

Clinical characteristics, treatment and prognosis of angiocentric glioma

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Received August 30, 2019; Accepted May 18, 2020

DOI: 10.3892/ol.2020.11723

Abstract. Angiocentric glioma (AG) is a rare subtype of neuro-epithelial tumor in children and young adults that commonly presents with seizures. To study the clinical characteristics, treatment and prognosis of patients with AG, the features of two cases of AG were described and 108 cases reported in the literature were assessed. The cases of the present study were two males aged 8 and 16 years, who mainly presented with seizures. MRI revealed superficial, non-enhanced lesions in the left temporal and right frontal lobe, respectively. The two patients underwent gross total resection (GTR) and remained seizure-free without neurological deficits after 3.5 and 2.5 years, respectively. Histopathological examination revealed that the tumors consisted of monomorphous cells that surrounded the blood vessels and neurons in the cerebral cortex, and formed concentric sleeves or pseudorosettes. Furthermore, immunostaining indicated that the diffuse infiltrative neoplastic cells were positive for glial fibrillary acidic protein and a dot-like pattern of epithelial membrane antigen was observed. AG mostly appeared similar to low-grade gliomas on MRI. GTR of the lesions was curative and radiation or chemotherapy were not required. AG typically has a favorable prognosis, with low mortality and incidence of disability.

Introduction

Angiocentric glioma (AG) is a rare central nervous system (CNS) neoplasm that was first reported by Lellouch-Tubiana *et al* (1) and Wang *et al* (2) in 2005. AG was

recognized as a distinct clinicopathologic entity by the World Health Organization (WHO) classification of CNS tumors in 2007 and was defined as 'an epilepsy-associated, stable or slow-growing cerebral tumor primarily affecting children and young adults, histologically characterized by an angiocentric pattern of growth, monomorphous bipolar cells and features of ependymal differentiation (3,4). Since its initial description, an increasing number of cases of AG have been reported in the literature. In the 2016 WHO classification of CNS tumors (5), AG was considered as a WHO grade I tumor and was classified as 'other gliomas'. The majority of studies on AG focus on the cytological features of the disease (6,7), while there is a lack of clinical and imaging data, as well as descriptions of the surgical treatment. The present study describes two patients with AG that received surgical treatment and provides a review of all previously reported cases to date.

Case study

Case 1. The first case was an 8-year-old male who presented with a 3-month history of seizures, headaches and vomiting. The patient was admitted to the Chinese PLA General Hospital (Beijing, China) in June 2016. MRI revealed a left temporal non-enhancing lesion [T1 hypointense, T2 hyperintense, diffusion-weighted imaging hyperintense, fluid-attenuated inversion recovery (FLAIR) hyperintense], measuring 2x2x1.5 cm, with a peripherally enhanced 1x1 mm cystic lesion and obvious brain edema around the lesion. The patient underwent magnetic resonance spectroscopy, which revealed a decrease in the N-acetylaspartate peak and no significant increase in the choline peak (Fig. 1). The patient was then subjected to a left craniotomy and underwent gross total resection (GTR). The tumor was located in the inferior temporal lobe and had a relatively clear boundary. Part of the tumor tissue was fish flesh-like in appearance and the patient had recurrent hemorrhage without vascular changes. Intra-operative frozen histological analysis suggested low-grade glioma. The final pathological assessment (Fig. 2) revealed that tumor cells surrounded the blood vessels and neurons in the cortex. The infiltrating tumor cells were glial fibrillary acidic protein (GFAP)-positive and epithelial membrane antigen (EMA) staining was observed in a distinct dot-like pattern in the cytoplasm. The Ki-67 proliferative rate

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Key words: angiocentric glioma, neuroepithelial tumor, low-grade glioma, children/young adults, pseudorosettes

