeal immature teratoma with malignant ependymoma component. *Pediatr Blood Cancer.* 2009;53:680–1.
9. Guerrieri C, Jarlsfelt I. Ependymoma of the ovary. A case report with immunohistochemical, ultrastructural, and DNA cytometric findings, as well as histogenetic considerations. *Am J Surg Pathol.* 1993;17:623–32.
10. Mikami M, Komuro Y, Sakaiya N, Tei C, Kurahashi T, Komiyama S, et al. Primary ependymoma of the ovary, in which long-term oral etoposide (VP-16) was effective in prolonging disease-free survival. *Gynecol Oncol.* 2001;83:149–52.
11. Manasa PL, Uppin MS, Sundaram C. Analysis of squash smear cytology of ependymomas. *Acta Cytol.* 2012;56:183–8.

12. Lehman NL. Central nervous system tumors with ependymal features: A broadened spectrum of primarily ependymal differentiation? *J Neuropathol Exp Neurol*. 2008;67:177–88.
13. Lehman NL. Patterns of brain infiltration and secondary structure formation in supratentorial ependymal tumors. *J Neuropathol Exp Neurol*. 2008;67:900–10.
14. Burger PC. Ependymoma. In: Burger PC, editor. *Diagnostic Pathology Neuropathology*. 1st ed. Canada: Amirsys Publisher; 2012. pp. 96–113.
15. Yurt A, Selçuki M, Ertürk AR, Küpelioglu A. Large supratentorial cortical ependymoma in a child. *Clin Med Res*. 2010;8:25–7.
16. Lehman NL, Jordan MA, Huhn SL, Barnes PD, Nelson GB, Fisher PG, et al. Cortical ependymoma. A case report and review. *Pediatr Neurosurg*. 2003;39:50–4.
17. Lum DJ, Halliday W, Watson M, Smith A, Law A. Cortical ependymoma or monomorphous angiocentric glioma? *Neuropathology*. 2008;28:81–6.
18. Miyazawa T, Hirose T, Nakanishi K, Uozumi Y, Tsuzuki N, Shima K. Supratentorial ectopic cortical ependymoma occurring with intratumoral hemorrhage. *Brain Tumor Pathol*. 2007;24:35–40.
19. Mork SJ, Loken AC. Ependymoma: A follow-up study of 101 cases. *Cancer*. 1977;40:907–15.