They rarely arise within the cerebral cortex where they are known as cortical ependymomas (CEs) and often present with seizures. Pediatric supratentorial CEs are extremely rare; only 17 pediatric patients, including ours, have been reported to date. We encountered a 10-year-old boy with CE presenting with seizures and discuss its clinicopathological features.

**CASE REPORT**

This 10-year-old boy first suffered seizures in 2007 when he was 5 years old. They started on the left side of his face and be-
came generalized. Cranial magnetic resonance imaging (MRI) showed a 14-mm diameter cortical lesion in the right postcen-
tral gyrus (Fig. 1A, B). It was slightly hypointense on T1-weighted MRI, hyperintense on fluid-attenuated inversion recovery images and not Gd-contrast-enhanced. The preliminary differential diagnosis included DNT, ganglioglioma, low-grade glioma, and cortical dysplasia but not ependymoma. He was placed under observation because the tumor was thought to be benign. No anticonvulsants were prescribed because his electroencephalogram was normal. Repeat MRI studies performed 2 years later revealed no changes in the size and radiological features of the tumor. He was not seen in the next 3 years because he suffered no further seizures. However, in 2012 when he was 10 years old he again had generalized seizures. MRI showed a 40 mm diameter parietal tumor that was heterogeneously Gd enhanced (Fig. 1C, D). As there was radiographic evidence suggestive of malignant transformation we chose to remove the tumor.

After right frontoparietal craniotomy we observed a reddish-gray tumor on the surface of the postcentral gyrus; it was not covered by brain parenchyma (Fig. 2). It was well demarcated from surrounding brain tissue, had no attachment to the lateral ventricle, and was gross-totally removed. Histologically, the tumor cells were arranged around vessels. Their radially-oriented
cell processes were directed at the vessels, thereby producing the perivascular pseudorosettes typical of ependymoma (Fig. 3A). In some areas there were oligodendroglioma-like cells with round nuclei and clear cytoplasm. No true ependymal rosettes were observed. We noted the intermingling of an increased number of abnormal vessels but no other anaplastic features. Immunohistochemically the tumor cells were positive for glial fibrillary acidic protein (GFAP) (Fig. 3B), S-100 protein, and epithelial membrane antigen (EMA) (Fig. 3C). The MIB-1 labeling index was 6.4%. Electron microscopy confirmed the typical presence of microvilli (Fig. 3D). These additional findings confirmed our histopathological diagnosis of ependymoma, World Health Organization (WHO) grade II.

The patient made a good recovery and manifested no neurological deficits. We prescribed carbamazepine and he suffered no further seizures. On follow-up brain MRI scans there was no evidence of residual tumor. Neither adjuvant radiotherapy nor chemotherapy was delivered because the tumor had been resected gross-totally and exhibited no anaplastic features on histopathologic study. Follow-up brain MRI performed 6 and 12 months after the operation yielded no evidence of tumor recurrence.

DISCUSSION

Tumors are commonly the cause of seizures in pediatric patients. Prayson reported that the most common tumors encountered in this population included ganglioglioma (37.2%), DNT (13.2%), low-grade fibrillary astrocytoma (11.6%), and oligoastrocytoma (6.2%); he identified coexisting malformations of cortical development or cortical dysplasia in one-third of patients with these tumors. When we encounter pediatric supratentorial cortical mass lesions presenting with seizures, our differential diagnosis usually includes ganglioglioma, DNT, low-grade glioma, and cortical dysplasia but not ependymoma. Ependymomas comprise 2–9% of all neuroepithelial neoplasms, 40% are supratentorial and the others are infratentorial. Posterior fossa ependymomas are seen mostly in children, supratentorial ependymomas (STE)s tend to arise in older age groups. Ependymomas tend to arise in the lateral or fourth ventricles. Ectopic ependymomas at sites distant to the ventricular system are relatively rare and they are seldomly seen in the cerebral cortex where they are known as CEs. Hypotheses for the ectopic origin of ependymomas suggest their development from a heterotopic ependymal nest due to anomalous migration from the germinal matrix; glial cells with progenitor cell properties have also been proposed as the source of these tumors.

Pediatric supratentorial cortical mass lesions presenting with seizures are often followed without resection because they rarely progress. However, as evidenced in our patient, CEs must be followed carefully to alert to tumor progression. Ohwaki et al. reported a child with a parietal cortical mass measuring 8 mm in diameter whose size remained stable for 16 months and then grew to 4 cm in the course of 8 months. The histological diagnosis was ependymoma (WHO grade II) although the MIB-1 labeling index was high at 27.5%. Takeshima et al. encountered an adult with a small cortical mass in the frontal lobe whose size remained stable for 2 years before turning into a huge tumor during the subsequent 18 months. The histological diagnosis was anaplastic ependymoma: the MIB-1 labeling index was 20%. They concluded that the rapid tumor growth was due to malignant transformation. Although CEs may progress, compared to other STEs their prognosis is relatively favorable, possibly because of their early detection due to the elicitation of seizures, the possibility of complete resection facilitated by their superficial location, their distinct tumor margin, and their tendency to be histologically benign.