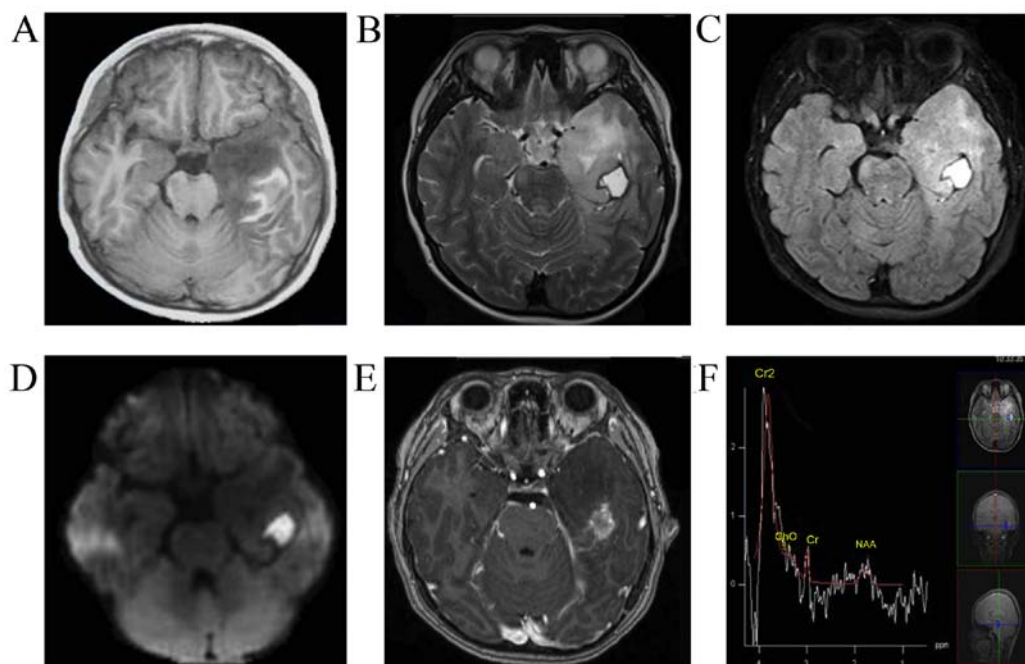


Figure 1. (Case 1) Axial brain MRI displaying a lesion in the left temporal lobe. (A) T1-weighted MRI hypointense. (B) T2-weighted MRI hyperintense. (C) Fluid-attenuated inversion recovery hyperintense with obvious brain edema around the lesion. (D) Diffuse weighted imaging hyperintense. (E) Contrast-enhanced T1-weighted MRI non-enhanced with an enhanced rim. (F) Magnetic resonance spectrum exhibiting a marked decrease in the NAA peak and no significant increase in the Cho peak. NAA, N-acetylaspartate; Cho, choline.

was 5% and the cells were S-100- and neurospecific nucleoprotein (NeuN)-positive, and protein 53 (p53)-, synaptophysin (Syn)-, oligodendrocyte transcription factor-2 (Olig-2)- and creatine kinase (CK)-negative. According the 2016 WHO classification of CNS tumors (3,4), tumors with an angiocentric pattern of growth, GFAP-positive, NeuN- positive and low Ki-67 proliferative rate were diagnosed as AG (WHO grade I). At the 3.5-year follow-up, the patient continued to be seizure-free and did not exhibit any neurological deficits. Post-operative MRI also revealed no recurrence of the tumor.

Case 2. The second case was a 16-year-old male who presented with a 23-day history of recurrent seizures. The patient was admitted to the Chinese PLA General Hospital (Beijing, China) in July 2017. A CT scan revealed a round, circumscribed, hypodense lesion in the right frontal lobe (Fig. 3). MRI revealed a right frontal non-enhanced lesion (T1 hypointense, T2 and FLAIR slightly hyperintense; Fig. 4). Low-grade glioma was diagnosed at the initial stage. The patient then underwent right craniotomy. At the time of this initial surgery, the tumor was fish flesh-like in appearance and soft, had a rich blood supply and was not distinctly different from the surrounding brain tissue. Intra-operative frozen histological analysis also suggested low-grade glioma. The tumor was completely resected as tumor cells were not detected in the surgical margin based on the intra-operative histology. The final pathological assessment (Fig. 5) revealed infiltrating round or ovoid tumor cells in and under the subcortex that were partly arranged around blood vessels and neurons in concentric sleeves and pseudorosettes, demonstrating an angiocentric and creeping pattern. The tumor was dense with irregular cell nuclei and new blood vessels. No mitotic figures or necrotic cells were observed. The Ki-67 proliferative rate was 2%. The tumor

was also immunoreactive for S100, vimentin, neurofilament and NeuN, but was negative for Olig-2, CD3, CD20, CD34 and CD68. These features supported the diagnosis of AG (WHO grade I). At the 2.5-year follow-up, the patient continued to be seizure-free and did not have any complications or neurological deficits. MRI at 5 months post-operatively (Fig. 6) revealed no recurrence of the tumor.

