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Intracranial chordoid glioma: A clinical, radiological and pathological study of 14 cases

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ABSTRACT

Background: Chordoid gliomas (CGs) are rare neuroepithelial tumors, which commonly arise from the anterior part of the third ventricle. Most studies on CGs included only one or two cases. To better understand the disease, we report 14 patients with pathologically confirmed CGs.
Method: The clinical characteristics, including radiological and histological examination, operative records, and prognoses were analyzed and reviewed.
Result: The case series included six male and eight female patients with an average age of 44.4 years. The most common preoperative symptom was headache (64.3%) and visual deterioration (57.1%). Radiological results showed that the third ventricle (12/14) was the most common site of the brain involved, and the lesions presented with solid (n = 9, 64.3%) or cystic-solid (n = 5, 35.7%) appearance. All patients were misdiagnosed as non-CG tumors. The operation approach was mainly determined by tumor location, thus trans-callosal approach (9/14) and trans-laminar terminalis approach were commonly used. Gross total resection (GTR) was achieved in all cases and none of them received any adjuvant therapy postoperatively. The most frequent postoperative complications were diabetes insipidus, electrolyte disturbance, hypopituitarism, cognitive dysfunction, and obstructive hydrocephalus. During an average follow-up period of 40.1 months, 2 cases (14.3%) were died of refractory hypopituitarism and pulmonary embolism, respectively. The preoperative symptoms and postoperative complications were all significantly improved in other 12 patients, and MRI showed no tumor recurrence.
Conclusion: According to our experience, we recommend GTR as the primary goal, which is associated with improved rates of tumor control and without increasing rates of postoperative complications.

1. Introduction

Chordoid glioma (CG) is a rare neuroepithelial tumor of the central nervous system (CNS). It usually originates from the anterior part of the third ventricle, and a few originate from other parts of the CNS [1–4]. The name of the CG is derived from the histological manifestation, that is comprised of clusters and cords of epithelial cells in a mucinous matrix background with a strong presence of glial fibrillary acidic protein (GFAP) staining on immunohistochemistry, and classified as grade II in the 2016 World Health Organization (WHO) Classification of Tumors of the CNS [5]. Their specific location of origin leads to unique symptoms, including intracranial hypertension symptoms associated with obstructive hydrocephalus, hypothalamic dysfunction, and visual impairment [4,6–8]. Although surgery is thus far the most efficient treatment for CG, post-operative diabetes insipidus (DI) and other neuroendocrine dysfunction due to their close association and sometimes tight adherence to the hypothalamus may result in en bloc resection difficult. Worldwide, less than 100 cases have been described merely until now and lack of large retrospective studies on these tumors due to their rare nature [4]. Here, we searched the

Abbreviations: CGs, chordoid gliomas; GTR, Gross total resection; STR, subtotal resection; CNS, central nervous system; WHO, World Health Organization; DI, diabetes insipidus; TTA, trans-laminar terminalis approach; TCA, trans-callosal approach.

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cases by pathological diagnosis of CG through the chart review from the medical records database in our hospital, which included all the patients who discharged from hospital since 2010. We reported a series of 14 patients with histologically confirmed intracranial CG and their long-term outcomes in a single center. To the best of our knowledge, this study is the largest case series of intracranial CG. We believe that the clinical, radiological and pathological features of intracranial CG we showed here will provide valuable information for better diagnosis and treatment in the future.

2. Materials and methods

This retrospective study included 14 cases of intracranial CG. All of them were surgically treated and pathologically confirmed at Beijing Tiantan Hospital between January 2010 and January 2020 (Table 1). The relevant clinical data (including presentation, radiological imaging, pathology, treatment, and follow-up outcomes) were collected through a chart review and telephone interviews with an approval of the institutional review board.

