Chordoid Glioma: Ten Years of a Low-Grade Tumor with High Morbidity

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ABSTRACT

We report two new cases of chordoid glioma of the third ventricle and review all previously published cases of this rare tumor with regard to presentation, optimum management, and outcome. Two new cases are reported with the radiological and histopathologic findings. We searched and cross-referenced PubMed and published reports of this tumor to retrieve an additional 51 cases of this tumor, which were then analyzed according to a proforma. Chordoid glioma has consistent radiological features, although some atypical elements including a cystic component are well described. The insidious presentation and morbidity of attempted surgical resection combine to give high overall morbidity. There is a high incidence of postoperative thromboembolic disturbance. Adjuvant radiosurgery has a promising role. The morbidity of this condition is likely to remain high but may be minimized by a less aggressive surgical approach together with surveillance and early use of radiosurgery.

KEYWORDS: Chordoid glioma, glioma, hypothalamus, amnesia, pituitary, thromboembolism

Chordoid glioma (CG) is an unusual, slow-growing, noninvasive primary neuroepithelial central nervous system (CNS) neoplasm of uncertain origin, classified by the World Health Organization in 2000. It was first named by Brat et al in 1998, although its features were first described by Wanschitz et al in 1995 in a 24-year-old woman.^{1,2} Clinical presentation varies. Headache, amnesia, and visual disturbance are common features. Focal cortical deficits are relatively rare.³ The delay from onset of symptoms to diagnosis is highly variable.

Differential diagnoses include tumors affecting the suprasellar region: meningioma, ependymoma, pilocytic astrocytoma, chordoma, and lymphoma.^{1,4-6} Radiological isodensity may lead to suspicion of an unruptured

aneurysm or lymphoma.^{4,5,7} Craniopharyngioma may be suspected due to a cystic component.⁸ Nonfunctioning pituitary tumors may be suspected, although on magnetic resonance imaging (MRI), CG should be distinct from pituitary gland and stalk.⁴ Inflammatory lesions such as neurosarcoid and histiocytosis may be suspected.^{4,5}

Radiological features of CG are remarkably consistent, although the exact site of origin can be difficult to identify.^{1,7} CG typically appears as a third ventricular or suprasellar mass often extending into the hypothalamus, isointense on T1-weighted MRI with strong uniform contrast enhancement.^{7,8} CG is well defined and usually has an ovoid shape, with greatest diameter in the craniocaudal direction.⁹ CG can produce obstructive hydrocephalus; development depends more on tumor

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location than size.⁷ Calcification is rare. CG may have a cystic component.⁷ Vasogenic edema may impinge on the optic apparatus.⁷ The tumor may be large at the time of diagnosis.⁸

The surgical findings vary, from tumors with a plane allowing easier tumor dissection to close adherence to the optic apparatus and hypothalamus.^{4,10,11} Few defining macroscopic features are described, but it is frequently attached to the lamina terminalis with a uniform gray or brownish color and variable consistency.¹²

Histopathologic findings are variable although glial fibrillary acidic protein (GFAP) CD34, and vimentin positivity suggest glial origin from tanyocytes, a subgroup of ependymal cells, possibly of embryonic origin. The midline location may favor an embryological origin.¹³ CG

has low mitotic activity (MIB-1 and Ki67 values below 5%). Ultrastructural features include intermediate junctions, intermediate filaments, microvilli-like projections, and focal basement membrane. CG is negative for p53 and other key tumor suppressor and proto-oncogenes, as well as genetic markers of other CNS tumors, such as *TP53*, *EGFR*, *CDK4*, and *MDM2* genes.¹⁴ It is also negative for estrogen and progesterone receptors, unlike intermediate- and high-grade gliomas.^{6,15}

We present two new cases of CG, one with unique histopathologic features. We review all previously reported cases of CG focusing on clinical and surgical aspects. We also review radiological and histopathologic findings and discuss the evidence for optimum management of this rare tumor.

Figure 1 (A) Axial T2-weighted, (B) sagittal, and (C) coronal T1-weighted gadolinium-enhanced magnetic resonance imaging scan showing round, well-defined tumor with avid uniform contrast enhancement in the floor of the third ventricle.

Figure 2 Coronal T1-weighted gadolinium-enhanced magnetic resonance imaging scan (A) 6 months and (B) 1 year postoperatively showing stable residuum of disease.

CASE REPORTS

Case 1

A 48-year-old woman presented with 5 months of fluctuating visual failure and progressive intermittent headaches. Examination found left optic pallor but normal visual fields. There was no history of endocrine or memory disturbance. MRI showed a 2-cm round, homogeneously enhancing, well-defined lesion adjacent to the floor of the third ventricle, with compression of the optic chiasm from above with little surrounding edema (Fig. 1). The presumed diagnosis was of an intraventricular meningioma.

A bifrontal craniotomy and interhemispheric approach to the tumor was performed, and the tumor was debulked extensively. A poor plane was established superiorly and posteriorly between the tumor and brain, and resection was incomplete.

Histopathology showed a tumor with low mitotic proliferation and clusters of ovoid epithelial cells with eosinophilic cytoplasm and mucinous stroma. The tumor was positive for GFAP, CD-34, S-100, and vimentin. The diagnosis was of CG.

Postoperatively, the patient developed severe electrolyte disturbance with alternating diabetes insipidus and syndrome of inappropriate antidiuretic hormone secretion. She also suffered prolonged confabulatory amnesia. After 3 months, her electrolyte imbalances settled. She underwent follow-up MRI at 6 months and 1 year, showing stable residual disease (Fig. 2).