Discussion

A literature review of studies on AG published in English or Chinese between January 2005 and December 2019 (Fig. 7), performed using Medline, PubMed and the China National Knowledge Infrastructure database, revealed that a total of 108 cases, including the two cases of the present study, have been reported (Table SI) (1,2,6-54). Of the 108 patients with AG, 61 were male and 47 were female (male/female ratio, 1:0.77). The age at the time of admission ranged between 1.5 and 83 years (median age, 13 years) and the majority of the patients were children and young adults. A total of 90 patients (85.7%; 90/105) presented with a long history of several types of intractable seizures (symptoms in three cases were not described). Only 15 patients exhibited different symptoms. Of these 15 patients, eight had headaches, four of which had decreased vision (7,12,47), three experienced dizziness, one of which had otalgia (9,19,49), two had ataxia (11,47), two had swallowing disorders (34,37), two presented with weakness and numbness of the left side of the body (17,45) and one had strabismus (34). The majority of the AG tumors (94.4%; 102/108) were in a supratentorial location situated within/under the cerebral cortex and 81.5% (88/108) were located in a single lobe. A total of six tumors were located in the brain-stem (11,34,37,47). In a total of 46 cases, the tumor was in the

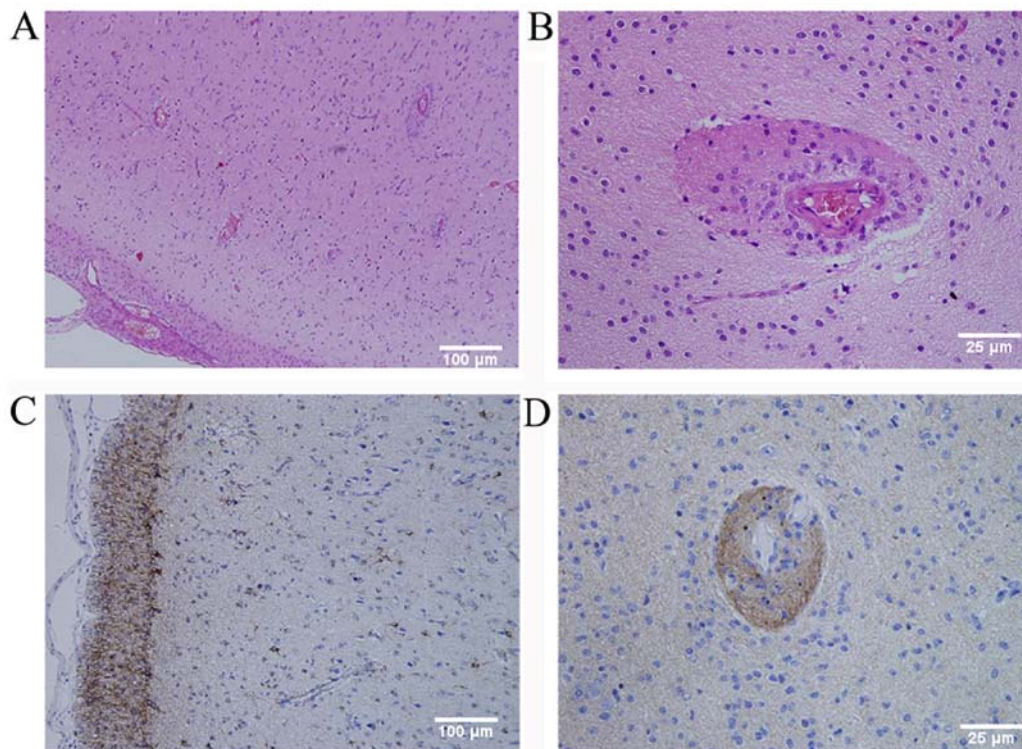


Figure 2. (Case 1) (A and B) Histopathological examination revealed that tumor cells surrounded the blood vessels and neurons in the cortex (H&E staining; original magnification, x100 in A and x400 in B; scale bar, 100 µm in A and 25 µm in B). (C) Immunohistochemical staining demonstrated cytoplasmic immunoreactivity for GFAP (original magnification, x100; scale bar, 100 µm) and (D) dot-like staining for EMA (original magnification, x400; scale bar, 25 µm). GFAP, glial fibrillary acidic protein; EMA, epithelial membrane antigen; H&E, hematoxylin and eosin.

