Posterior Cranial Fossa Gangliogliomas

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ABSTRACT

Ganglioglioma (GG) is an uncommon primary lesion of the central nervous system that is typically located supratentorially. There are only a few reports of GG arising from the cerebellum. To the best of our knowledge this is the first case of a cerebellar GG with supratentorial extension and a longstanding history before its recognition. In fact, this 29-year-old male presented with an 11-year history of intermittent headaches. A cranial computerized tomography (CT) performed at the onset of his complaints failed to reveal the tumor. After a particularly longstanding cephalalgic episode, the patient underwent a new CT scan that was also negative. However, magnetic resonance (MR) imaging of the brain revealed a space-occupying lesion in the right cerebellar hemisphere with extension to the level of the superior colliculi and pineal recess. The tumor was partially removed through a midline suboccipital craniotomy and supracerebellar approach. Pathological examination of the tumor showed composition of atypical ganglion cells and astrocytes, indicating the diagnosis of cerebellar GG. At last follow-up, 24 months after surgery, the patient reported a marked improvement of his clinical condition with significant reduction of intensity and frequency of the headache. The present report illustrates how cerebellar GG may remain undetectable by CT and may therefore present with a longstanding history and nonspecific signs and symptoms. MR investigation can lead to the proper diagnosis. Even after partial removal the prognosis remains good and remission of the symptoms may be achieved. In this article, we review the literature and summarize the current understanding of infratentorial GGs.

KEYWORDS: Ganglioglioma, posterior cranial fossa, cerebellum, brainstem, neuronal tumors, history

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Ganglioglioma (GG) is an uncommon primary lesion of the central nervous system, accounting for 0.4 to 1.3% of all intracranial neoplasms.¹ Eighty percent of the GGs occur in patients younger than 30 years with a peak age of incidence between 10 and 20 years.² GG is typically a supratentorial lesion with the majority of the tumors occurring in the temporal lobe, followed by the parietal and frontal lobes, respectively, although it may be found at virtually any location of the central nervous system.³ Infratentorial GGs are rather rare with only a few reports in the literature. To our knowledge, including our case, only 30 cases of cerebellar GG have been described^{1,4-32} (Table 1). Cerebellar GGs have a generally short mean history (1.6 years), whereas longer histories have been found in cases of brainstem GGs^{4,7,29,33-47} (Table 1).

We present here a rare case of cerebellar GG supratentorial extension to the superior colliculi and pineal recess. This is, to the best of our knowledge, the first case of a cerebellar GG with a longstanding history of more than 10 years. This case adds to the growing knowledge about the findings, presentation, and management of this entity. We review the literature and summarize the current understanding of infratentorial GGs.

CASE REPORT

History and Findings

A 29-year-old male with an 11-year history of intermittent headache was referred to our institution. A previous cranial computerized tomography (CT) obtained 10 years before had not shown any pathological findings. One month before admission the headaches worsened and the patient experienced a longstanding cephalalgic episode. A new neuroradiological workup was performed. A cranial CT scan was done, the results of which were interpreted as negative (Fig. 1A). A brain magnetic resonance (MR) imaging study was also obtained and revealed a space-occupying lesion of the right cerebellar hemisphere

with extension to the level of the superior colliculi and pineal recess and compression of the right cerebral peduncle. On a T2-weighted MR image the tumor had mixed signal intensity, with cystic-appearing areas of high signal intensity and solid-appearing areas of low signal intensity. The tumor also showed mixed areas of contrast enhancement (Fig. 1B, C, D). No gait disturbance, limb ataxia, nystagmus, or cranial nerve deficits were noted at neurological examination. The results of other medical and hematological examinations were also unremarkable.

Operation and Pathology

The patient underwent a removal of the lesion via a median suboccipital craniotomy in a semisitting position using a supracerebellar approach to provide tumor decompression and tissue sample. The tumor appeared as a vascular mass and had a mixed soft and hard consistency. Removal of the dorsal aspects of the lesion was performed. However, total resection of the tumor was not feasible since the tumor was found to be extremely adhesive to the surrounding structures. In particular it could not be separated from normal vessels. Neuropathological examination confirmed the diagnosis of GG.

Postoperative Course

The postoperative course was uneventful. At the last follow-up 24 months after surgery, clinical examination revealed no neurological deficits. The patient reported a significant reduction of intensity and frequency of the headache. Control MR 12 and 24 months after surgery showed a tumor remnant without any tumor growth (Fig. 2).