Case 2

A 36-year-old man presented with a 1-week history of worsening headache and a 1-year history of polyuria and polydipsia. Neurological and ophthalmologic

examinations were unremarkable. Endocrine studies showed hypothalamic dysfunction. MRI showed a well-defined, solid suprasellar mass, suspicious of craniopharyngioma. Total resection was achieved via bifrontal craniotomy and translamina terminalis approach. No adjuvant therapy was given. The early postoperative course was eventful with diabetes insipidus, metabolic myopathy, diencephalic amnesia, partial hypopituitarism, and hypothalamic syndrome with weight gain. At 40 months of follow-up, the patient continues to suffer intrusive amnesia and hypopituitarism. Further MRI scans have shown no evidence of recurrence.

Case 2—Pathological Findings

The tumor was built up by cords, files, and clusters of epithelioid tumor cells with abundant eosinophilic cytoplasm and separated by dense, collagen-rich fibrous stroma and infiltrated by large numbers of lymphoplasmacytic cells (Fig. 3). Numerous Russell bodies were noted in plasma cells. There was only little mucin deposition in the stroma. Focal microcalcification was also noted. Mitotic figures, vascular proliferation, and necrosis were absent. In the adjacent atrophic, non-neoplastic brain tissue, reactive astrocytes, and Rosenthal fibers were seen. GFAP immunohistochemistry showed strong cytoplasm staining and highlighted the multipolar fibrillary processes of the tumor cells, although some tumor cells remained negative. CD34 was strongly positive in all tumor cells. EMA, MNF116, chromogranin, and synaptophysin were negative. The inflammatory cells revealed a mixture of CD20-positive B cells, CD68 (PGM1)-positive macrophages, and some CD3-positive T cells. Ki67 was expressed in less than 1% of the tumor cell population.

Figure 3 Histopathologic findings, case 2. (a, b) Chordoid glioma showing clusters of epithelioid tumor cells with abundant eosinophilic cytoplasm separated by dense, collagen-rich fibrous stroma (hematoxylin and eosin; a $\times 10$; b $\times 20$). Large numbers of lymphoplasmacytic inflammatory cells present. The tumor cells are immunoreactive with CD34 (c) and GFAP (d) (c $\times 20$, d $\times 40$).

The features were consistent with a CG of the third ventricle, although the prominent desmoplasia and the lack of mucinous stroma were highly unusual.

Published Cases

Fifty-three cases of CG have been reported to date, including the presented cases. Their details according to our analysis are presented in Tables 1 and 2. Nineteen cases (36%) occurred in male patients and 34 (64%) in female patients. Only three pediatric cases (5.7% of the total) are reported, in two boys and one girl aged 5, 12, and 7, respectively. Ages range from 5 to 71 years.

Presenting Symptoms

Clinical presentation was described in 50 cases, most commonly headache (20 patients, 40%) and visual disturbance (15 cases, 30%). Twelve patients (24%) reported memory deficits (summarized in Table 3).

Six patients (12%) presented with endocrine symptoms. Other symptoms reported include sweating,

gastrointestinal disturbance, speech dysfunction, and sleep apnea.

Thirty-nine patients (78%) were polysymptomatic. Eleven cases (22%) were clearly monosymptomatic, with visual disturbance being the most common sole symptom in these cases.

The timescale of clinical presentations was described in 39 cases. Twenty-nine cases (74%) presented chronically, 5 (13%) presented acutely, and 5 (13%) manifested in an acute-on-chronic manner. The duration of chronic symptoms was a few weeks to 20 years.

Examination Findings and Investigations

Twelve of 32 patients (37.5%) had a visual field defect. Visual acuity was reported as abnormal in six cases (19%). Two patients (6%) had optic neuropathy, two patients (6%) had other visual signs, and one patient (3%) had papilledema. In total, 23 out of 32 patients (72%) displayed visual signs. Seven cases (22%) showed endocrine dysfunction, mainly affecting the sex hormones. Seven patients (22%) had a focal neurological deficit.

Table 1 Clinical and Radiological Features (All Published Cases of Chordoid Glioma)