left brain and in 43 cases, it was in the right brain. As AG arises in a superficial location in the cerebrum, which is typically completely removed, the literature indicated that gross total resection (GTR) of this lesion is curative. A total of 64.9% (61/94) of the patients with AG underwent GTR and 27.7% (26/94) underwent subtotal resection (STR). Furthermore, 7 cases only had a biopsy (7,34,37,47). The follow-up time ranged between 0.25 and 168 months (mean, 23.5 months). Of the patients with resection, 93.1% (81/87) were free of seizures during the follow-up time and the 6 patients with seizure recurrence had all undergone STR. A total of 10 patients received adjuvant therapy including radiation or chemotherapy, of which six received STR and four had a biopsy as the lesions were located in the brainstem (6,14,34,37). Histopathological evaluation revealed that the tumors were WHO grade I, with one exception, which was WHO grade III-IV (17).

The present case study reported on two patients that were surgically treated at the Chinese PLA General Hospital (Beijing, China). The cases were similar to the ones reported in the literature, regardless of age, symptoms, tumor location and prognosis. The 8- and 16-year-old patients of the present study presented with intractable seizures and the 16-year-old patient also presented with headaches and vomiting. In the two patients, the tumor was located in the superficial cerebrum, in the left temporal lobe and in the right frontal lobe, respectively. The two patients underwent GTR. During the follow-up period of 42 and 30 months, the patients were seizure-free and did not experience any tumor recurrence.

Only a few studies have reported on the appearance of AG on CT scan (17,19). Hu *et al* (19) reported one case of AG

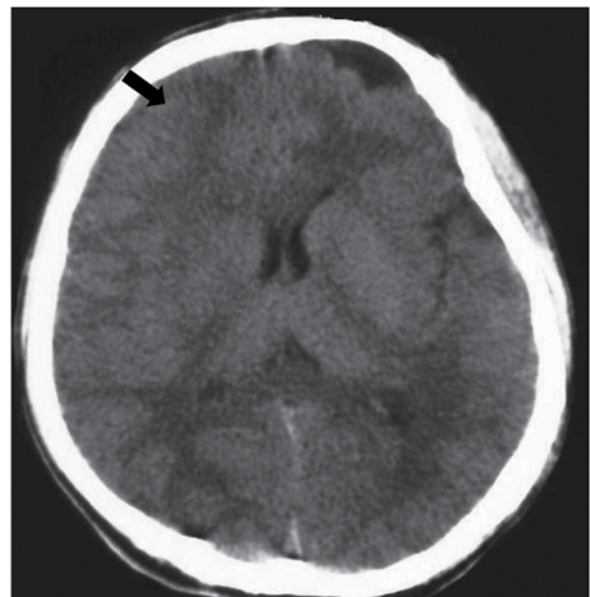


Figure 3. (Case 2) Axial brain CT scan revealed a round, circumscribed, hypodense lesion in the right frontal lobe and an arachnoid cyst in the left frontal lobe (arrow).

that exhibited a hyperdense lesion on CT scan, which may be linked to hemorrhage and hemosiderin deposition. The 16-year-old patient in our cases showed a hypodense lesion on CT scan. The majority of AGs are similar to low-grade gliomas on MRI, i.e., hypointensity on T1-weighted imaging, hyperintensity on T2-weighted imaging, hyperintensity on

