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## Case Report

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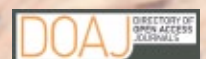
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# Case Report

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## ■ Case Report

An 11-year-old girl visited the Department of Neurology, complaining of increased frequency of complex partial seizures. She had an 8-years history of partial epilepsy.

Neurological examination showed no focal or false-localizing signs. Examination of other systems was unremarkable. A magnetic resonance imaging (MRI) revealed a well-circumscribed right-sided frontal lesion. The lesion was diffusely hyperintense on T1- and T2-weighted images and did not exhibit significant enhancement after Gadolinium administration (Fig. 1A and C). Electroencephalography (EEG) was characteristic with focal epileptic changes in the right frontal region. The index of epileptic discharges was approximately unchanged. Brain 18FDG-positron emission tomography (PET) showed a diffuse hypometabolism of the lesion as it is observed in low-grade tumors (Fig. 1B). Hypometabolism may also show the epileptic focus because PET scan is a good interictal method to localized epileptic foci.

*En-bloc* resection of the right superior frontal gyrus was performed. The resection included the tumor and the surrounding cortex.

The patient remained free from seizures with antiepileptic medications and showed no evidence of tumor recurrence 6 months later.

## ■ Diagnosis

Angiocentric glioma.

Histopathological examination revealed a tumoral proliferation compound of spindle-shaped cells with palisade arrangements (Fig. 2A) and formation of perivascular rosettes (Fig. 2B). Tumor cells have small, round and regular nuclei with fine chromatin without atypia or mitosis.

On immunohistochemistry, the neoplastic cells strongly expressed GFAP (Fig. 2C) and characterized by cytoplasmic dot-