Case No.	Author	Age and Sex	Presenting Features	Acute or Chronic	Signs and Investigations	Imaging
1	Brat et al, 1998 ¹	50 F	Lethargy, urinary incontinence, ataxia			3v mass and hydrocephalus
2	Brat et al, 1998 ¹	70 M	Ataxia	Chronic, 2 y		Suprasellar and 3v mass with solid homogenously enhancing and multicystic components extending into the right temporal lobe
3	Brat et al, 1998 ¹	59 F	Obstructive hydrocephalus			Solid and cystic 3v mass
4	Brat et al, 1998 ¹	47 F	Obstructive hydrocephalus			3v mass
5	Brat et al, 1998 ¹	31 F	Headaches, nausea, vomiting, LOC	Chronic, 6 mo		Homogenously enhancing 3v mass
6	Brat et al, 1998 ¹	56 F	Endocrine disturbance		Hypothyroidism; diabetes insipidus	Solid 3v mass
7	Brat et al, 1998 ¹	31 F	Gastrointestinal disturbance, weight loss		Hypothyroidism	Solid, enhancing 3v 2.5-cm mass
8	Brat et al, 1998 ¹	35 F	Amenorrhea, psychosis	Chronic, 1 y		Solid, enhancing 3v mass
9	Reifenberger et al, 1999 ¹⁴	56 F	Headache, fatigue	Chronic, 6 mo	Scotomata	Well-demarcated, uniformly enhancing, 23 × 19 × 20-mm 3v mass
10	Reifenberger et al, 1999 ¹⁴	53 F	Visual disturbance, weight gain	Chronic, 20 y	VFD	2.5 × 2.5 × 2-cm suprasellar mass
11	Reifenberger et al, 1999 ¹⁴	65 M	Speech disturbance, right-sided facial weakness	Acute	Hypertension, resolving right-sided facial nerve palsy	Well-demarcated, homogenously enhancing 3v mass
12	Reifenberger et al, 1999 ¹⁴	35 M	Psychosyndrome			Well-demarcated, enhancing 3v mass
13	Ricoy et al, 2000 ²³	41 F	Visual disturbance	Chronic, 18 mo	Bilateral ↓ VA, VFD; endocrine profile normal	3 × 2.4 × 2.5-cm homogenously enhancing suprasellar mass
14	Vajtai et al, 1999 ²⁴	60 F	Headache, amnesia, somnolence		Generalized hypotonia; Romberg's positive test	Irregularly round, well-defined, 3v 3.5-cm enhancing mass with hydrocephalus
15	Castellano-Sanchez et al, 2000 ⁸	36 M	Progressive myopia	Chronic, 12 y	VFD, ↓ VA	Homogenously enhancing 3-cm suprasellar mass protruding into 3v
16	Tonami et al, 2000 ²⁵	42 F	Amenorrhea, amnesia, LOC	Chronic, 1 y		Well-defined, hyperdense 3v mass with homogenous enhancement and a cystic component
17	Castellano-Sanchez et al, 2001 ¹⁷	12 M	Visual disturbance		Left optic neuropathy	Solid, suprasellar 3.6 × 3 × 3-cm heterogeneously enhancing mass
18	Cenacchi et al, 2001 ²⁶	34 M				Well-demarcated, solid, homogenously enhancing 3v mass
19	Cenacchi et al, 2001 ²⁶	40 M				Well-demarcated, homogenously enhancing, solid 3v mass
20	Cenacchi et al, 2001 ²⁶	43 F				Well-demarcated, homogenously enhancing, solid 3v mass
21	Galloway et al, 2001 ²⁷	54 M	Left-sided weakness, ataxia	Acute	Bilateral nystagmus, VFD; left-sided weakness.	Well demarcated, 2-cm, contrast-enhancing suprasellar mass; also low-density area in left internal capsule, probably ischemic
22	Hanbali et al, 2001 ²²	57 M	First presentation: headache, ataxia, anosmia, malaise, anorexia; Second presentation: headache, amnesia, confusion	Chronic	Hyponatremia and SIADH	Midline, 1.5-cm well-defined, solid, uniformly enhancing cystic 3v mass
23	Grand et al, 2002 ⁴	41 F	Visual disturbance, headache			Well-defined, solid, homogenously enhancing, 3v 2.8 × 3 × 3.5-cm mass with cystic component.
24	Oda et al, 2002 ²⁸	25 M	Hyperphagia, amnesia	Chronic, 2 y		Well-defined, oval, 4.5-cm, homogenously enhancing 3v mass with cystic component
25	Pasquier et al, 2002 ¹²	35 M	Headache, nausea, insomnia, visual disturbance, weakness	Chronic, 18 mo		Midline, solid, 1.5-cm homogenously enhancing 3v mass

Table 1 (*Continued*)

Case No.	Author	Age and Sex	Presenting Features	Acute or Chronic	Signs and Investigations	Imaging
26	Pasquier et al, 2002 ¹²	39 M	Headache, diplopia	Chronic, 18 mo	VFD	Well-defined, partially cystic, midline 2.8 × 2.5 × 2-cm uniformly enhancing 3v mass
27	Nakajima et al, 2003 ²¹	49 F	Headache, amnesia, urinary incontinence	Chronic, 1 y	Papilledema; normal pituitary function	Well-defined, homogenously enhancing, hyperdense 4 × 4 × 3-cm suprasellar mass with hydrocephalus
28	Raizer et al, 2003 ⁵	57 F	Incoordination, drop attacks, amnesia	Chronic	Right-sided hearing loss	Well-defined, homogenously enhancing, 3v 2.3 × 2.1 × 1.9-cm mass
29	Sato et al, 2003 ¹³	65 F	Headache	Chronic, 2 mo	None	Well-defined, solid 2.5-cm homogenously enhancing 3v mass
30	Suh et al, 2003 ¹⁵	48 F	Headache, dizziness	Chronic, 1 y	VFD, ↑ PRL	Well-defined, enhancing, solid cystic 1.7-cm suprasellar mass
31	Taraszkewska et al, 2003 ²⁹	62 M	Headache, polyuria, polydipsia	Chronic, 6 mo	None	Solid, well-defined, hyperdense, 1.5-cm homogenously enhancing suprasellar mass
32	Taraszkewska et al, 2003 ²⁹	51 F	Somnolence, visual disturbance	Chronic, 1 y		Well-defined, isointense, 2.5-cm, homogenously enhancing suprasellar mass
33	Buccoliero et al, 2004 ³⁰	56 F	Incidental finding			Well-defined, solid, cystic, 2-cm, enhancing, 3v mass
34	Bensalah et al, 2005 ³¹	47 F	Sleep apnea, somnolence, headache	Chronic	VFD	Midline 3v mass of heterogenous density, strongly enhancing
35	Kurian et al, 2005 ⁶	32 F	Visual disturbance, headache, amenorrhea, weight gain	Chronic, 5 y	VFD, ↓VA, ↓E3, ↓progesterone	Lobulated, enhancing suprasellar 2.1 × 2.2 × 2.4-cm 3v mass
36	Kurian et al, 2005 ⁶	37 F	Amnesia, confusion	Acute	Acute hydrocephalus, VFD, ↓VA	Well-defined, ovoid, enhancing mass in suprasellar cistern
37	Baehring and Bannykh, 2006 ¹⁸	71 F	Visual disturbance	Chronic	VFD, normal pituitary function	Solid, suprasellar, enhancing, semicystic mass
38	Jung and Jung, 2006 ¹¹	50 F	Cognitive dysfunction, hyperphagia	Chronic, 4 y	↓VA	Calcified, multilobulated, enhancing 4.5 × 3.5-cm 3v mass with hydrocephalus
39	Leeds et al, 2006 ⁹	57 M	Fatigue, anosmia, sweating, headache, anorexia	Chronic, several months		Well-defined, uniformly enhancing, 1.8-cm suprasellar mass
40	Nga et al, 2006 ³²	49 F	Drowsiness, visual disturbance, weakness	Acute on chronic	GCS 11; dilated and sluggish right pupil; left hemiparesis and hypertonia	Solid, well-defined, heterogeneously enhancing, 3 × 2 × 0.5-cm 3v mass with hydrocephalus
41	Takei et al, 2006 ³³	42 F	Amnesia, confusion	Acute		5-cm, 3v mass lesion
42	Goyal et al, 2007 ¹⁶	5 M	Headache, vomiting, sensorimotor disturbance	Acute	Raised ICP, right-sided hypertonia,	Left temporoparietal 7 × 5.5 × 4-cm hemorrhagic mass; midline shift and compression of lateral ventricle
43	Jain et al, 2008 ¹⁰	7 F	Fever, headache, vomiting	Chronic, 8 mo	Right facial nerve palsy, bilateral lower-limb weakness	Well-demarcated, mixed-density, heterogenous, contrast-enhancing, juxtaventricular mass in right corona radiata extending to the thalamus
44	Jain et al, 2008 ¹⁰	55 M	Amnesia, urinary incontinence, seizures	Chronic	↓VA on left	Homogenously enhancing 3v mass with a cystic component
45	Ortega-Martínez et al, 2007 ³⁴	M 51	Amnesia, ataxia, urinary incontinence	Acute on chronic	No visual or endocrine abnormalities	Well-defined, 3 × 4 × 2.5-cm, uniformly contrast enhancing 3v mass with hydrocephalus
46	Gallina et al, 2007 ³⁵	56 F	Incidental finding		None	Solid, well-defined, homogenously enhancing suprasellar mass
47	Vanhauwaert et al, 2008 ³⁶	58 F	Amnesia	Chronic		Solid 3v mass
48	Carrasco et al, 2008 ³⁷	53 F	Visual disturbance, confusion, dizziness	Acute on chronic	VFD, ↓E3, ↑PRL; delayed VEPs	Well-defined, 3 × 2.5-cm, semicystic uniformly enhancing 3v mass