CONCLUSION

We reported a pediatric patient with supratentorial CE. In the differential diagnosis of pediatric supratentorial cortical mass lesions presenting with seizures, ependymoma should be considered and these tumors must be followed carefully because they may progress. The prognosis of these tumors is favorable if they are diagnosed early and accurately and if they are addressed properly.
References

1. Afra D, Müller W, Slowik F, Wilcke O, Budka H, Tiurczy L: Supratentorial leioh ependymomas: reports on the grading and survival periods in 80 cases, including 46 recurrences. Acta Neurol (Wien) 69: 243-251, 1983

2. Fujimoto K, Ohnishi H, Koshimaa N, Ida Y, Kanemoto Y, Motoyama Y, et al.: Brain surface clear cell ependymoma: case report. No Shinkei Geka 27: 843-846, 1999

3. Ghanis A, Abdullah JM, Ghazali M, Ahmad F, Ahmad KA, Madhavan M: Recurrent paediatric supratentorial extraventricular ependymoma associated with genetic mutation at exon 4 of p53 gene. J Korean Neurosurg Soc 50: 150-153, 2010

4. Hiniker A, Lee HS, Chang S, Berger M, Perry A: Cortical ependymoma with unusual histologic features. Clin Neuropathol 32: 318-323, 2013

5. Lehman NL: Patterns of brain infiltration and secondary structure formation in supratentorial ependymal tumors. J Neuropathol Exp Neurol 67: 900-910, 2008

6. Lehman NL, Jordan MA, Huhn SL, Barnes PD, Nelson GB, Fisher RG, et al.: Cortical ependymoma. A case report and review. Pediatr Neurosurg 39: 50-54, 2003

7. Lellouch-Tubiana A, Boddart N, Bourgeois M, Fohlen M, Jouvet A, Delalande O: Angiocentric neuroepithelial tumor (ANET): a new epilepsy-related clinicopathological entity with distinctive MRI. Brain Pathol 15: 281-286, 2005

8. Liu Z, Li J, Liu Z, Wang Q, Famer P, Mehta A, et al.: Supratentorial cortical ependymoma: case series and review of the literature. Neuropathology 34: 243-252, 2014

9. Liu NJ, Halliday W, Watson M, Smith A, Law A: Cortical ependymoma or monomorphic angiocentric glioma? Neuropathology 28: 81-86, 2008

10. Massimino M, Buttarelli FR, Antonelli M, Gandola L, Modena P, Giangaspero F: Intracranial ependymoma: factors affecting outcome. Future Oncol 5: 207-216, 2009

11. Miyazawa T, Hirose T, Nakanishi K, Uozumi Y, Tsuzuki N, Shima K: Supratentorial ectopic cortical ependymoma occurring with intratumoral hemorrhage. Brain Tumor Pathol 24: 35-40, 2007

12. Molina OM, Collina JL, Luzardo GD, Mender OE, Cardozo D, Velasquez HS, et al.: Extraventricular cerebral anaplastic ependymomas. Surg Neurol 51: 630-635, 1999

13. Nakamizo S, Sasayama T, Kondoh T, Inoue S, Shimori R, Tanaka H, et al.: Supratentorial pure cortical ependymoma. J Clin Neurosci 19: 1453-1455, 2012

14. Oltowski K, Tanishima T, Yoshimasa N, Hojo S, Fujimaki T, Kirino T: Rapidly enlarging supratentorial ependymoma in a child presenting initially with a small calcified lesion: case report. No Shinkei Geka 25: 713-718, 1997

15. Prayson RA: Tumours arising in the setting of paediatric chronic epilepsy. Pathology 42: 426-431, 2010

16. Preusser M, Hoischen A, Novak K, Czech T, Prayer D, Haflinger JA, et al.: Angiocentric glioma: report of clinicopathologic and genetic findings in 8 cases. Am J Surg Pathol 31: 1709-1718, 2007

17. Rigante L, Novello M, Massimi L, Caldarelli M: A cortical cystic epileptogenic lesion: tanyctopic ependymoma. Acta Neurol Belg 113: 523-525, 2013

18. Roncaroli F, Consales A, Fioravanti A, Ceracchi G: Supratentorial cortical ependymoma: report of three cases. Neurosurgery 57: E192: discussion E192, 2005

19. Takeshima H, Kawahara T, Uchiwa H, Hirano S, Nakazato Y, Kuratsu J: Brain surface ependymoma with repeated episodes of intratumoral hemorrhage: case report. Neurol Med Chir (Tokyo) 42: 166-169, 2002

20. Van Gompel JJ, Koeller KK, Meyer FB, Marsh WR, Burger PC, Roncaroli F, et al.: Cortical ependymoma: an unusual epileptogenic lesion. J Neurosurg 114: 1187-1194, 2011

21. Vernet O, Farmer JP, Meagher-Villemeur K, Montes JL: Supratentorial ectopic ependymoma. Can J Neurol Sci 22: 316-319, 1995

22. Vinchon M, Soto-Ares G, Riiffaud L, Ruchoux MM, Dhemelmannes P: Supratentorial ependymoma in children. Pediatr Neurosurg 43: 77-87, 2007

23. Wang M, Tihan T, Rojiani AM, Bodhireddy SR, Prayson RA, Iacuone JJ, et al.: Monomorphic angiocentric glioma: a distinctive epileptogenic neoplasm with features of infiltrating astrocytoma and ependymoma. J Neuropathol Exp Neurol 64: 875-881, 2005

24. Yadav YR, Neha, Chandrakar SK: Pure cortical supratentorial extraventricular ependymoma. Neurol India 57: 213-215, 2009

25. Yurt A, Selçük M, Ertürk AR, Kipelioğlu A: Large supratentorial cortical ependymoma in a child. Clin Med Res 8: 25-27, 2010