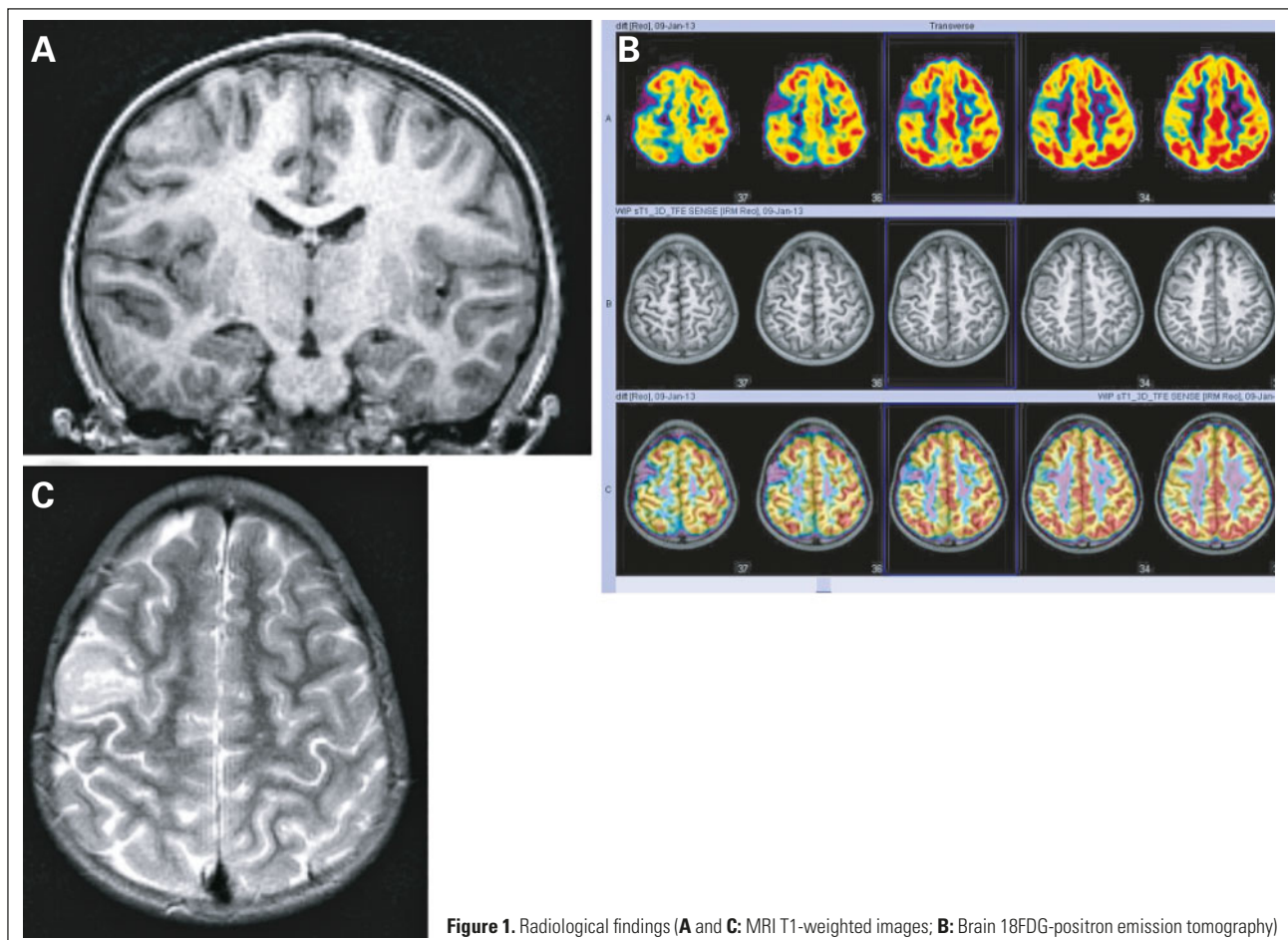
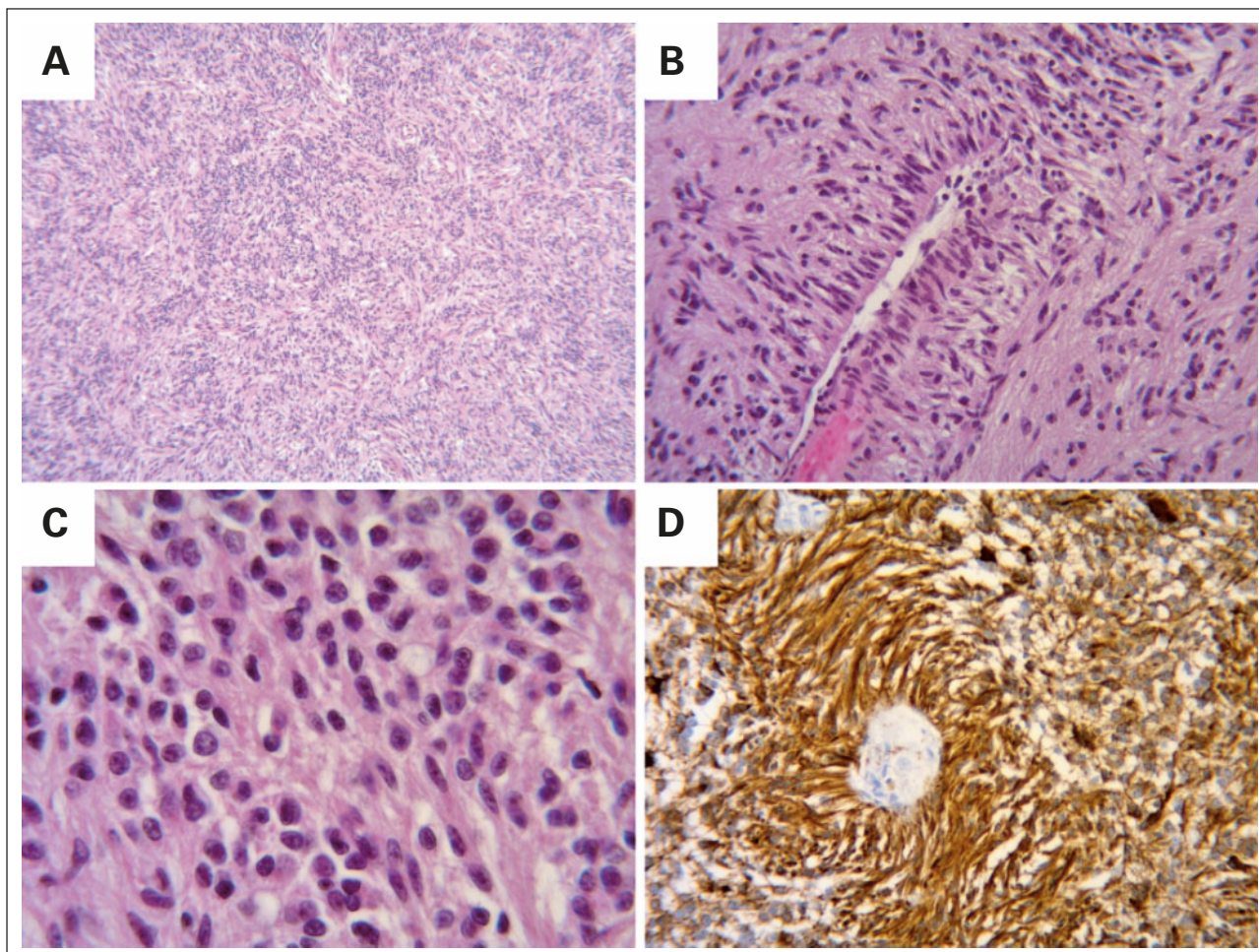