Table 1 (Continued)

Case No.	Author	Age and Sex	Presenting Features	Acute or Chronic	Signs and Investigations	Imaging
49	Iwami et al, 2009 ²⁰	61 F	Syncope	Chronic	None	Well-demarcated, uniformly contrast-enhancing, partially cystic 3.5-cm 3v mass with calcification
50	Kawasaki et al, 2009 ¹⁹	42 F	Headache, LOC, visual disturbance	Acute on chronic		Well-demarcated, homogenously enhancing hemorrhagic 3v mass with hydrocephalus
51	Kawasaki et al, 2009 ¹⁹	51 M	Left-sided visual disturbance	Chronic, 6 mo		Well-demarcated homogenously enhancing 3v partially cystic mass
52	Present case 1	36 M	Polyuria, polydipsia, headache	Acute on chronic	Endocrine tests abnormal	Well-defined, solid suprasellar mass
53	Present case 2	48 F	Visual disturbance, headache	Chronic, 5 mo	Left optic pallor	Well-defined 2-cm homogenously enhancing 3v mass

E3, estrogen; GCS, Glasgow Coma Scale; ICP, intracranial pressure; LOC, loss of consciousness; PRL, prolactin; SIADH, syndrome of inappropriate antidiuretic hormone secretion; VFD, visual field defect; VA, visual acuity; VEP, visual evoked potential; 3v, third ventricle.

Radiological Investigations

Thirty of 53 cases conformed to the typical radiological description of CG: a solid, well-defined, ovoid, isointense suprasellar/third ventricular mass with strong uniform contrast enhancement. Unusual radiological features are summarized in Table 4. One tumor was sited in the temporoparietal region, another in the corona radiata and thalamus. Both were pediatric cases. One of the pediatric cases also exhibited hemorrhage within the tumor. Seven cases (13%) had radiological evidence of hydrocephalus. The largest tumor, a pediatric case, measured $7 \times 5.5 \times 4$ cm. The smallest was 1.5 cm in diameter.

Management

Forty-nine of 52 cases (94%) were debulked or removed completely. Eleven cases (21%) were biopsied. In 3 (6%) of these 11 cases, biopsy was the only management used without resection. Nine (17%) patients received adjuvant radiotherapy: external beam in three, radiosurgery in four, and radioactive implants in two. Shunting was required in 5 (9.6%) patients.

Details of the surgical approach used are available for 26 patients. Four cases (15%) were managed by the transcallosal approach and 5 (19%) by the translamina terminalis approach. Three tumors (11.5%) were approached interhemispherically (no further details) and 3 (11.5%) by a transventricular route. A subfrontal approach was taken in two cases (8%). Unspecified bifrontal craniotomy, anterior approach, and transsphenoidal routes, respectively, were taken in one patient each.

The extent of resection was reported in 44 patients. Two patients required more than one resection, making a total of 46 resections. Twenty-four out of the 46 resections (52%) were reported total and the remaining 22 (48%), subtotal. Of the subtotal and partial resections (22 operations), eight received adjuvant radio-

therapy and 14 did not. Twenty-one patients of the 44 reported were left with residual tumor following surgery.

Postoperative Complications

Postoperative complications were discussed in 39 cases. Table 5 shows the frequency of the most common complications. As expected, these relate to the presenting symptoms and location, with hypothalamic disturbance and amnesia being most common.

Outcome

Data on outcome are variably reported. The range of follow-up is 0 to 68 months in living patients and one postmortem report in a patient who died of unrelated causes 6 years later. Visual improvement was noted in four patients. No new neurological deficit was reported in nine cases. Formal functional assessments are seldom available.