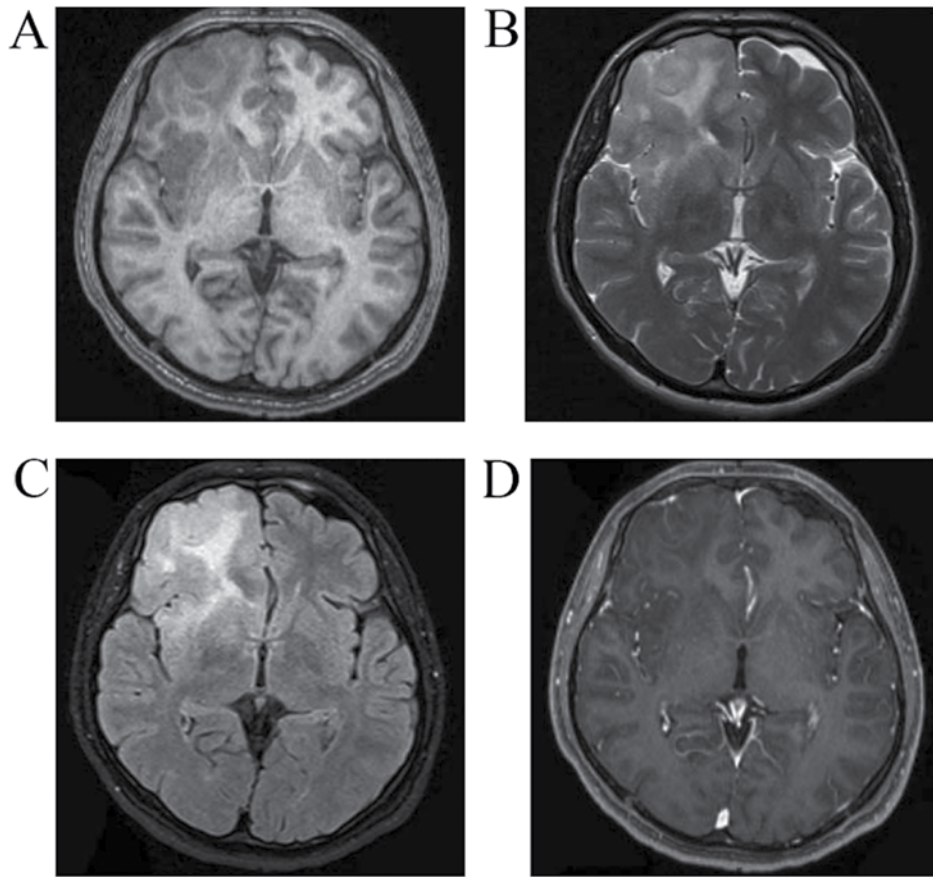


Figure 4. (Case 2) Axial brain MRI indicating a lesion in the right frontal lobe. (A) T1-weighted MRI hypointense; (B) T2-weighted MRI slight hyperintense. (C) Fluid-attenuated inversion recovery slight hyperintense with obvious brain edema around the lesion. (D) Contrast-enhanced T1-weighted MRI non-enhanced.