DISCUSSION

The term *ganglioglioma* was originally used in 1870 by Loretz and was later popularized by

Table 1 Summary of Reported Cases of Brainstem and Cerebellar Gangliogliomas in the Literature

Year	Authors	Case(s)	Age	Sex	Duration of		Manifestations	Site of Tumor	Therapy	Reference
					Symptoms					
1911	Bielschowsky	1	26	M	15 years		–	Brainstem	Autopsy	4, 33
1911	Pick and Bielschowsky	1	24	F	–		–	Brainstem	Autopsy	4, 33
1932	Foerster and Gagel	2	11	M	3 years		Nystagmus, quadripareisis, syncope	Brainstem	Autopsy	4, 33
1966	Tommasi et al	1	14	M	6 years		Nystagmus, ataxia, syncope	Brainstem	Autopsy	
1966	Richmond et al	1	23	M	3–4 months		–	Brainstem	Partial removal	4
1974	Feigin and Budzilovich	2	4	M	2 weeks		Headache	Brainstem (pons)	Autopsy	4, 34
			18	F	Several months		Headache	Brainstem	Autopsy	4
1976	Scalley	1	9	F	7 years		Headache, vomiting, nystagmus, ataxia	Brainstem	Autopsy	
1978	Garrido et al	3	–	–	–		–	Cerebellum	–	4
			12	F	2 years		–	Brainstem	Partial removal	35
			12	M	1 year		–	Brainstem	Biopsy	
1979	Probst et al	1	13	F	4 months		–	Brainstem	Partial removal	
			2	M	–		Increased ICP, cerebellar symptoms	Cerebellum	Surgical removal	5
1979	Friedman et al	1	9	F	7 years		Dizziness, swallowing difficulties, syncope	Brainstem (medulla)	Biopsy	33
1979	Mork et al	1	22	F	1 year		Headache, unsteadiness, slurred speech	Cerebellar hemisphere	Total removal	6
1982	Epstein et al	1	6	M	16 months		Hemiparesis, hearing loss,	Brainstem	Partial removal	36
1984	Garcia et al	1	19	M	1 year		Hemiparesis, diplopia, headache	Brainstem (pons)	Shunting followed by biopsy	4
1985	Fukuoka et al	1	Infant	–	–		–	Cerebellum	Surgical removal	7
1986	Martin et al	1	–	–	–		–	Brainstem	Partial removal	37
1987	Nelson et al	1	5	M	5 years		Vomiting, apneic spells	Brainstem (medulla)	Autopsy	38
1987	Dhillon	1	8	F	1 year		Hearing loss	Cerebellar hemisphere, peduncle	Biopsy	8
1987	Mizuno et al	1	–	–	–		–	–	–	39
1988	Harada et al	1	22	F	–		Headache, vomiting, gait disturbance	Cerebellar hemisphere, vermis	Total removal	9

Table 1 (Cont)

Year	Authors	Case(s)	Age	Sex	Duration of Symptoms	Manifestations	Site of Tumor	Therapy	Reference
1990	Turgut and Ozcan	1	-	-	-	-	Cerebellar vermis	-	10
1990	Kimura and Suzuki	1	5	F	1 year	Headache	Cerebellar hemisphere	Subtotal removal	11
1991	Bills and Hanieh	1	2	M	Newborn	Facial spasm	Cerebellar peduncle, fourth ventricle	Subtotal removal	12
1991	Nishizawa et al	1	14	M	-	Severe headache, confusion	Cerebellum	Total removal	13
1992	Hirose et al	1	12	F	-	-	Cerebellar peduncle, medulla, Brainstem	Autopsy	14
1992	Mickle	3	-	-	17 years	-	Brainstem	Partial removal	40
1992	Lindboe et al	1	27	M	1 year	Headache, nausea, ataxia	Cerebellar hemisphere, vermis	Subsequent total removal	15
1992	Geyer et al	1	-	-	-	-	Cerebellum	Surgical removal	16
1993	Chang et al	1	-	-	-	-	Posterior fossa	-	17
1993	Pollack et al	1	-	-	-	-	Brainstem	Subtotal removal	41
1993	Lang et al	9	-	-	-	Cranial nerve dysfunction	Brainstem	-	42
1994	Handa et al	1	53	M	-	-	Cerebellum	Total removal	18
1994	Osanal et al	1	44	M	-	Hypoventilation, Apnea	Superior cerebellar peduncle, Posterior Fossa	-	19
1994	Nixon et al	1	-	-	-	-	Posterior Fossa	-	43
1994	Zentner et al	2	-	-	-	-	Infratentorial	Surgical removal	1
1995	Blatt et al	1	4	M	4 years	Gait disturbance, hemiparesis, ataxia	Cerebellar hemisphere, peduncle	Shunting followed by biopsy	20
1995	Karamitopoulou et al	1	38	M	16 years	Headache, gait disturbance, hemiparesis	Brainstem	Partial removal	44
1995	Wang et al	1	14	-	-	-	Cerebellum	-	21
1996	Harvey et al	1	6 months	F	At birth	Hemifacial contraction, nystagmus	Cerebellar hemisphere, peduncles	Total removal	22
1997	Jay and Greenberg	1	14	M	-	-	Cerebellum	Surgical removal	23
1997	Yachnis	1	-	-	-	-	Brainstem	-	45
1998	McLone et al	1	1 month	M	At birth	Epilepsy	Cerebellar peduncles, vermis	Subtotal removal	24
1998	Gleckman and Smith	1	3	F	-	Apnea	Brainstem (medulla)	Autopsy	46