There were 15 deaths. One-third of these were due to pulmonary embolism. Four deaths (27%) were caused by infection. Two patients (13%) had a cardiac arrest. Two deaths (13%) were considered directly attributable to tumor. Two-thirds of the 15 deaths occurred in the first 4 weeks postoperatively.

Recurrence

Little follow-up data are reported. Recurrence was noted in one patient following an apparently total resection. Progression was noted in 5 of the 22 cases where subtotal resection was performed. Following total resection, no recurrence has been found in 15 patients. Twelve patients remain progression free after a subtotal or partial resection.

Of the nine patients receiving adjuvant radiotherapy, progression was observed in three. All these

Table 2 Management, Complications, and Outcome (All Published Cases of Chordoid Glioma)

Case No.	Author	Age and Sex	Treatment Summary	Operative Approach	Operative Procedure	Complications	Outcome and recurrence (where stated)
1	Brat et al, 1998 ¹	50 F	Resection	Craniotomy	Total resection	Multiple medical complications	No growth of residual tumor 1 y postop
2	Brat et al, 1998 ¹	70 M	Resection		Subtotal resection		
3	Brat et al, 1998 ¹	59 F	Resection, postop radiation		Subtotal resection		
4	Brat et al, 1998 ¹	47 F	Resection		Subtotal resection		
5	Brat et al, 1998 ¹	31 F	Resection		Total resection		
6	Brat et al, 1998 ¹	56 F	Resection		Subtotal resection		
7	Brat et al, 1998 ¹	31 F	Resection, postop radiation		Subtotal resection		
8	Brat et al, 1998 ¹	35 F	Resection	Transventricular approach	Subtotal resection	PE	Death from PE
9	Reifenberger et al, 1999 ¹⁴	56 F	Stereotactic biopsy followed by resection; gamma knife radiosurgery; bilateral ventricular shunts		Partial resection	Amnesia, somnolence, hydrocephalus	MRI at 3.5 y shows no recurrence
10	Reifenberger et al, 1999 ¹⁴	53 F	Microsurgical resection	Bifrontal craniotomy	Extent of resection not stated	Uneventful	Death a few days postop from PE
11	Reifenberger et al, 1999 ¹⁴	65 M	Stereotactic biopsy followed by resection	Craniotomy	Total resection	PE	
12	Reifenberger et al, 1999 ¹⁴	35 M	Resection		Total resection	PE	
13	Ricoy et al, 2000 ²³	41 F	Resection	Unilateral subfrontal approach	Complete resection	Uneventful	Improved VA; no evidence of recurrence at 13 mo
14	Vajtai et al, 1999 ²⁴	60 F	Resection	Right frontal transventricular	Subtotal resection	Hyponatremia; bronchopneumonia	Death 10 d postop
15	Castellano-Sanchez et al, 2000 ⁸	36 M	2 resections	Right frontotemporal craniotomy	Subtotal resection	Seizures	No neurological deficits
16	Tonami et al, 2000 ²⁵	42 F	Resection; postop radiotherapy and stereotactic radiosurgery	Translaminar terminalis approach	Partial resection		No enlargement of residual at 9 mo
17	Castellano-Sanchez et al, 2001 ¹⁷	12 M	Debulking and biopsy	Right pterional craniotomy	Total resection	PE	No recurrence at 2 y, patient well
18	Cenacchi et al, 2001 ²⁶	34 M	Resection				
19	Cenacchi et al, 2001 ²⁶	40 M	Resection				
20	Cenacchi et al, 2001 ²⁶	43 F	Resection				
21	Galloway et al, 2001 ²⁷	54 M	Decompression of optic chiasm and resection of tumor	Right pterional craniotomy	Total resection	Uneventful	Recovery of VF and VA; independent with all ADLs; no recurrence at 9 mo
22	Hanbali et al, 2001 ²²	57 M	Two operations—May 1999 and April 2000; radiotherapy after first operation	First—craniotomy and subfrontal approach; Second—right frontal craniotomy and transcallosal approach	First—biopsy and subtotal resection; Second—gross total resection	First—left parietal stroke, MI; Second—central DI, confusion	First—progression, prompting 2nd presentation; Second—no residual at 3 mo; death from 2nd MI months later
23	Grand et al, 2002 ⁴	41 F	Resection	Anterior approach	Near complete resection; tumor encapsulated and nonadherent	Central DI, weight gain, amnesia	No residual mass on postop MRI

Table 2 (Continued)

