

Pure cortical supratentorial extraventricular ependymoma

To the Editor,

We read the report by Yadav *et al.*, describing a case of “pure cortical ependymoma”.^[1] The authors described a case of a young boy with a short duration of raised intracranial pressure and progressive hemiparesis. Only a computed tomography scan was available which showed a solid cystic mass with the solid component and thick cyst wall showing significant enhancement and surrounding perilesional edema. The boy was treated with near total excision and subsequent follow-up withholding adjuvant therapy. Histology revealed a classical Grade 2 ependymoma. The authors contend that this represented a case of “pure cortical ependymoma”. There are, however, concerns regarding labeling this case as such.

Cortical ependymoma (CE) is a nomenclature reserved for a very small subset of supratentorial extra-ventricular ependymomas which are exclusively restricted to the cortex as the name suggests.^[2] This nomenclature excludes extraventricular supratentorial ependymomas which arise in the white matter and reach up to the cortex. In large tumors it may often be difficult to make this differentiation and such tumors should not be called cortical ependymomas. The case by Yadav *et al.*, was a large tumor which (as evident in the pictures provided) involved the cortex but extended into the white matter too. The authors report five cases of CE reported in literature. However, a few more cases have been described.^[3-5] Almost all the CEs reported have been well-circumscribed small cortical lesions. Magnetic resonance imaging (MRI) features though not diagnostic would have helped ascertain the site of origin of the tumor in this case. This again highlights the importance of a complete preoperative MRI in patients with brain tumors and reiterates its importance in the optimal management of such patients, especially when the pathological diagnosis needs to be interpreted in the light of the imaging findings. This is exemplified in the case of CEs where histologically they share the same features as any other supratentorial ependymoma. It is only the site of the tumor which confers upon it a unique biological behavior. The clinical presentation of the case in discussion is also atypical for a CE. The CEs reported so far have all presented with longstanding seizures.^[2] The boy in the case described had a rather short and progressive history of raised pressure symptoms and neurological deficits, both of which

are odd for a very low-grade lesion of the type of a CE. Intratumoral hemorrhage may cause an acute deterioration,^[5] however, the case described did not show any hemorrhage. Supratentorial ependymomas on the other hand, especially the malignant ones may have an acute presentation. Histologically, there is little to differentiate CE from other classical ependymomas. Often it may be difficult to differentiate CE from astroblastomas. Electron microscopy may be helpful.^[2-4] Lum *et al.*,^[4] have recently reported one more case of a CE in which they discuss the similarities of CE with the newly designated entity of angiocentric glioma which in fact has been accorded a World Health Organization grade of 1.^[6] In fact Lehman suggests that the two could be variants of the same ependymal tumor group.^[3] All the available evidence points towards a very benign course for CE, unlike other supratentorial ependymomas. Their indolent behavior, however, contrasts with the often poor outcomes seen in other supratentorial ependymomas. Hence it is imperative to be cautious in labeling a case as a CE. Whereas follow-up after a radical excision is appropriate in CEs, adjuvant radiation would be indicated in other supratentorial ependymomas (especially with residue) as was the case in this report.

We feel that the case described was a supratentorial extra-ventricular ependymoma and not a true cortical ependymoma. Given the clinical presentation and imaging findings we would have favored adjuvant radiotherapy, especially considering the residue.

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References

1. Yadav YR, Neha, Chandrakar SK. Pure cortical supratentorial extraventricular ependymoma. *Neurol India* 2009;57:213-5.
2. Roncaroli F, Consales A, Fioravanti A, Cenacchi G. Supratentorial cortical ependymoma: Report of three cases. *Neurosurgery* 2005;57:E192; discussion E192.
3. Lehman NL. Patterns of brain infiltration and secondary structure formation in supratentorial ependymal tumors. *J Neuropathol Exp Neurol* 2008;67:900-10.
4. Lum DJ, Halliday W, Watson M, Smith A, Law A. Cortical ependymoma or monomorphous angiocentric glioma? *Neuropathology* 2008;28:81-6.
5. 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.
6. 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

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