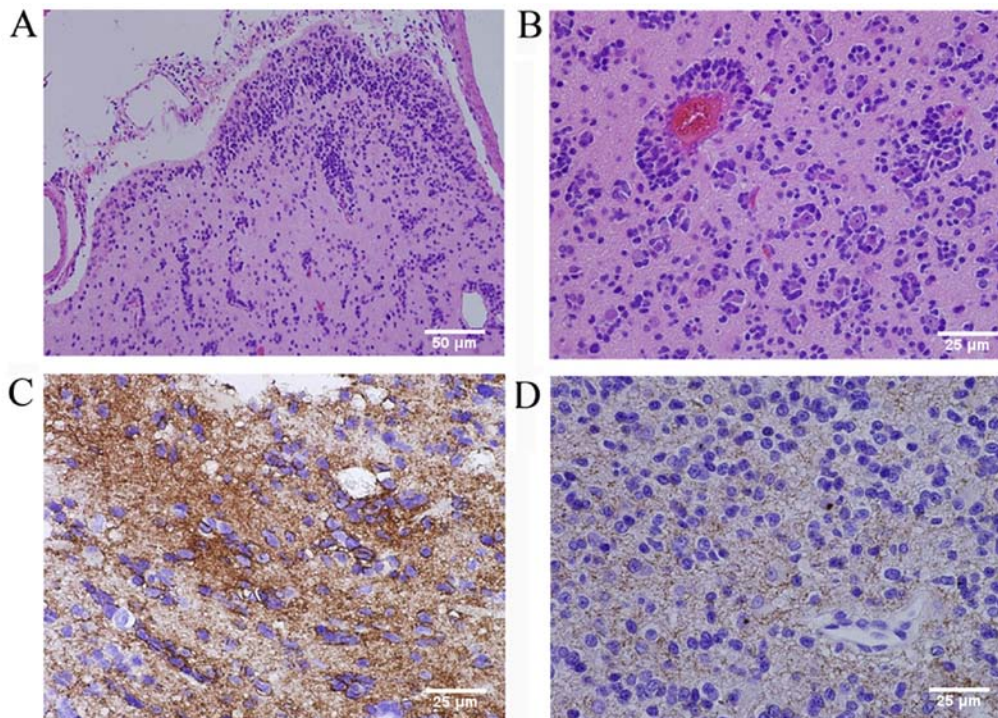


Figure 5. (Case 2) Histopathological examination revealed infiltrative round or ovoid tumor cells in and under the cortex, and partly arranged around blood vessels and neurons in concentric sleeves and pseudorosettes and demonstrated an angiocentric and creeping pattern (H&E staining; original magnification, x200 in A and x400 in B; scale bar, 50 μ m in A and 25 μ m in B). (C) Immunohistochemical staining demonstrated strong cytoplasmic immunoreactivity for GFAP (original magnification, x400; scale bar, 25 μ m) and (D) dot-like staining for EMA (original magnification, x400; scale bar, 25 μ m). GFAP, glial fibrillary acidic protein; EMA, epithelial membrane antigen; H&E, hematoxylin and eosin.

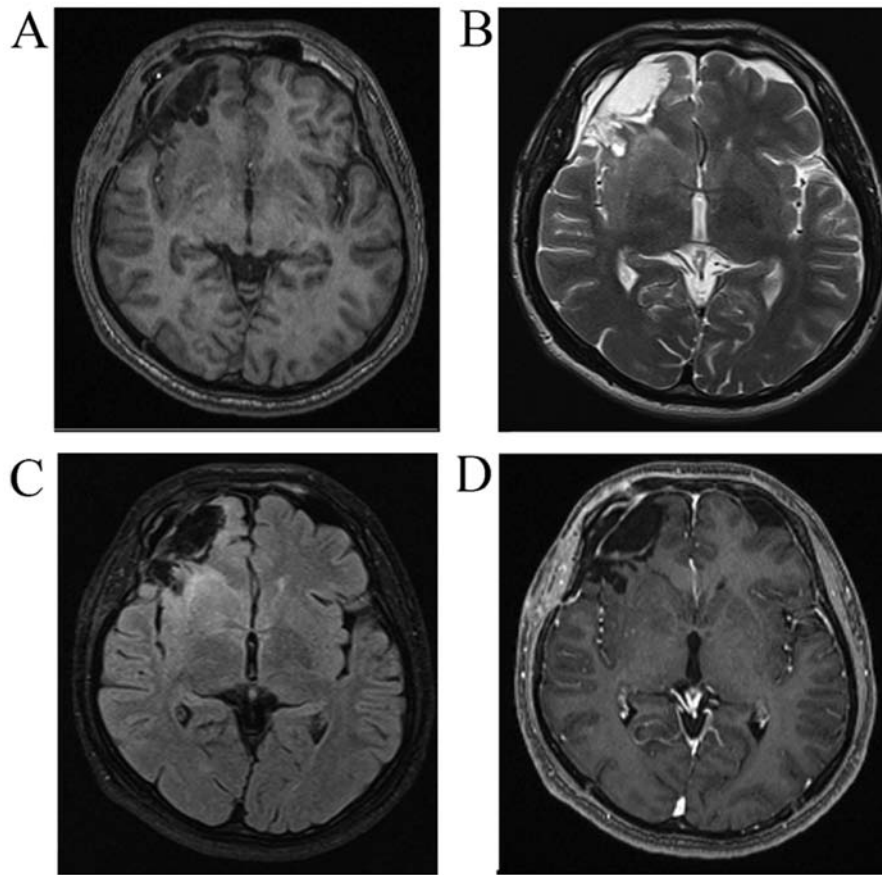


Figure 6. (Case 2) Axial brain MRI at 5 months post-operatively revealing no recurrence of the tumor in the right frontal lobe. (A) T1-weighted MRI. (B) T2-weighted MRI. (C) Fluid-attenuated inversion recovery. (D) Contrast-enhanced T1-weighted MRI.