Case No.	Author	Age and Sex	Treatment Summary	Operative Approach	Operative Procedure	Complications	Outcome and recurrence (where stated)
24	Oda et al, 2002 ²⁸	25 M	Stereotactic biopsy; resection 1 mo later	Anterior transcallosal interforniceal	Complete resection	Central DI, hyperthermia, amnesia, hyponatremia	No recurrence at 17 mo; symptoms gradually improved
25	Pasquier et al, 2002 ¹²	35 M	Resection		Near total resection		No growth of residual at 68 months
26	Pasquier et al, 2002 ¹²	39 F	Resection		Subtotal resection	Amnesia; weight gain of 20 Kg	No growth of residual at 16 mo
27	Nakajima et al, 2003 ²¹	49 F	Resection; gamma knife	Bifrontal craniotomy with translamina terminalis approach	Estimated 70% resection	Hypothalamic dysfunction, PE	No recurrence of residual at 2-y follow-up
28	Raizer et al, 2003 ⁵	57 F	Endoscopic biopsy; open resection	Subfrontal/ interhemispheric	Gross total resection	Partial DI, AF, meningitis	No recurrence at 2-y follow-up; no new neurological deficits
29	Sato et al, 2003 ¹³	65 F	Resection		Subtotal resection	Uneventful recovery	No recurrence at 2 y; death from unrelated causes
30	Suh et al, 2003 ¹⁵	48 F	Resection for suspected macroadenoma	Sublabial incision for transsphenoidal approach	Gross total resection; cystic portion found to be coexistent Rathke's cyst	Central DI	VFD resolved; no recurrence at 17 mo
31	Taraszewska et al, 2003 ²⁹	62 M	Resection	Pterional approach	Total resection	Respiratory failure, coma, central DI, visual disturbance, MI	Death from cardiac arrest at 6 wk
32	Taraszewska et al, 2003 ²⁹	51 F	Resection	Subfrontal translamina terminalis	Total resection	Uneventful	Residual tumor seen at 4 mo
33	Buccoliero et al, 2004 ³⁰	56 F	Resection		Gross total resection	Uneventful	No recurrence 8 mo postop; patient well
34	Bensalah et al, 2005 ³¹	47 F	Biopsy				
35	Kurian et al, 2005 ⁶	32 F	Surgery with radioactive implants		Subtotal resection	Hypothalamic dysfunction	Poor condition at 15 mo
36	Kurian et al, 2005 ⁶	37 F	VP shunt; stereotactic biopsy and radioactive seed implants; resection at 9 mo			Central DI, sepsis, hypothermia, hypotension	Death 2 wk postop from sepsis
37	Baehring and Bannykh, 2006 ¹⁸	71 F	Biopsy, VF monitoring				No deterioration
38	Jung and Jung, 2006 ¹¹	50 F	Resection; VP shunt 2 mo later	Anterior transcallosal approach	Subtotal resection	Progressive drowsiness; hydrocephalus; pneumonia	Recurrence at 3 mo and death at 4 mo postop
39	Leeds et al, 2006 ⁹	57 M	Biopsy				
40	Nga et al, 2006 ³²	46 F	Resection	Craniotomy and transcortical and transventricular resection		Hypotension; hypothermia	Death 18 d postop from pneumonia
41	Takei et al, 2006 ³³	42 F					
42	Goyal et al, 2007 ¹⁶	5 M	Resection				Death 2 d postop from cardiac arrest
43	Jain et al, 2008 ¹⁰	7 F	Biopsy followed by resection		Gross total resection		
44	Jain et al, 2008 ¹⁰	55 M	Resection		Gross total resection	Hematoma; bacterial meningitis	Death 1 wk postop from meningitis
45	Ortega-Martínez et al, 2007 ³⁴	51 M	VP shunt and resection	Transcallosal approach	Total resection	PE	Death 3 d postop from PE
46	Gallina et al, 2007 ³⁵	56 F	Resection	Right pterional craniotomy	Total resection	None	No recurrence at 48 mo follow-up; patient remained asymptomatic
47	Vanhouwaert et al, 2008 ³⁶	58 F	Resection		Gross total resection	Central DI, hypothermia	No recurrence at 18 mo follow-up
48	Carrasco et al, 2008 ³⁷	53 F	VP shunt followed by resection	Translamina terminalis approach via left pterional craniotomy	Complete resection	Amnesia; disorientation	Endocrine improvement, no improvement in VF, no recurrence at 2 mo

Table 2 (Continued)

Case No.	Author	Age and Sex	Treatment Summary	Operative Approach	Operative Procedure	Complications	Outcome and recurrence (where stated)
49	Iwami et al, 2009 ²⁰	61 F	Resection; gamma knife radiosurgery	Subfrontal interhemispheric	Partial resection	Amnesia	No regrowth at 12 mo follow-up
50	Kawasaki et al, 2009 ¹⁹	42 F	Resection		Total resection	Organic psychosyndrome; amnesia, central DI	No recurrence at postmortem 6 y later—cause of death unrelated
51	Kawasaki et al, 2009 ¹⁹	51 M	Resection		Partial resection		Improved VA. No recurrence at 40 mo, patient well
52	Present case 1	36 M	Resection	Interhemispheric translamina terminalis	Total resection	Central DI, amnesia, weight gain, partial hypopituitarism, myopathy	No recurrence on MRI at 40 mo
53	Present case 2	48 F	Resection	Interhemispheric approach via bifrontal craniotomy	Partial resection	Central DI, SIADH, confabulatory amnesia	No recurrence of residual at 24 mo

ADL, activities of daily living; AF, atrial fibrillation; DI, diabetes insipidus; MI, myocardial infarction; MRI, magnetic resonance imaging; PE, pulmonary embolism; postop, postoperatively; SIADH, syndrome of inappropriate antidiuretic hormone secretion; VFD, visual field defect; VA, visual acuity; VF, visual field; VP, ventricoperitoneal.

patients had received conventional radiotherapy following an incomplete resection. One patient received both conventional radiotherapy and radiosurgery after an incomplete resection with no subsequent recurrence. Three patients received gamma knife treatment follow-

ing an incomplete resection and none showed progression. Two patients had radioactive implants. One of these patients remains recurrence free and the other died of unspecified causes in the early postoperative period.

Table 3 Presenting Features

Clinical Feature	Number of Cases (total = 50)
Headache	20 (40%)
Visual disturbance	15 (30%)
Memory impairment	12 (24%)
Ataxia/incoordination	6 (12%)
Other	6 (12%)
Limb weakness	5 (10%)
Confusion	5 (10%)
Urinary incontinence	4 (8%)
Lethargy/malaise	4 (8%)
Nausea and vomiting	4 (8%)
Changes in consciousness	4 (8%)
Increased weight/hyperphagia	4 (8%)
Somnolence	4 (8%)
Reduced weight/anorexia	3 (6%)
Amenorrhea	3 (6%)
Polyuria/polydipsia	2 (4%)
Incidental	2 (4%)
Obstructive hydrocephalus	2 (4%)
Anosmia	2 (4%)
Dizziness	2 (4%)
Syncope/drop attacks	2 (4%)
Psychiatric disturbance	2 (4%)
Cranial nerve palsy	1 (2%)
Seizures	1 (2%)
Insomnia	1 (2%)
Other endocrine	1 (2%)

Immunocytochemistry

Table 6 shows the immunochemical patterns observed.