Table 1 (Cont)

Year	Authors	Case(s)	Age	Sex	Duration of		Manifestations	Site of Tumor	Therapy	Reference
					Symptoms					
2000	Vinchon et al	1	10	F			NF1, asymptomatic	Cerebellum	Surgical removal	25
2000	Evans et al	1	60	M	1 month		Headache, dizziness	Cerebellar vermis	Subtotal removal	26
2001	Farmer et al	3	9	-	1 year		-	Brainstem, cervicomedullary	Radical subtotal removal	47
			12	-	1 year		-	Brainstem, cervicomedullary	Radical subtotal removal	
			6		1 year		-	Brainstem, cervicomedullary	Subtotal removal	
2001	Kwon et al	1	2	M	2 years		Developmental delay	Brainstem, cervicomedullary	Total removal	27
								Cerebellum, cerebellopontine angle		
2001	Chae et al	1	4 months	M	At birth		Paroxysmal facial contraction, nystagmus	Superior cerebellar peduncle	Partial removal	28
2001	Lagares et al	3	59	M	1 year		Dizziness, syncope	Floor of fourth ventricle	Shunting followed by removal	29
			19	F	1 year		Headache	Brainstem, vermis	Shunting followed by removal	
2002	Kinoshita et al	1	36	F	1 year		Gait instability, diplopia	Brainstem	Subtotal removal	30
2002	Mesiwala et al	1	28	M	-		Headache, ataxia	Cerebellar vermis	Subtotal removal	31
			11 weeks	M	At birth		Focal seizure with second generalized	Cerebellum	Surgical removal	
2003	Mink et al	1	22 months	M	-		Progressive myoclonus	Cerebellar peduncles	Surgical removal	32
2006	Safavi-Abbasi et al	1	29	M	10 years		Headaches	Cerebellar hemisphere with supratentorial extension	Subtotal removal	Present case

Figure 1 The (A) axial computed tomography study does not reveal the lesion that becomes clearly evident in (B) axial (C) contrast-enhanced coronal T1-weighted, and in (D) axial T2-weighted magnetic resonance imaging images. The tumor appears to arise from the right cerebellar hemisphere with compression of the right cerebral peduncle and extension to the level of the superior colliculi and pineal recess.

Perkins in 1926.^{3,48} Zuelch found GG in 0.4% and Cushing in 0.3% of a comprehensive series of brain tumors.¹ In more recent reports GG accounts for 0.4 to 7.6% of pediatric and up to 1.3% of adult central nervous system neoplasms.^{1,3,48} In 1911 Pick and Bielschowsky, and later in 1932 Foerster and Gagel, provided the first documented reports of infratentorial and medullary GG.³³

Histology

In 1930 Courville^{49,50} correctly acknowledged the mixed histologic composition of mature ganglion cells and glial cells of varying proportions and degrees of differentiation. Both components usually exhibit low grades of malignancy, but it is the grade of the glial element that predicts the biological behavior.

Figure 2 Contrast-enhanced sagittal T1-weighted MR (A) preoperative and (B) 24-month postoperative studies. The remnant of the tumor does not compress the surrounding structures.

GG is part of the family of mixed glial-ganglion cell tumors that are included within grade I or II of the WHO classification; however, occasional cases of anaplastic GG (WHO grade III) have been reported.⁵¹ Malignant degeneration is also rare, with an estimated incidence of 6%. One report⁵² describes such degeneration even 23 years after the initial diagnosis. Although anaplastic transformation of both the glial and neuronal components has been reported, malignant changes appear to be confined mostly to the glial element of the tumor.^{51,53} Leptomeningeal and subarachnoid spread is uncommon but may occur.⁵⁴

Epidemiology of Infratentorial GG

Infratentorial location of GG is rarely found. In the series by Chang and associates,¹⁷ of 133 posterior cranial fossa tumors, only 1 case of GG was described. Farmer et al,⁴⁷ in their 10-year review of brainstem gliomas in 2001, found 3 cases of GG and Goncalves-Ferreira and coworkers⁵⁵ found one case of GG in their series of 30 cases with focal brainstem lesions in 2003.