Perioperative magnetic resonance imaging (MRI) was performed as standard radiological evaluation, including routine T1-weighted imaging (T1WI) and T2-weighted imaging (T2WI) with gadolinium contrast enhancement, diffusion weighted imaging (DWI), and magnetic resonance spectrum (MRS). The radiological features, including tumor location, size (the maximum diameter), cystic or solid appearance, signal intensity, and contrast enhancement, were reviewed extensively by two independent neuroradiologists. The estimation of the extent of tumor removal was acquired from the operation records or postoperative MRI. Gross total resection (GTR) and subtotal resection (STR) were defined as total and subtotal macroscopic removal, respectively.

The diagnosis of CG was pathologically confirmed by the Department of Neuropathology at Beijing Neurosurgical Institute using the 2016 WHO Classification of Tumors of the CNS [5]. Fresh paraffin-embedded tumor tissue was cut into 5-μm slices and stained with hematoxylin and eosin (H&E). Immunohistochemical staining for Ki-67 labelling index, glial fibrillary acidic protein (GFAP), epithelial membrane antigen (EMA), cytokeratin (CK), CD34 and transcription termination factor-1 (TTF-1) were used for differential diagnosis of the samples. Two independent neuropathologists reviewed the microscopic pathologies of the CG samples.

3. Results

3.1. Patient demographics

The clinic characteristics and treatment of the 14 patients of intracranial CG were described in Table 1. The tumors occurred in 6 men and 8 women, ranging in age from 27 to 67 years (mean age, 44.4 years). The duration of symptoms ranged from 1.0 to 24.0 months with a mean length of 6.0 months. The preoperative symptoms included headache (n = 9, 64.3%), visual deterioration (n = 8, 57.1%), mental status/memory changes (n = 3, 21.4%), lethargy/somnolence/fatigue (n = 6, 42.9%), ataxia (n = 1, 7.1%), diabetes insipidus (n = 1, 7.1%), sexual dysfunction/menstrual disorder (n = 2, 14.3%), epileptic seizure (n = 1, 7.1%) and dizziness (n = 4, 28.6%). None of the patients received previous radiotherapy or surgery.

3.2. Radiographic characteristics

The tumors were mainly located in the third ventricle (12/14), as shown in Fig. 1, but in a few cases, the tumors were outside the third ventricle (2/14), including temporal–parietal–occipital lobe involving the lateral ventricle and cerebellar hemisphere. The tumor maximum diameter ranged from 15 to 72 mm with a mean size of 36.7 ± 15.1 mm. The morphology of the lesions was defined as solid (n = 9, 64.3%) and cystic-solid (n = 5, 35.7%). The solid element of the tumor presented with hypointense T1 and hyperintense T2 signals in three cases (21.4%), isointense T1 and hyperintense T2 signals in five cases (35.7%), and isointense T1 and T2 signals in six cases (42.9%). After Gd-diethylenetriamine pentaacetic acid (Gd-DTPA) administration, the tumors showed homogeneous (2/14) or heterogeneous (12/14) enhancement. Additionally, hydrocephalus was present in five patients. The preoperative suspected diagnosis included craniopharyngioma (6/14), glioma (4/14), choroid plexus papilloma (2/14) and meningioma (2/14). The radiological characteristics of chordoid gliomas were summarized in Table 1.

3.3. Surgical findings and outcomes

All tumors were removed through craniotomy under operative microscope. Over half of the patients underwent frontal craniotomy with trans-callosal (TCA) approach (9/14). A translaminar terminalis (TTA) approach was documented in 3 cases of frontal craniotomies (3/14). One patient underwent temporal-parietal-occipital craniotomy, and one patient utilized suboccipital approach. Intraoperatively, the solid portion of chordoid gliomas typically appeared grayish-white or grayish-red, soft, crisp, hypervascularity, cystic portion containing xanthochromic fluid and the border ill-defined, partially adhesion to the surrounding parenchyma. Postoperative MRI indicated that GTR was achieved in all the patients (Fig. 1).

