

## Angiocentric glioma, a recently added WHO grade-I tumor

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Angiocentric glioma (AG) is a distinct group of tumors, grade I according to the 2007 WHO classification of tumors of the central nervous system. It is a rare group predominantly seen in children and young adults, with good prognosis following gross total resection. A young child presented with progressively worsening seizure activity; MRI revealed a lesion involving the left posterior frontal lobe. The lesion was surgically resected, and the child is now free of seizure medications, with good neurological function.

### Introduction

Seizure disorder is one of the less well understood but frequently encountered conditions in children and young adults. Since it has varied etiology, much of which is not known, the treatment is often symptomatic and long term, and progression of the disease to refractory epilepsy is not uncommon. Angiocentric glioma has recently been recognized as a distinct group of tumors presenting with refractory epilepsy as the clinical symptom and predominantly involving children and young adults. The radiological features studied in the described literature have been consistent, and the prognosis following gross surgical resection is very good.

### Case report

A 4-year-old boy presented with progressively worsening seizure activity that included staring episodes, with some limpness on the right side of the body. In the interictal period, he had no weakness or numbness and was neurologically intact.

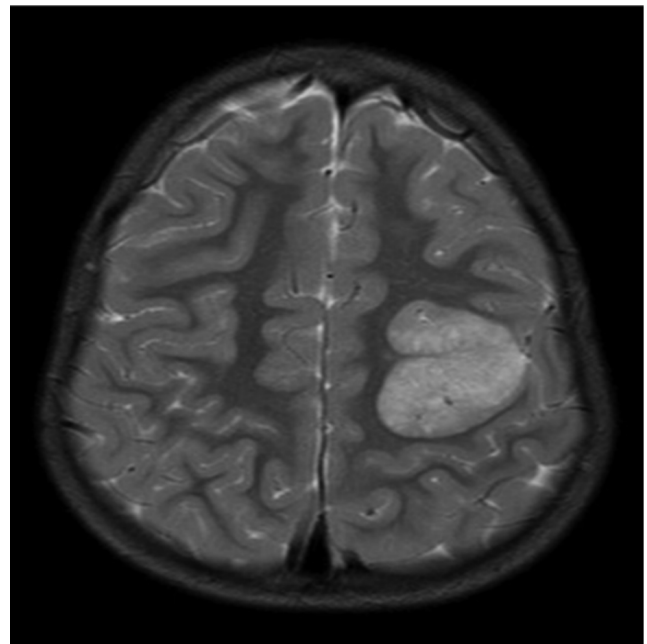


Figure 1. 4-year-old boy with angiocentric glioma. Axial, T2-weighted image shows a hyperintense, expansile, predominantly cortical lesion involving the left posterior frontal lobe.

MRI of the brain revealed a 2.8 x 3.8 x 2.2cm expansile lesion involving the left posterior frontal lobe. The lesion was predominantly cortical, involving the gray matter of the two gyri anterior to the motor cortex. It was predominantly hyperintense on the T2-weighted images (Figs. 1, 2) and hypointense on the T1-weighted images (Fig. 3). It

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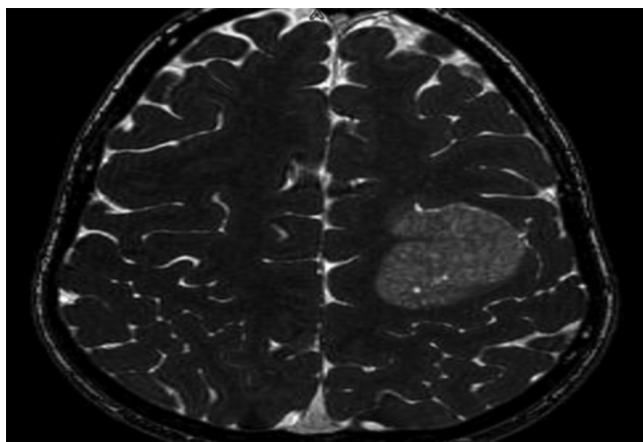