Figure 1. Radiological findings (A and C: MRI T1-weighted images; B: Brain 18FDG-positron emission tomography)



**Figure 2.** Histopathological and immunohistochemical findings (A: HES \*50, B: HES \*200, C: HES \*400 and D: GFAP \*400)

like staining of EMA (epithelial membrane antigen). The Ki-67 labeling index was less than 1% throughout the specimen.

### ■ Comments

The last revised WHO Classification of Tumors of Nervous System (WHO 2007) has introduced this new tumor type and is now included under the category “Other Neuroepithelial Tumors”.

Angiocentric glioma is a rare and slow-growing cerebral hemispheric tumor arising preferably in children and young adults. Approximately 65 cases have been reported in the literature. Presenting symptom is seizure and patients are all intractable to antiepileptic medications. The second most common symptom is headache.

Radiologically, this tumor is solid and well-circumscribed and is generally localized in the cortex of frontal lobe. On MRI, angiocentric glioma appears hypointense on T2-weighted images and FLAIR-hyperintense. This lesion lacks enhancement following contrast administration. Calcifications are rare.

Histologically, they are characterized by monomorphic, bipolar glial tumor cells oriented along vascular structures. The

nuclei of tumor cells are slender with stippled granular chromatin. This lesion does not present high-grade features such as elevated mitotic activity, necrosis and endothelial proliferation.

Cortical dysplasia, associated with other epileptogenic tumors, such as ganglioglioma and dysembryoplastic neuroepithelial tumor, has not been reported in angiocentric glioma.

Immunohistochemically, tumor cells express GFAP, S-100 protein, vimentin and EMA with a dot-like pattern but do not express neuronal markers. Ki-67 labeling index is less than 5%.

Prior to other epilepsy-associated tumors such as ganglioglioma and pleomorphic xanthoastrocytoma, there is no mutation of BRAF gene in angiocentric glioma. Indeed, one recent study failed to reveal any BRAF mutation in three cases and another did not evidence any expression of BRAF V600E on immunohistochemistry in nine tested cases.

Their precise histogenesis remains unclear. Based on the cytology, morphology and immunohistochemistry, ependymal derivation has been postulated.

The treatment of choice is total resection. The prognosis of this tumor is favorable if totally resected. Gross total resection

leads to children becoming seizure free in all documented cases of the literature. Nevertheless, postoperative seizure freedom is not achieved in most of cases with subtotal resection. No recurrence and progression has been reported after gross total or subtotal removal of tumor, except for one patient who died of tumor progression. There are no reported cases of malignant transformation.

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