Both cerebellar and brainstem GG are rather rare entities, with few anecdotal cases in the literature (Table 1). In 1984, reviewing the literature, Garcia and coworkers⁴ reported only 14 cases of brainstem GG, and more recently in 2001, Lagares et al²⁹ found 31 cases of brainstem GG in the literature. Lang and associates⁴² in their series of

58 patients with GG have the largest series on infratentorial GG, including 9 brainstem GGs.

Similarly, though cerebellar location of GG was considered previously as the second most frequent site of GG,⁶ in recent series, the temporal lobe has been found to be most commonly affected, followed by the parietal and frontal lobes, respectively.^{1-3,48,56}

Brainstem localization seems to be generally associated with a longer history than cerebellar GG (Table 1). To our knowledge, the case we present here is the first documentation of an infratentorial cerebellar GG extending supratentorially to the pineal recess with a longstanding history.

Clinical Presentation

Clinical presentation of infratentorial GG varies depending on the structures involved. Associated symptoms include cranial nerve deficits (hearing loss, intractable facial pain, hemifacial seizures), hemiparesis, cerebellar signs, gait disturbance, and headache.^{8,20,22,33,36,29,57}

Because of their location, brainstem GGs may cause respiratory problems, syncope, and sudden death.^{33,29} In fact, many of the previously reported brainstem GGs were only detected post-mortem (Table 1). In our patient, the location and size of the tumor as well as the need for pathologic tissue sample more than the symptoms initiated the decision for surgical resection.

Interestingly, there are some reports describing epilepsy of cerebellar origin in patients with cerebellar GG.^{22,24,28,31,32} Whether the cerebellar tumor either initiates the seizure or lowers their threshold is still debated.²⁴ Although the exact mechanism for the epilepsy arising in the cerebellum is not known, invasive electrophysiologic monitoring with depth electrodes has confirmed the cerebellum as the site of seizure origin.²² It is thought that the seizure arises in the cerebellum and may generalize secondarily, spreading throughout the cortical surface.^{22,28,31} Surgical resection of GG has resulted in remission of the seizures in all reviewed cases.^{22,24,28,31}

The age at presentation in infratentorial GG shows a wide range, beginning at 2 weeks to 59 years in brainstem GG and from 11 weeks to 60 years in cerebellar tumors (Table 1). In general, the tumor affects mostly patients younger than 30 years with a peak age of incidence between 10 and 20 years.^{1,3,29}

Radiology Findings

GGs have variable manifestations on CT scans, mostly presenting with low density.^{1,3,20} Calcifications are seen occasionally and were reported in the literature in 20 to 50% of the cases.^{1,3,20,58} The degree of iodinated contrast enhancement is variable, occurring in 16 to 80% of the patients.³ The neoplasm may be, as in our case, undetectable on CT.^{1,3} This may delay the diagnosis and affect the patient's outcome, especially since it has been reported that early operative intervention improves the outcome.⁴² It is particularly important to take this into account in patients with seizures of unknown origin, with a long history of headache, as in our case, or with pathologic cerebellar or brainstem signs without CT findings. In fact, MR imaging has proved to be more sensitive in identifying GGs. However, the MR appearance is also variable and nonspecific, with a spectrum of signal intensity ranging from hypointense to hyperintense on short MR images and hyperintense relative to the gray

matter on long TR images.^{3,20} The lesion may appear to be cystic, solid, or mixed with nonspecific gadolinium enhancement.¹ Cystic components have been reported in 57%^{1,59,60} and gadolinium enhancement in 44% of the cases.¹ Overall the CT and MR appearance of infratentorial GGs do not seem to be specific or different from other supratentorial tumors.^{20,37,29}

Although the tumor usually appears as an intra-axial avascular mass, extra-axial or vascular presentation may occur.^{61,62} Furthermore, angiographically avascular and nonenhancing tumors on MR may be found to be vascular during surgery.²⁷ Our case presented as a cystic, enhancing mass on T2, with heterogeneous signal enhancement on the T1 images. The tumor was found to be extremely vascular intraoperatively.