Histological Findings

All 50 cases that were histologically analyzed exhibited the typical cord and clusters of polygonal epithelioid cells. Twenty-nine cases were observed for Russell bodies and were present in 28 (97%). Twenty-seven cases showed round-oval nuclei. Thirty-six out of 36 cases had eosinophilic granules or cytoplasm. The stroma of 49 cases was described as mucinous or myxoid and in all, although sparse in a few cases and very sparse in the second present case. Reactive gliosis or Rosenthal fibers were found in 14 of 16 cases (88%). Lymphocytic and plasma cell infiltrates were present in 48 of 50 cases (96%), albeit sparsely in 5 (10%). Significant mitoses were only found in 1 of the 45 cases. Necrosis was present in the same case and was looked for in 28 cases.

Table 4 Atypical Radiological Features in Chordoid Glioma

Feature	Number of Cases (total = 53)
Cystic component	14 (26%)
Heterogenous enhancement	3 (6%)
Unusual site	3 (6%)
Hemorrhage	2 (4%)
Calcification	2 (4%)

Table 5 Postoperative Complications following Resection of Chordoid Glioma

Complication	Number of Cases (total = 39)
Diabetes insipidus	12 (31%)
Amnesia	10 (26%)
No complications	7 (18%)
Pulmonary embolism	6 (15%)
Other	6 (15%)
Hypothermia/hyperthermia	4 (10%)
Hyponatremia	3 (8%)
Weight gain	3 (8%)
Hypothalamic syndromes	3 (8%)
Hypotension	2 (5%)
Deepened consciousness	2 (5%)
Confusion	2 (5%)
Myocardial infarction	2 (5%)
Meningitis	2 (5%)
Pneumonia	2 (5%)
Hydrocephalus	2 (5%)
Visual deficit	2 (5%)

The MIB-1 was recorded in 17 cases with a mean value less than 5%. Ki67 was recorded in 22 cases and ranged from 0.3 to 5%.

Ultrastructure

Ultrastructural studies were performed in 22 cases. The results are shown in Table 7.

DISCUSSION

Demographics

CG occurs mainly in adults, with only 3 of 53 cases in pediatric age group.^{10,16,17} A female-to-male ratio of 2 to 3:1 has been described, similar to our findings of ~3:2.^{4,6} CG has no known risk factors or syndromic

Table 6 Immunochemical Staining in Chordoid Glioma

Marker	Number of Cases Stained for Marker	Number of Cases Positive for Marker
CD34	31	30 (97%)
S100	36	25 (69%)
EMA	44	30 (68%)
Vimentin	45	45 (100%)
Cytokeratin	34	24 (70.5%)
GFAP	43	43 (100%)
Synaptophysin	24	0 (0%)
Neurofilaments	20	2 (10%)
Desmin	10	0 (0%)

Table 7 Ultrastructural Features of Chordoid Glioma

Feature	Number of Cases Observed for Feature	Number of Cases Demonstrating Feature
Intermediate junctions	18	17 (94%)
Intermediate filaments	20	20 (100%)
Microvilli	22	19 (86%)
Focal basement membrane	21	21 (100%)
cilia	21	6 (29%)
Well-formed desmosomes	15	1 (7%)

associations and no etiologic factor could be identified in this review. One case had coexisting Rathke's cyst, probably unrelated.¹⁵

Location

CG typically arises in the suprasellar region and the third ventricle, with all but two of the cases reported arising in these structures, supporting embryological origin. CG can extend to occupy the entire ventricle, suprasellar, and hypothalamic regions.

CG rarely arises elsewhere, with no reports of this in adults and only two cases overall. In one, CG arose from the left temporoparietal cortex in a 5-year-old boy and in another from the corona radiata and thalamus in a 7-year-old girl.^{10,16} The cystic component of one tumor invaded the right temporal lobe of a 70-year-old man, but did not originate here.¹ The unusual site of CG at the extremes of age may be relevant. The pediatric cases were atypical in other aspects and are considered later. Metastasis is not described.

Presentation

Overall, chronic presentations predominate (months to 20 years, with 6 months to 2 years being the most common), occurring in almost 75%. Although CG is described as slow growing, rapid periods of growth are reported in one case where tumor remained stable for 3 years and then underwent a sixfold expansion over 6 months.⁵ This may create an acute on chronic mode of presentation, mistaken for hemorrhage within a tumor or misdiagnosed as another more typically aggressive tumor.

CG frequently presents with multiple nonspecific symptoms (39 cases) rather than with one presenting feature (11 cases), probably due to location. Endocrine presentations are uncommon, occurring in only six cases. Memory impairment, occurring in 12 patients, is more site specific, and combined with radiological factors may be sufficient to include CG on the list of differential

diagnoses. Visual field deficit is the most common abnormality on examination. Ophthalmic assessment should therefore be thorough and may be used for monitoring purposes.

Radiological Findings

Only 32 of the 53 cases were radiologically typical for CG: midline in the third ventricle, well circumscribed, round or ovoid, and uniformly enhancing. The most common atypical feature was a cystic component, present in 14 cases. The rarity of hemorrhage within CG (two cases) may be testimony to its slow-growing nature.