Figure 2. 4-year-old boy with angiocentric glioma. High-resolution, T2-weighted image shows that the hyperintense, expansile lesion is predominantly cortical and lies anterior to the central sulcus.

showed no significant postcontrast enhancement (Figs 4, 5, 6), restricted diffusion, or gradient susceptibility. The lesion had a small stalklike extension that was oriented inferiorly

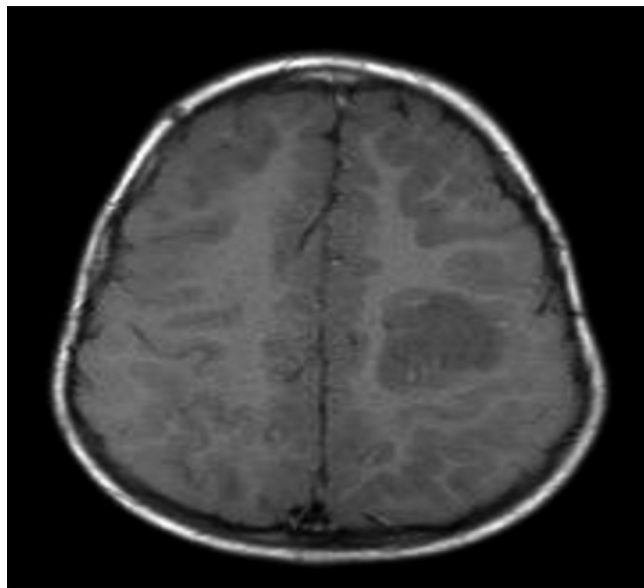


Figure 3. 4-year-old boy with angiocentric glioma. Axial, noncontrast, T1-weighted image shows a hypointense, expansile, cortical-based lesion in the left posterior frontal lobe.

toward the ventricle but did not reach the ventricular/ependymal surface (Figs. 5, 6). On diffusion-tensor imaging, the corticospinal tract (CST) was seen to be displaced posteriorly by this mass lesion (Fig. 7), without significant interruption or loss of volume of the CST.

The mass lesion was surgically resected with left frontal craniotomy, using stealth image guidance, with intraopera-

tive cortex mapping and EMG monitoring. The mass consisted of tan brown, soft, friable tissue.

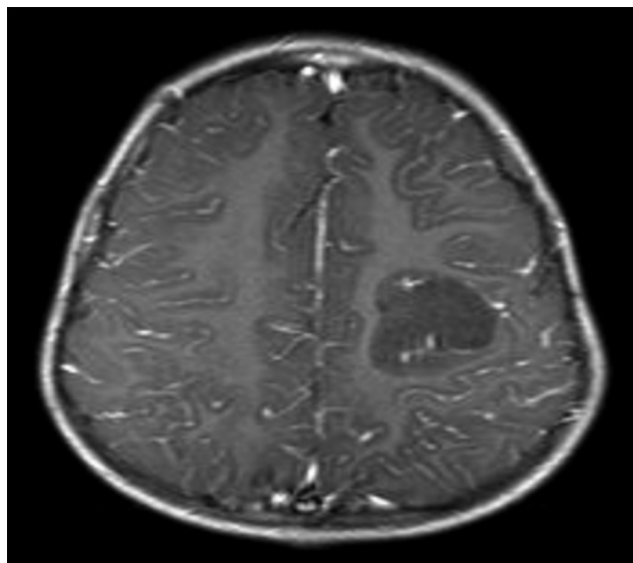


Figure 4. 4-year-old boy with angiocentric glioma. Postcontrast, axial, T1-weighted image shows no significant enhancement in the left posterior frontal lobe lesion.