Postoperative complications occurred in 11 patients. Eight patients developed central diabetes insipidus, which resolved following desmopressin administration. Seven patients experienced transient electrolyte disturbance (hypernatremia or hyponatremia) which resolved spontaneously within 1–2 weeks. Hypopituitarism was observed in 9 patients, which was improved after hormone replacement therapy except for one patient (case 5), who was died of refractory hypopituitarism. Three patients developed obstructive hydrocephalus during the postoperative follow-up and the symptoms were relieved after ventriculoperitoneal shunting surgery. Five patients developed varying degrees of memory deterioration, which improved spontaneously in the postoperative follow-up. Bulimia, bipolar affective disorder, epileptic seizure, delirium, menstrual disorder, deep vein thrombosis and pulmonary embolism were each observed in one patient postoperatively and all of them were transient. Unfortunately, the patient (case 3) was died of pulmonary embolism. It was worth mentioning that TCA approach correlated with significantly increased numbers of memory deterioration, and TTA was associated with the significantly more postoperative hypopituitarism, diabetes insipidus and electrolyte disturbance than other surgical approaches. During an average follow-up period of 40.1 months, all the patients didn't receive any adjuvant therapy postoperatively. The preoperative symptoms and postoperative complications were all significantly improved in other 12 patients, and MRI showed no tumor recurrence.

3.4. Pathological examination

Histopathological examination revealed eosinophilic epithelioid tumor cells with medium-sized oval/elongated nuclei, arranged in cords or clusters and a mucinous matrix with infiltration of lymphocytes and plasma cells. There were slight to moderate nuclear pleomorphism, with no mitotic activity, nor necrosis. Immunohistochemical staining showed positive for GFAP, EMA, CK, CD34 and

Table 1
Clinical, radiologic, and pathological characteristics of 14 intracranial chordoid gliomas.

No.	Sex/ Age, years	Signs and symptoms	Location	Tumor diameter (mm)	Radiology	Surgical approach	Extent of resection	Postoperative complications	Adjuvant therapy	Follow-up time (months)	Outcome
1	M/ 39	Headache, fatigue, visual deterioration	Right temporal-parietal-occipital lobe involving the lateral ventricle	72	Hypointense T1WI, hyperintense T2WI and heterogeneous enhancement of cystic-solid mass without hydrocephalus	Temporal-parietal-occipital approach	GTR	Uneventful	None	119	No recurrence
2	M/ 27	Visual deterioration, somnolence and sexual dysfunction	Third ventricle	40	Isointense T1WI, hyperintense T2WI and homogeneous enhancement of cystic-solid mass with hydrocephalus	Trans-lamina terminalis approach	GTR	Hypernatremia, diabetes insipidus, hypopituitarism	None	72	No recurrence
