Clinical Characteristics and Management of Angiocentric Gliomas: A Case Series of 3 Patients and Review of Outcomes

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INTRODUCTION

Angiocentric gliomas (AGs) are World Health Organization (WHO) grade I gliomas characterized by a perivascular growth pattern and a low-grade mutational profile. Defined in 2007, the literature describing AGs is mostly limited to case reports and reviews.[1] They are generally described as pediatric gliomas that arise in the cerebral hemispheres and present with seizures refractory to medical management.[2]

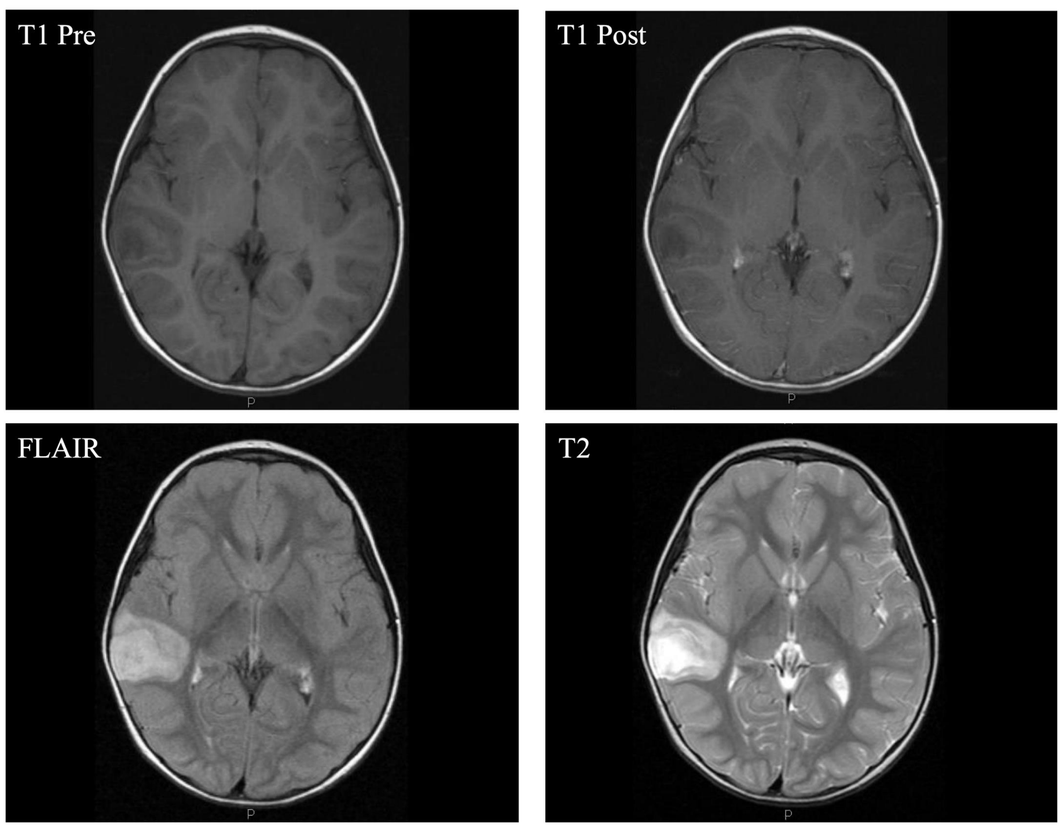
PURPOSE

Given the limited number of cases reported in the literature, we aimed to describe the clinical characteristics and outcomes of three patients with AG treated at our institution. We sought to expand the existing body of literature and inform diagnostic evaluation and treatment decision-making for clinicians faced with a similar picture.