On pathology, the mass lesion consisted of groups of elongated glial cells with bland cytologic features, forming groups of variable sizes. They showed a tendency to aggre-

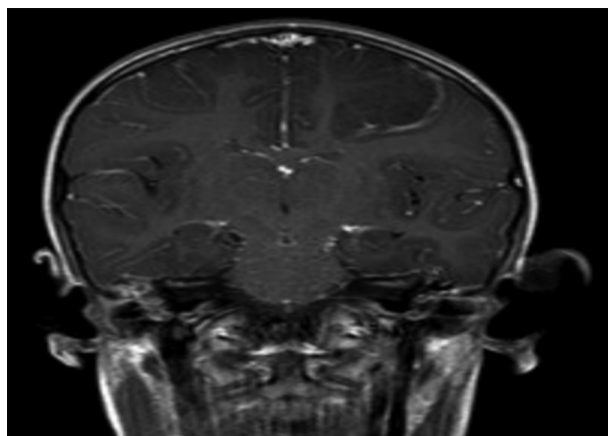


Figure 5. 4-year-old boy with angiocentric glioma. Postcontrast, coronal, T1-weighted image shows no significant enhancement in the left posterior frontal lobe lesion. A small stalklike inferior extension is oriented toward the ventricle but does not reach the ventricular surface.

gate around the blood vessels, forming pseudorosettes. No significant mitosis, necrosis, nor vascular proliferation was seen. The Ki-67 proliferation index was 2-3%. The neoplastic cells were positive for Glial fibrillary acidic protein

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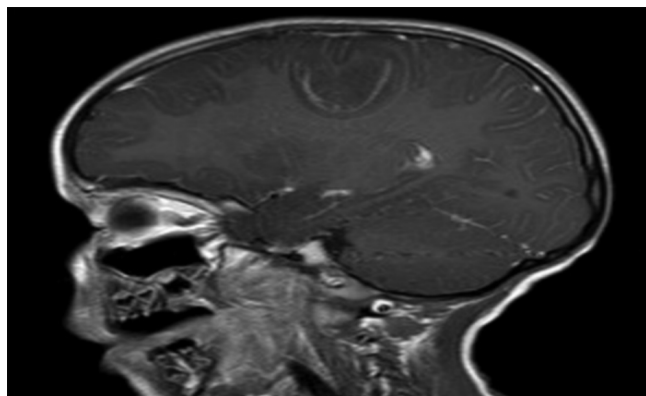


Figure 6. 4-year-old boy with angiocentric glioma. Postcontrast, sagittal, T1-weighted image shows no significant enhancement in the left posterior frontal lobe lesion. A small stalklike inferior extension is oriented toward the ventricle but does not reach the ventricular surface.

(GFAP) and S-100 protein. A subpopulation showed a cytoplasmic-dot-like EMA positivity.

### Discussion

In the 2007 WHO classification of tumors of the central nervous system, the working group proposed eight new entities. Angiocentric glioma (AG) was accepted as a newly identified WHO grade-I tumor (1). AG had been first identified as a distinct lesion in 2005 in two different independent study reports (2, 3).

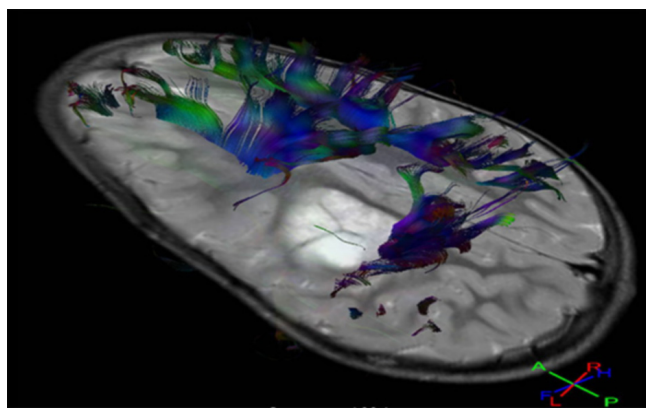


Figure 7. 4-year-old boy with angiocentric glioma. DTI image shows the corticospinal tract displaced posterior to the lesion, without any interruption or significant loss of volume.