3	F/35	Visual deterioration, menstrual disorder, fatigue, irascibility and memory deterioration	Third ventricle	41	Hypointense T1WI, hyperintense T2WI and homogeneous enhancement of solid mass without hydrocephalus	Trans-callosal approach	GTR	Diabetes insipidus, obstructive hydrocephalus, hypopituitarism, deep vein thrombosis and pulmonary embolism	None	2	Died of pulmonary embolism
4	F/51	Headache, ataxia	Left cerebellar hemisphere	47	Hypointense T1WI, hyperintense T2WI and heterogeneous enhancement of cystic-solid mass without hydrocephalus	Suboccipital approach	GTR	Uneventful	None	54	No recurrence
5	F/41	Dizziness, fatigue and diabetes insipidus	Third ventricle	31	Isointense T1WI, isointense T2WI and homogeneous enhancement of solid mass without hydrocephalus	Trans-lamina terminalis approach	GTR	Hypernatremia, hyponatremia, Diabetes insipidus, hypopituitarism	None	6	Died of hypopituitarism
6	M/ 67	Dizziness, visual deterioration	Third ventricle	28	Isointense T1WI, hyperintense T2WI and homogeneous enhancement of cystic-solid mass without hydrocephalus	Trans-callosal approach	GTR	Obstructive hydrocephalus, hypopituitarism and memory deterioration	None	44	No recurrence
7	M/ 66	Shuffle, urine incontinence, memory deterioration and visual deterioration	Third ventricle	26	Isointense T1WI, isointense T2WI and homogeneous enhancement of solid mass with hydrocephalus	Trans-callosal approach	GTR	Obstructive hydrocephalus, memory deterioration	None	39	No recurrence
8	F/32	Headache, menstrual disorder, epileptic seizure	Third ventricle	29	Isointense T1WI, isointense T2WI and homogeneous enhancement of solid mass without hydrocephalus	Trans-callosal approach	GTR	Diabetes insipidus, delirium, menstrual disorder, epileptic seizure, hyponatremia, bipolar affective disorder	None	8	No recurrence
9	F/50	Headache, visual deterioration and lethargy	Third ventricle	35	Isointense T1WI, isointense T2WI and homogeneous enhancement of solid mass with hydrocephalus	Trans-callosal approach	GTR	Hypernatremia, hyponatremia, Diabetes insipidus, hypopituitarism	None	54	No recurrence
10	F/30	Progressive headache	Third ventricle	26	Isointense T1WI, isointense T2WI and homogeneous enhancement of solid mass without hydrocephalus	Trans-callosal approach	GTR	Uneventful	None	7	No recurrence
11	F/43	Headache, dizziness, visual deterioration	Third ventricle	62	Isointense T1WI, hyperintense T2WI and homogeneous enhancement of solid mass with hydrocephalus	Trans-callosal approach	GTR	Memory deterioration, hypopituitarism, hyponatremia, Diabetes insipidus	Radiotherapy	15	No recurrence
12	M/ 43	headache, memory deterioration	Third ventricle	15	Isointense T1WI, hyperintense T2WI and homogeneous enhancement of solid mass without hydrocephalus	Trans-callosal approach	GTR	hyponatremia, Diabetes insipidus, Memory deterioration, hypopituitarism and bulimia	None	41	No recurrence