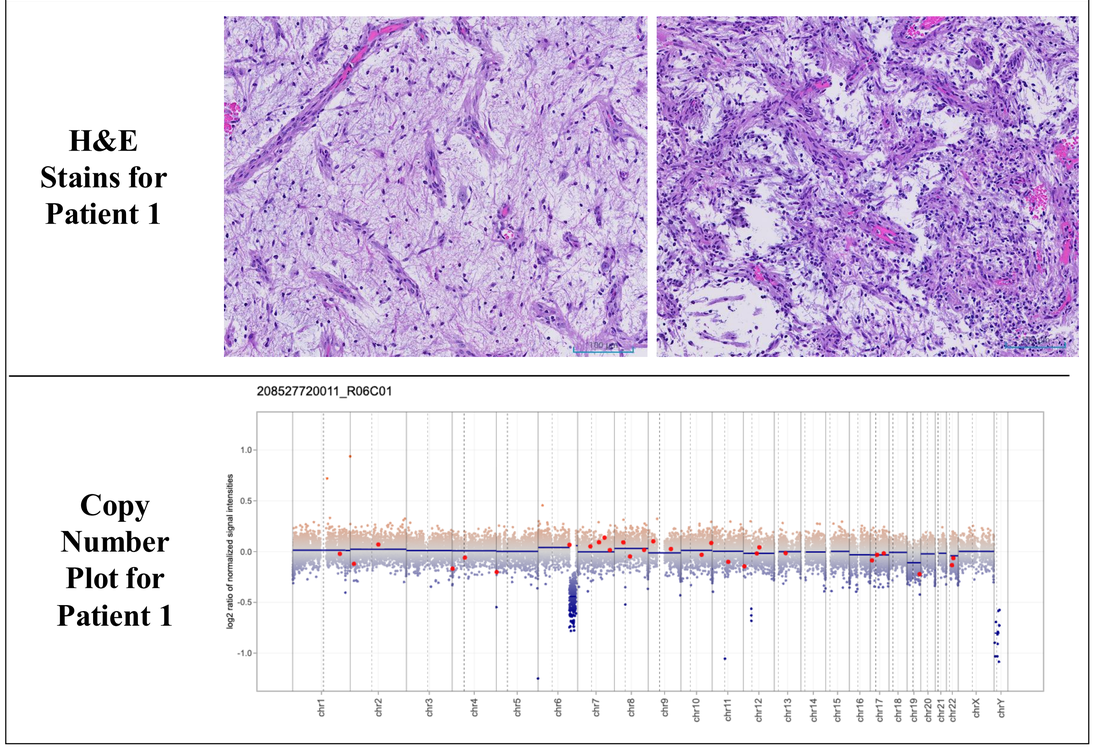
METHODS

We retrospectively identified pathologically confirmed cases of AG treated at our institution from 2012 - 2025 via institutional tumor databases. Inclusion criteria consisted of histopathologic confirmation of AG and availability of clinical and imaging data. We then performed a detailed chart review to extract clinical, radiographic, and pathological characteristics from the electronic health record.