Proton magnetic resonance spectroscopy, positron emission tomography, and single-photon emission computed tomography have also been used as tools for differentiating cerebral and cerebellar GGs from other tumors and determining the degree of anaplasia.^{21,59,62,63} However, there seems to be a discrepancy between the high metabolic activity and membrane synthesis, indicative of malignancy, and the usually benign histologic appearance of GG.^{3,63}

Management of Infratentorial GG

In the past, radiotherapy has been used for treatment of some brainstem GGs but its usefulness has yet to be proven.^{5,29,64-66} The deleterious side effects on the developing nervous system have been a strong argument against use of this treatment modality in children with this lesion.^{56,67} In fact, some reports further suggest that postoperative radiation may predispose a GG to malignant degeneration.^{51,53,58,66,68} In one comparative study of supratentorial lesions, malignant progression occurred in 4 cases of 14 irradiated GGs, but in none of 14 nonirradiated GGs.⁶⁸ It has been therefore stated that radiation therapy and chemotherapy should be reserved for unresectable and malignant GGs or for those patients with tumor progression.^{1,35}

Gross total resection is the recommended treatment of choice for these tumors and complete tumor resection is associated with good prognosis.^{1,3,29} However, because of the location and vicinity to vital and sensitive brain areas and their adherence to the tumor, complete resection of infratentorial GG is often not possible without producing severe deficits or even death, thus leaving only space for subtotal resection.^{4,8,20,26,29,33,34} Nevertheless, the diagnosis of GG is of great importance because of its comparatively better prognosis than other tumors in this location. Farmer and coworkers⁴⁷ in 2001 in their series of brainstem gliomas identified the diagnosis of GG in this location as a favorable prognostic factor.

Infratentorial GGs usually have a benign clinical course. In fact, although infratentorial GGs frequently are resected only partially, the overall survival time does not seem reduced as compared with supratentorial GG where a more radical resection can be achieved.²⁹ Further, invasiveness on radiographic studies or anaplasia within a GG seem not to correlate with clinical outcome, but the risk of recurrence is higher for brainstem and spinal GG.^{20,42} In general, patients with GG, regardless of the location, tend to have a good prognosis.⁴²

CONCLUSIONS

Infratentorial GGs are rare tumors that may remain undetectable on cranial CT scans and may present with a longstanding history of nonspecific complaints. Even with partial resection, prognosis remains good because of the benign tumor biology. The treatment for posterior fossa GG should therefore include subtotal resection, if gross resection is not possible.

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Commentary

This is a very well-described case and literature review of an unusual posterior fossa tumor presenting with long-term headache. The tumor failed to be detected by two CTs and was diagnosed with an MRI. Only a subtotal resection was possible, as would be expected in this location. The review of the literature is excellent. In summary, it should be noted that ganglioglioma should be in the differential diagnosis of cerebellar and brainstem intrinsic tumors. No definite distinguishing features on MRI have been found, although they do enhance with gadolinium. Consideration of this tumor may support biopsy of an intrinsic brainstem tumor since the prognosis is good for overall longevity compared with other glial tumors. In their review, the tumor infrequently undergoes malignant degeneration. I commend the authors on their presentation.

Chandranath Sen, M.D.¹

Commentary

The authors illustrate an interesting case of a posterior fossa ganglioglioma that presented with over a decade of chronic headaches. An initial CT study performed 10 years prior had failed to reveal the lesion. Ultimately, an MRI study showed a cerebellar mass and direct biopsy secured the diagnosis.

This case is interesting for a variety of reasons. The long duration of symptoms before diagnosis could no doubt have been shortened by early MR imaging, and this raises the possibility that patients with prolonged or severe headache histories

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should be aggressively imaged. The fact that the headaches improved after subtotal resection and biopsy suggests that surgery may still play a role even for tumors that cannot be fully resected. While I do not typically employ the sitting position for a supracerebellar approach, I agree that this avenue of attack offers the best exposure to the region.

It is notable that the chronic presentation (11 years) more closely followed the expected history of brainstem gangliogliomas rather than similar cerebellar tumors. With its localization at the right cerebellar peduncle with extension toward the tectum and pineal recess, the tumor might also be classified as a brainstem lesion. Without serial

studies, it would not be possible to define precisely exactly where it arose.

The article also very succinctly summarizes the known literature for cerebellar gangliogliomas and offers reasonable recommendations regarding adjuvant treatment. Given the generally favorable prognosis, even for brainstem locations, chemotherapy and radiation are reserved for progressive growth or malignancy. The impact of radiosurgical treatment has yet to be defined. No doubt, this patient must have regular follow-up with serial MR imaging to assess for tumor growth and hydrocephalus.

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