(*continued on next page*)

Table 1 (*continued*)

No.	Sex/ Age, years	Signs and symptoms	Location	Tumor diameter (mm)	Radiology	Surgical approach	Extent of resection	Postoperative complications	Adjuvant therapy	Follow-up time (months)	Outcome
13	M/ 44	Headache, dizziness and fatigue	Third ventricle	34	Isointense T1WI, isointense T2WI and homogeneous enhancement of solid mass with hydrocephalus	Trans-callosal approach	GTR	Memory deterioration, hypopituitarism	None	23	No recurrence
14	F/53	headache, visual deterioration	Third ventricle	28	Isointense T1WI, hyperintense T2WI and homogeneous enhancement of cystic-solid mass without hydrocephalus	Trans-lamina terminalis approach	GTR	Hypernatremia, hyponatremia, Diabetes insipidus, hypopituitarism	None	77	No recurrence

GTR: gross total resection, T1WI: T1 weighted image, T2WI: T2 weighted image.

TTF-1. The Ki-67 proliferative indexes were all less than 5%, and the tumor had low-grade features consistent with CG (as shown in Fig. 2). According to these findings, it was diagnosed as CG of the third ventricle (WHO grade II).

4. Discussion

Chordoid gliomas are extremely rare tumors in the CNS. The pathogenesis of this tumor is still unclear. The mainstream hypothesis believes that CG arises from embryonic tanycytes, rather than mature ependymal cells, which line the ventricles, in the vicinity of the lamina terminalis [9,10]. Tanycytes, as a specialised subgroup of ependymal cells, primarily reside in the anterior part of the third ventricle and are completely absent in the dorsal hypothalamus [11,12]. This explains that the location of CG trends to the anterior part of the third ventricle with extension to the suprasellar region and lateral ventricles. In the 2016 revision of the WHO classification of CNS tumors [5], chordoid glioma of the third ventricle was defined as a slowly-growing, non-invasive glioma located in the third ventricle. Microscopically, CG mainly consists of solid neoplasms composed of clusters and cords of epithelioid tumor cells within a variably mucinous stroma that typically contains a lymphoplasmacytic infiltrate. The nuclei are moderately sized, ovoid and relatively uniform, with few mitotic figures. Regarding immunohistochemical characteristics, these tumor cells showed diffuse reactivity for GFAP [4,10,13]. In addition, CG was found to strongly express CD34 and TTF-1 [4,7,14,15]. CD34, a transmembrane glycoprotein, expressed on hematopoietic stem cells, vascular endothelia, and mesenchymal cells [16]. In general, CD34 is consistently positive in chordoid glioma, neurofibroma, solitary fibrous tumor, and meningeal hemangiopericytoma, while negative in chordomas, meningiomas, and common forms of glioma such as diffuse astrocytomas, ependymomas, and oligodendrogliomas [17–19]. TTF-1, the nuclear transcription factor, which is strongly expressed in the neurons and ependymal and subependymal cells of the third ventricle, as well as in hypothalamic astrocytes [20]. Huo et al. reported that staining for both TTF-1 and CD34 is specifically expressed in CG of the third ventricle, which provides the most useful markers for accurate diagnosis of chordoid glioma of the third ventricle [7]. In our 14 cases, all the tumors comprised clusters and cords of epithelioid cells that typically contained lymphoplasmacytic infiltration on morphological analysis. The immunohistochemical analyses showed immune-positive for GFAP, CD34 and TTF-1. These findings were consistent with the diagnosis of chordoid glioma.

In previous literatures, CG has been reported to occur predominantly in middle-aged and elderly patients (25–75 years) with a female predominance of 2:1 [21,22]. The majority of CGs were located in the anterior part of third ventricle, and only five cases had reported that the tumor locations were outside the third ventricle, including the parieto-temporal lobe, the corona radiata, the thalamus, the temporal-parietal-occipital lobe, and the cerebellar hemisphere [1–4]. The delay from the onset of symptoms to diagnosis is highly variable and depends more on tumor location rather than size [23]. The typical symptoms were associated with raised intracranial pressure (headache, nausea, vomiting, and confusion), visual disturbances due to optic chiasm compression, memory disturbances, and endocrine dysfunction (including amenorrhea, hypothyroidism, hypocortisolism and polydipsia et al.) [4,6,22]. It may be difficult to diagnose this tumor by radiological examinations preoperatively. In our study, the preoperative suspected diagnoses included craniopharyngioma, glioma, choroid plexus papilloma and meningioma. In general, MRI depicts CG as a well-circumscribed, round or oval-