FLAIR and non-enhanced on the contrast. AG tumors have a well-delineated boundary but may grow diffusely without an obvious edge. Previous studies reported that AG had a cortical rim of hypointensity on T1 and T2 around the lesion, which may be related to long-term compression by the slow-growing low-grade AG (1,14,19). The two patients in the present study also revealed similar characteristics to low-grade glioma, and a rim around the lesion was seen in the second patient.

As there is a lack of specific clinical manifestations and radiological features for AG, diagnosis still depends primarily on histopathological examination. Monomorphic, diffusely infiltrating bipolar spindled cells are commonly arranged around cortical blood vessels or neurons in concentric sleeves and pseudorosettes, which was the typical angiocentric gliomas pattern (15). Immunohistochemical staining results are generally positive for GFAP, S-100 and vimentin, and EMA has a dot-like pattern (8). Based on these findings, the main entities that were considered in the differential diagnosis were ependymoma and AG (6). The neuronal markers NeuN, Syn and chromogranin A (CgA) were usually negative. The Ki-67 proliferative index was ~1% and not >5%. This tumor type is also similar to other benign brain tumor types, including focal cortical dysplasia, ganglioglioma and certain neuroepithelial neoplasms, including pilomyxoid astrocytoma and supratentorial cortical ependymoma. Differences between AG and various easily misdiagnosed brain tumors are presented in Table I (5,55-61).

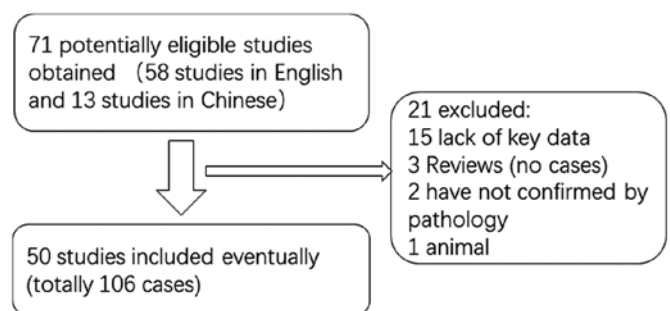


Figure 7. Flowchart of the selection and identification process of the cases included in the present study. After careful screening, 50 studies that included 106 cases were evaluated.

While AG has been established as a distinct tumor type, its cytogenesis remains elusive. Wang *et al* (2) posited that AG arises from ependymoma and astroblastoma. Lellouch-Tubiana *et al* (1) suggested that AG has radial glia or neuronal origin. Bandopadhyay *et al* (62) and Qaddoumi *et al* (63) observed that an myeloblastosis quaking (MYB-QKI) gene rearrangement occurred in the majority of tumors, which contributed to the tumorigenesis, and this rearrangement was specific to angiocentric gliomas. Therefore, MYB-QKI gene fusion may be a defining genetic alteration typical for AG. While mutations of isocitrate dehydrogenase-1 most commonly result in the replacement of arginine

Table I. Differential diagnosis between AG and other types of brain tumor.