Natural History of CG

Experience is limited to two cases: one a 71-year-old woman presenting with chronic visual failure.¹⁸ The patient received only serial visual field monitoring, which did not change. Follow-up duration is not specified. The second, a 37-year-old woman, was managed by shunt, biopsy, and radioactive implants for 9 months before interval MRI showed considerable extension necessitating surgery.⁶ A conservative approach to this tumor may therefore only be considered in asymptomatic or minimally symptomatic patients, and given reported periods of rapid tumor enlargement, this strategy is likely to have an appreciable failure rate. Periods of rapid growth in tumors under surveillance may be more common in female patients.^{5,6,19}

Surgical Details

Surgical approaches are typically midline. Subfrontal translamina terminalis approaches offer excellent chiasmal exposure with lesser access to the superior components of the tumor. For tumors with a large intraventricular component, a transcallosal or transcortical transventricular approach may be used, especially in the presence of preoperative hydrocephalus. The transphenoidal approach has been used once; we suspect that it would be unlikely to offer appropriate access for such a tumor, especially given the propensity for CG not to cause sellar expansion. Pterional craniotomy has been used to approach lateralized CG, but achieving major resection with this approach is very challenging. There will surely be a future role for endoscope-assisted resection of the tumor in the third ventricle.

Ventricular drainage has been used infrequently. It was not associated with infection in the five patients who were subsequently shunted.

Postoperative Complications

Postoperative complications following resection of CG are common with only seven patients following an

uneventful postoperative course. There is little information available on performance status following treatment. The commonest outcome measures are absence of new neurological deficit, visual improvement, and death.

Venous thromboembolism prophylaxis and recognition are vital as CG appears to be associated with a high rate of thrombotic complications postoperatively, especially fatal pulmonary embolism.

Postoperative hypothalamic-pituitary-axis disturbance is frequent and may be severe. Diabetes insipidus, typically transient, was the most common. Amnesia (a common presenting symptom) was the next most common postoperative complication, occurring in 10 of the 39 cases that reported complications.

Death is most likely to occur in the first month postoperatively, with 10 out of 15 deaths in this period. This number includes deaths from pulmonary embolism and cardiovascular causes, which if ignored, make death in the first month postoperatively unlikely.

Adjuvant Treatment

Adjuvant radiotherapy has been used following subtotal resection.¹ The type of radiotherapy given is associated with recurrence risk. Iwami et al discuss the promising role of adjuvant gamma knife radiosurgery as it was not associated with recurrence in the three patients who received it.²⁰ External beam radiotherapy, however, was associated with recurrence in all three patients treated. Nakajima et al used gamma knife as an adjuvant after resecting only 70% of the tumor mass, finding stable residuum at 3.5-year follow-up.²¹ More experience is required with postoperative radiosurgery to establish if it should routinely follow subtotal resection to minimize recurrence. Using adjuvant radiosurgery may reduce the need for aggressive resection and the attendant morbidity. The efficacy of radioactive implants is not established. It has been attempted on two patients, one of whom had no recurrence and one of whom died of sepsis in the early postoperative period. There is no report of adjuvant chemotherapy.

Progression following incomplete removal was recorded in five cases. Recurrence was rare following total macroscopic resection, occurring once in 24 cases, with maximum follow-up available 68 months. Early recurrence is reported: Jung and Jung report recurrence in a 50-year-old woman who underwent subtotal resection via the transcallosal approach. Recurrence occurred 3 months postoperatively, and the patient died a month later after no further intervention.¹¹ One of Brat's cases showed recurrence within the residual tumor at 5 months and death at 8 months of unrelated causes in a 47-year-old woman.¹ Late recurrence has also been described, occurring 3 years following subtotal resection and adjuvant external beam radiotherapy.¹ This CG was found

Table 8 Pediatric Cases of Chordoid Glioma

12-y-old boy ¹⁷	Radiological features were unusual as the mass was heterogeneously contrast enhancing. Histologically, there was less lymphocytic infiltration and no Russell bodies. This case had considerable chordoid metaplasia, which is not a recognized feature of adult cases.
5-y-old boy ¹⁶	Histological findings were consistent with chordoid glioma except lack of lymphoplasmic infiltrates with Russell bodies and lack of reactive tissue plane surrounding the tumor. It is possible that the tumor was growing so rapidly in this case, as manifest by the acute presentation (2-d history), that there was insufficient time for reactive tissue formation.
7-y-old girl ¹⁰	Again, radiological features were unusual. The tumor was located in the corona radiata, was of mixed density, and showed heterogenous contrast enhancement. Ultrastructural analysis showed neurosecretory granules and intermediate filaments, which may suggest tanyctic origin.

at postmortem to fill the entire third ventricle, again supporting the observation of rapid growth periods in women. Another patient, who received external beam radiotherapy following subtotal resection, showed small and asymptomatic recurrence at 4 years.

Reoperation for recurrence is little described. Hanbali et al performed subtotal resection and radiotherapy for CG.²² The patient represented 1 year later and underwent a second operation where total resection was achieved. The patient died after 3 months from myocardial infarction. Reoperation for raised intracranial pressure was performed 5 days after the first operation in a 36-year-old man, with no detailed discussion of outcome.⁸ From the cases of aggressive recurrence and rapid growth presented, cautious reoperation in the context of recurrence or progression is probably advisable with adjunctive radiotherapy or radiosurgery. No factors, other than incomplete resection and possibly use of conventional rather than gamma knife adjuvant radiosurgery, predisposing to recurrence could be found from our review.

Only three pediatric cases have been described and all have unique clinicopathologic features, summarized in Table 8. It is difficult to comment on the long-term outcome of CG in children. One child died 2 days postoperatively from a cardiac arrest.¹⁶ However, the tumor may be more aggressive in children, manifest by the acute presentations, short history, hemorrhage within the tumor, and large size.¹⁶

CONCLUSION

The treatment-related morbidity of CG is very high. Outcomes will only be improved by better preoperative recognition of this tumor entity, allowing a more cautious approach to definitive surgery. A low threshold should be maintained for the use of radiosurgery in the context of subtotal removal or early recurrence.

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