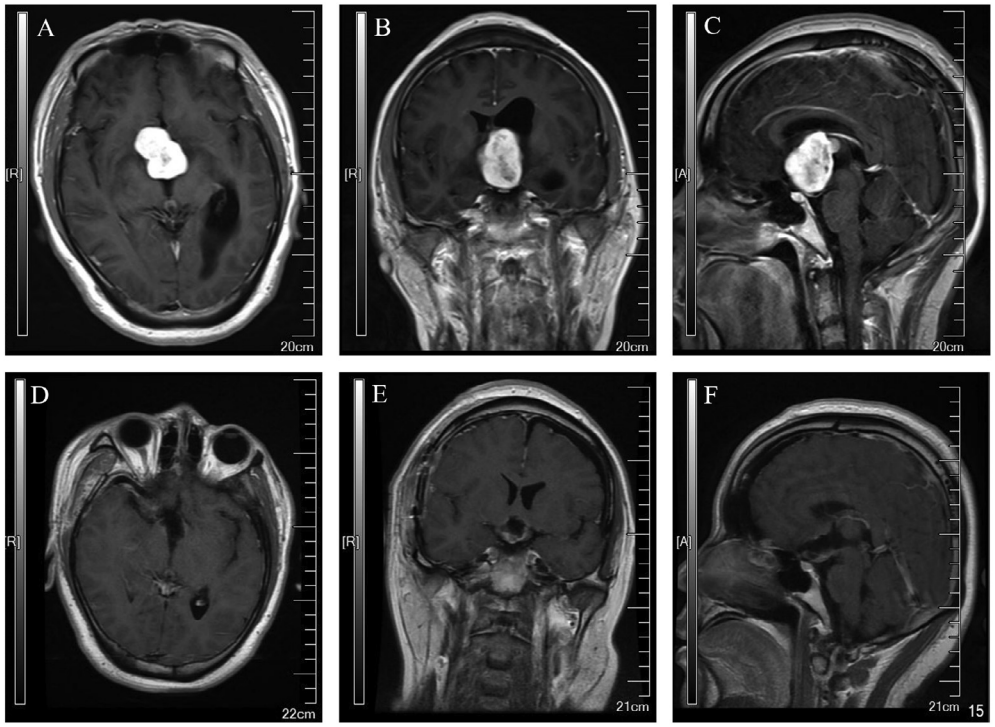

Fig. 1. Preoperative and postoperative neuroimaging findings of the chordoid glioma. (A-C) Pre-operative axial, coronal and sagittal images show an isointense, well-demarcated suprasellar lesion, which demonstrated homogenous strong enhancement after intravenous injection of gadolinium contrast on T1-weighted imaging. The tumor grew upward into the third ventricle. (D-F) Post-operative contrast T1-weighted axial, coronal and sagittal images confirm a gross total resection.

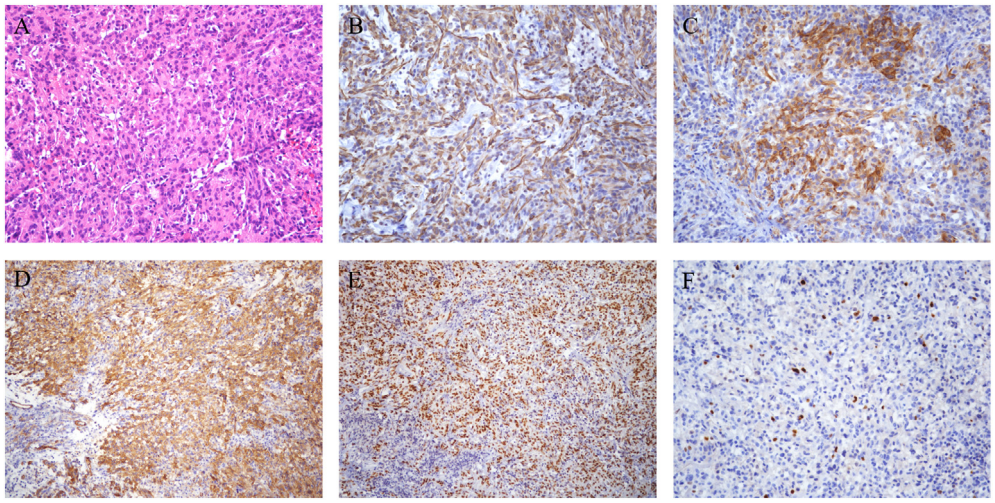

Fig. 2. Histomorphologic and immunohistochemical profile of chordoid glioma. (A) High-power photomicrograph of the chordoid glioma demonstrating eosinophilic epithelioid tumor cells with medium-sized oval/elongated nuclei, arrange in cords or clusters and a mucinous matrix with infiltration of lymphocytes and plasma cells. There are slight to moderate nuclear pleomorphism, with no mitotic activity, nor necrosis. (Hematoxylin and Eosin, original magnification ×200); (B) Tumor cells show diffuse and strong positive for GFAP (original magnification ×200); (C) Tumor cells show moderate multifocal positivity for CK (original magnification ×200); (D) Tumor cells show diffuse and strong positive for CD34 (original magnification ×100); (E) Tumor cells show diffusely positive for TTF-1 (original magnification ×100); (F) Tumor cells show a low Ki-67 proliferative indexes (<5%) (original magnification ×200).

shaped tumor. CGs have hypo- to isointensity on T1WI and iso- to hyperintensity on T2WI, with apparently homogenous enhancement in contrast imaging [7,24]. Cystic changes and necrosis may be present, but calcifications are rare. In addition, Hewer et al. described the perifocal edema in the optic tracts or basal ganglia on fluid-attenuated inversion recovery (FLAIR) MRI was extremely interesting and seemed to be the key finding for accurate preoperative diagnosis of CG [15].