Tumor type	Predilection age	Symptoms	Tumor location	Radiological features	Histopathological examination	Immunoreactive factors	WHO grades	(Refs.)
AG	Children and young adults	Epilepsy	Frontal and temporal lobe	Similar to low-grade glioma, non-enhanced on the contrast MRI, small mass	Angiocentric pattern of growth, monomorphous bipolar cells	GFAP, S-100, vimentin, EMA	I	(5,55)
Focal cortical dysplasia	Early childhood	Intractable epilepsy	Frontal lobe	Normal or increased cortical thickness, subtle changes in the smoothness of gyri or sulci, not enhanced with gadolinium	Heterotopic neurons in white matter, hypertrophic neurons and abnormal dendrites	None	None	(58)
Gangliogliom	Children and young adults	Chronic seizures and iCP	Temporal lobe and cerebellum	Nodular, cystic, rim-like or entirely solid, enhanced on contrast MRI	Mixed population of ganglion and glial cells	GFAP, NeuN, NF	I	(59)
Pilomyxoid astrocytoma	Children	Focal neurological deficits and iCP	Suprasellar-hypothalamic region and cerebellum	Bordered lesion with heterogeneous contrast enhancement	Prominent mucoid matrix and angiocentric arrangement of monomorphous, bipolar tumor cells	GFAP, S-100, NSE, Olig2	II	(60)
Supratentorial cortical ependymoma	Adults	Epilepsy and focal neurological deficits	Frontal and parietal lobe	Solid or mixed solid/cystic appearance, enhanced in a heterogeneous pattern on the contrast MRI, mostly larger than 4 cm	Perivascular pseudorosettes and to a lesser extent in true ependymal rosettes	MIB-1, L1 and Ki-67	II-III	(56)
Astroblastoma	Children and young adults, female	Seizures, focal neurologic deficits and iCP	Occipital and frontal lobe	Large, well-demarcated, lobulated mass, solid and cystic components with a characteristic bubbly appearance, with inhomogeneous contrast enhancement	Perivascular pseudorosettes and prominent perivascular hyalinization	GFAP, vimentin	Not established	(57)
Diffuse gliomas	Adults and elderly individuals	Seizures, focal neurologic deficits, cognitive dysfunction and iCP	Cerebral hemisphere	Non-enhanced in diffuse low-grade gliomas, garlanded enhancement with a heterogeneous pattern and cystic appearance in high-grade gliomas on contrast MRI	Diffusely infiltrating, necrosis, cell nucleus dissociation and hemorrhage	GFAP, S-100, Olig2, EMA, nestin and high Ki-67	II-IV	(5,61)

Characteristics listed above are for the majority. Diffuse gliomas mainly include oligodendroglioma, astrocytoma, anaplastic glioma and glioblastoma. iCP, increased intracranial pressure; AG, angiocentric glioma; WHO, World Health Organization; GFAP, glial fibrillary acidic protein; EMA, epithelial membrane antigen; NF, neurofilament; NeuN, neurospecific nucleoprotein; NSE, neurospecific enolase; Olig2, oligodendrocyte transcription factor-2; MIB-1, mind bomb enzyme 3 ubiquitin protein ligase 1; L1, ligand 1.

at position 132 by histidine (R132H) in WHO grade II and III diffuse gliomas and secondary glioblastomas, this was not identified in AG (20,34,43). Therefore, this may facilitate the differential diagnosis of AG from tumors with a higher potential for recurrence.

AG is a slow-growing, stable tumor and lesions in the cerebral cortex are generally benign and may be cured by surgical excision alone. Adjuvant therapy, including chemotherapy or radiation, are not typically required (45). However, angiocentric gliomas arising in structures that are not amenable to surgical resection, including the brainstem, may require stereotactic biopsies and adjuvant therapy (37). Seizure control was dependent on the degree of tumor resection. According to the present review, AG is associated with a more favorable prognosis, with low mortality and incidence of disability. In two cases of tumor recurrence (2,42), the histopathological evaluation revealed a malignant neoplasm (WHO grade III) that was ultimately fatal.

AG is a recently described rare tumor of the CNS and exhibits radiological features on MRI that usually resemble those of diffuse low-grade glioma. AG tends to be non-malignant and curable and typically has a favorable prognosis. However, certain tumors may undergo malignant transformation. Longer follow-up periods are required to accurately establish the time to recurrence, to determine whether additional treatment is required and to establish the overall survival time.

Acknowledgements

Not applicable.

Funding

No funding was received.

Availability of data and materials

All data generated or analyzed during this study were included in this published article.

Authors' contributions

GQH, JSZ and YM conceived the present study. GQH performed the data analysis. QPG and SY provided and analyzed the imaging and pathological data. GQH performed software analysis of the data and figures. YM supervised the research. GQH and JSZ drafted and reviewed the initial manuscript, and performed the literature review. YM edited the manuscript. All authors read and approved the manuscript.

Ethics approval and consent to participate

Not applicable.

Patient consent for publication

Written informed consent for publication was provided by the patients' guardians as the patients were both under the age of 18.

Competing interests

The authors declare that they have no competing interests.

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