AG is seen predominantly in children and young adults. Refractory epilepsy is the leading presentation, and is often drug-resistant. A case report of a patient who presented without seizures was documented by Rho et al (4). These lesions are predominantly supratentorial, are often seen superficially, and have predominant cortical involvement. Involvement of the fronto-parietal cortex, temporal lobe,

and hippocampal region has been documented (1). Involvement of the thalamus has been reported in the series by Marburger et al (5).

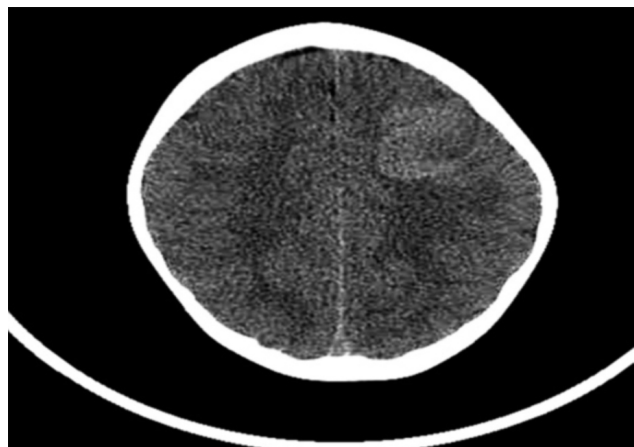


Figure 8. 4-year-old boy with angiocentric glioma. Axial, noncontrast CT image shows that the lesion is iso- to hyperdense, without any calcification.

On MRI images, the different series have reported consistent findings. AGs are T2 hyperintense, T1 hypointense, and show no significant postcontrast enhancement. They are predominantly cortical based and well delineated, and often show a stalklike extension to the subjacent ventricle (6). The case report by Rho et al demonstrated calcifications in their lesion, which is an unusual finding in AG (4). Calvarial remodeling overlying a parietal lobe AG was documented by Pokharal et al (7).

Miyata et al described a rare but distinct clinicopathological subset of AG characterized by adult onset, mesial temporal localization, and epitheloid-cell-predominant histology (8).

Association with coexistent malformation of cortical development was described in the series by Marburger et al (5).

Our case was consistent with the reported MRI appearance, with an expansile, predominantly cortical lesion involving the left posterior frontal lobe, anterior to the motor cortex, with a small stalk extending inferiorly to (but not reaching) the ventricular surface. The lesion was T2 hyperintense, T1 hypointense, and showed no postcontrast enhancement.

AG has been found to have distinct histopathological features, though its cytogenesis is still unclear. Wang et al suggested astrocytic and ependymal lineages (3), whereas Lellouch-Tubiana et al suggested origins from the neurons or radial glia (2).

AG has been observed to be a low-grade glioma with excellent prognosis. The tumors are composed of monomorphous bipolar cells and have an angiocentric growth pattern, with features of astrocytic/ependymal differentiation but no neoplastic neuronal features. They are immunoreactive to epithelial membrane antigen (EMA), GFAP,

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S-100 protein, and vimentin, but not to neuronal antigens (1).

The major radiological differential diagnosis includes dysembryoplastic neuroepithelial tumor (DNET), oligodendrogliomas, and gangliogliomas (9). DNET has a more pronounced, multicystic “bubbly” appearance, may show focal nodules or enhancement, and rarely may show focal calcification. Oligodendrogliomas typically are seen to originate at the gray white junction, often show calcifications, and may show some enhancement. They are seen typically in an older age group. Gangliogliomas are usually slow-growing, well-defined lesions that present with seizures, but these lesions often have enhancing components and focal calcifications, in addition to cystic components.

The role of the radiologist, therefore, is of paramount importance in the workup of a child or young adult presenting with seizures, with a supratentorial, predominantly cortical, nonenhancing, T2-hyperintense lesion. AG needs to be considered in the differential diagnosis of such lesions, as the recognition of this entity suggests a favorable prognosis, with excellent results after gross total resection.

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