Due to its rare occurrence, the best treatment strategy for CG is still controversial. Surgical resection is the preferred treatment for CG. Commonly used surgical approaches include trans-cortical, trans-callosal, and trans-lamina terminalis, which should be selected depending on the individual tumor growth pattern [7,25]. By reviewing the relevant cases in the literature, Huo et al. suggested that the trans-lamina terminalis approach may be associated with significantly reduced morbidity and mortality

rates, and incomplete resection is significantly associated with the increased recurrence rate [7]. In our cases, the trans-callosal approach was adopted by the majority patients, which may be based on anterior callosal section provides adequate surgical exposure for tumors that may be obscured by the anterior corpus callosum and other adjacent structures (including the foramen of Monro, fornix, and thalamostriate veins) [26]. Postoperatively, the patients suffered with temporary cognitive dysfunction were significantly improved during follow-up. Considering the deep location of the tumor and its proximity to vital neurovascular structures and the hypothalamus, GTR may be technically challenging. Postoperative diabetes insipidus, electrolyte disturbance, hypopituitarism, cognitive dysfunction and obstructive hydrocephalus are the most frequent complications [4,7]. In our cases, three patients developed obstructive hydrocephalus during the postoperative follow-up and the symptoms were relieved after ventriculoperitoneal shunting surgery. Other complications in our cases were reversible and improvement during the postoperative follow-up. The role of chemotherapy or radiotherapy as an adjunct therapy for CG remains unclear [8,22,27]. The use of adjuvant chemotherapy for CG has not been reported in literature. The use of adjuvant radiotherapy may minimize surgical complications and improve tumor control [28,29]. However, due to the small number of patients in literature, it is difficult to verify the role of radiotherapy in the treatment of CG. If GTR cannot be reached, tumor biopsy/partial resection with adjuvant radiotherapy are alternative treatment strategies [29–31]. The rationale was to balance the maximal reduction of tumor burden with avoidance of complications. Although CG appears to be highly recurrent in STR cases, and a number of deaths were reported, no death was attributed to disease progression. The most common causes of death are pulmonary embolism, myocardial infarction and infection [1,6,32]. In our study, there were 2 death early postoperative. The causes of death were pulmonary embolism and severe pituitary dysfunction, respectively. In other patients, no focal recurrence or metastasis was noted during the follow-up period after complete excision of the tumor. Given the high incidence of thromboembolic events, proper prevention of thromboembolic events is crucial [22]. Recently, several reports have found that CGs harbor the $PRKCA^{D463H}$ mutation, which enhances proliferation of astrocytes and tanycytes, the cells of origin of CG [33,34]. In addition, the expression of mutant PRKCA in immortalized human astrocytes leads to an increase in phosphorylated extracellular signal-regulated kinase (ERK) and anchorage-independent growth, while methylated methyl ethyl ketone (MEK) Inhibition may prevent tumor growth [33,34]. Therefore, the clarification of this genetic alteration may change the treatment paradigm for this uncommon tumor.

5. Conclusion

We described the largest single-center cases of CG. By reviewing all patients' demographics, radiological and pathological features, symptoms on presentation, surgical approaches and outcomes, we provided a meaningful insight into the management of CGs. Surgical resection remains the preferred treatment for these tumors. GTR should be the primary goal, as it is associated with improved rates of tumor control and has not been shown to increase rates of postoperative complications. Adjuvant radiotherapy has mainly been utilized in the management of residual tumor in cases where GTR may not be achievable. Molecular characterization of this lesion will also shed light on CGs oncogenesis, which may potentially lead to the development of targeted molecular therapies for CGs in the future.

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Compliance with Ethical Standards

Ethical approval: All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent: Informed consent was obtained from all individual participants included in the study.

Conflicts of interest: No potential conflicts of interest were disclosed.

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