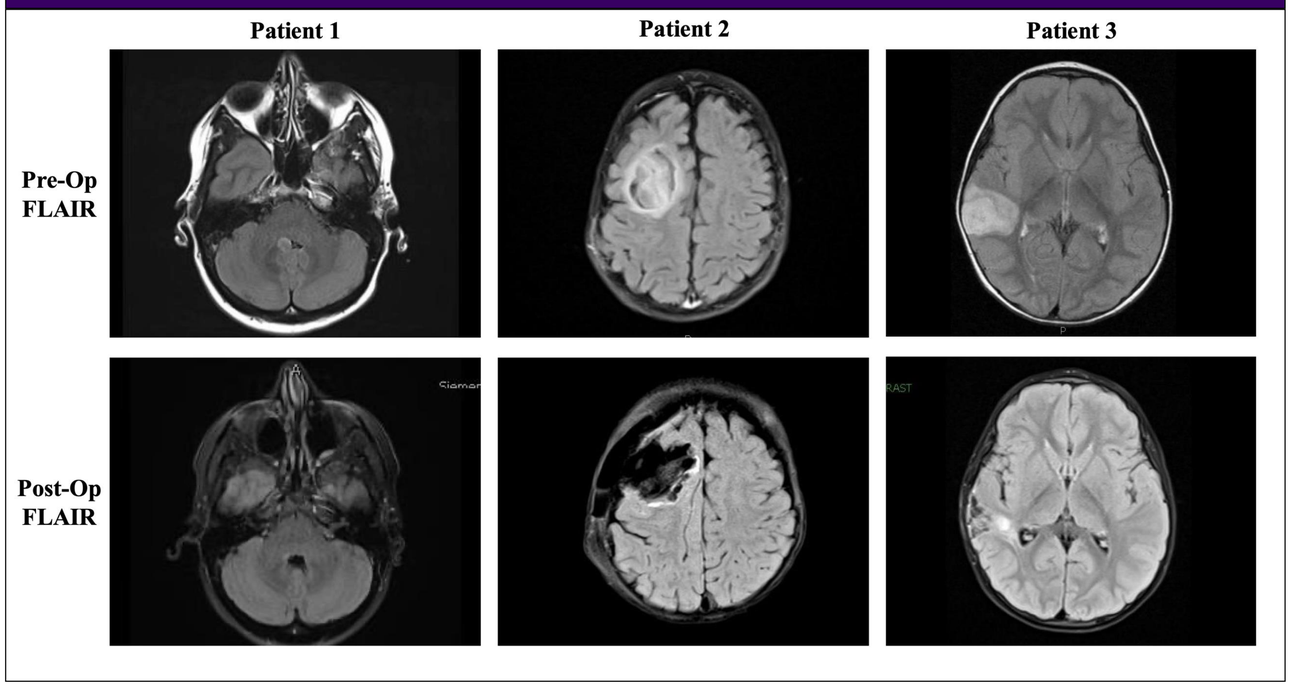
Patient 3 Preoperative Imaging



Patient 1 Notable Pathology



Pre and Post Operative FLAIR Comparison



RESULTS

Patient 1 was a 32-year-old left-handle female who presented with tingling in bilateral fourth toes and a long-term history of headaches. Brain MRI revealed a T1 hypointense, T2 and FLAIR hyperintense, non-enhancing lesion in the floor of the fourth ventricle. Interval growth on follow-up imaging prompted treatment with gross total resection (GTR) via a suboccipital craniotomy. The patient experienced transient diplopia postoperatively and has required no further treatment during the subsequent year of follow up.

Patient 2 was a 2-year-old female with medically managed epilepsy due to cortical dysplasia who presented to the emergency department with breakthrough seizures. Physical examination revealed increased tone and a dysconjugate gaze. Brain MRI showed a cystic lesion with surrounding edema and mass effect; the solid component had imaging characteristics similar to Patient 1's AG. GTR was achieved through a right frontal craniotomy. Despite no recurrence over 9 years of surveillance, seizure control was not achieved, likely due to the superimposed cortical dysplasia.

Patient 3 was a 3-year-old female with no medical history who presented with a generalized tonic-clonic seizure. Brain MRI revealed a temporal lesion without edema or mass effect, exhibiting the same T1 hypointense, T2 and FLAIR hyperintense, and non-enhancing pattern observed in the two previously described tumors. She has experienced nearly 10 years of seizure and disease control through GTR of the lesion.

Histologically, the tumors were defined by bipolar cells growing around blood vessels. The second patient's tumor had the highest Ki-67 index of 10%, while the other two did not exceed 5%. Tumors from the second and third patients exhibited positive GFAP and dot like EMA immunohistochemical expression. Notably, Patient 1's tumor was initially diagnosed as a pilomyxoid astrocytoma; however, the discovery of a MYB (exon 9):QKI (exon 5) fusion suggested AG as a more accurate diagnosis.[3,4] The copy number plot illustrates a microdeletion of chromosome 6q, highlighting a consequence of the MYB:QKI fusion.

CONCLUSION

AGs are a rare subtype of low-grade gliomas that can present with diverse clinical symptoms. Typically affecting pediatric patients with hemispheric lesions and seizures, we reported two such cases. Furthermore, we noted an atypical presentation of a young adult with a fourth ventricular tumor presenting for toe numbness. These tumors respond well to GTR. Further pathological characterization of AGs may inform multimodal treatment for lesions in more eloquent brain regions.

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