Journal of
Medical Case Reports

CASE REPORT

Open Access

Dorsally exophytic brain stem ganglioglioma extending to the foramen of Luschka: a case report

Ege Anil Ucar[1*], Utku Ozgen[2] and Talat Kiris[2]

Abstract

Background Gangliogliomas are rare tumors primarily arising from the central nervous system, mostly in the temporal lobes, with brain stem involvement being particularly infrequent. To the best of our knowledge, this is the first reported instance of a brainstem ganglioglioma exhibiting an extension to foramen of Luschka.

Case presentation We present a unique case of ganglioglioma of the brainstem. 23-year-old Turkish patient presented with flashing lights in the peripheral visual fields. Imaging studies revealed a distinct mass lesion adjacent to the brainstem, demonstrating an unusual exophytic growth pattern that extended towards the foramen of Luschka. Surgical intervention was performed to prevent tumor progression and obtain a definitive diagnosis. The surgical approach employed was the telovelar approach, which provides excellent visualization of the posterior fossa. Histopathological examination of the resected specimen confirmed the diagnosis of grade 1 ganglioglioma. Postoperative magnetic resonance imaging scans displayed gross total resection of the tumor. The patient's postoperative course was uneventful, and the initial symptom of flashing lights resolved in the postoperative period.

Conclusion This case report highlights the uniqueness of a dorsally exophytic brain stem ganglioglioma extending to the foramen of Luschka. Utilization of the telovelar approach and sodium fluorescein in the surgical management of this challenging case underscores its efficacy in managing deep-seated lesions within the posterior fossa. Although presenting infrequently, gangliogliomas should be considered in the differential diagnosis of lesions of the foramen of Luschka because early recognition is important for the management and prognosis.

Keywords Brainstem ganglioglioma, Case report, Foramen of Luschka, Dorsally exophytic, Telovelar approach, Sodium fluorescein

Background

Gangliogliomas are uncommon neoplasms of the central nervous system (CNS) composed of dysmorphic ganglion cells and a glial component. These tumors account for approximately 0.4% of all CNS tumors and 1–10% of pediatric CNS tumor [1]. The reported median ages of these tumors range from 10 to 30 years. While gangliogliomas are predominantly found in the temporal lobe, they can manifest in various CNS locations, presenting a diverse clinical picture [2].

Epilepsy is the most common presenting symptom of ganglioglioma patients, comprising 85% of cases [3]. Magnetic Resonance imaging (MRI) findings typically reveal a hyperintense T2-weighted image (T2WI) and iso/hypointense T1WI with variable contrast enhancement [4, 5]. The majority of gangliogliomas present as grade 1 lesions, accounting for 95% of the cases.

*Correspondence:
Ege Anil Ucar
eucar17@ku.edu.tr
[1] School of Medicine, Koc University Hospital, 34010 Zeytinburnu, Istanbul, Turkey
[2] Department of Neurosurgery, American Hospital, Istanbul, Turkey

The gold standard for treating gangliogliomas is gross total resection (GTR). With subtotal resection (STR), it is reported that progression-free survival significantly decreases [6].The utilization of chemotherapy and radiotherapy in case of STR is controversial since using radiotherapy exposes the patient to malignant transformation. Here, we present the first case of brain stem ganglioglioma that is dorsally exophytic from the medulla oblongata and extends to foramen of Luschka, thus allowing for GTR.

Clinical presentation

A 23-year-old Turkish man with no additional disease presented because of a 2-week history of flashing of lights on the right visual field. His physical examination was completely normal including cranial nerves (CN), motor and sensory function, reflexes, cerebellar examination, and visual fields. MRI of the patient revealed a 18×14 × 11 mm mass centralized in the right foramen of Luschka. The mass was well demarcated, hyperintense on T2WI, isointense on T1WI, and homogeneously enhanced under contrast (Fig. 1). Differential diagnosis of the MRI was evaluated by radiologists to be pilocytic astrocytoma, choroid plexus papilloma, or hemangioblastoma. Magnetic resonance angiography demonstrated that anterior medullar, lateral medullar, and telovelomedullar segments of posterior inferior cerebellar artery (PICA) surrounded the tumor (Fig. 2).

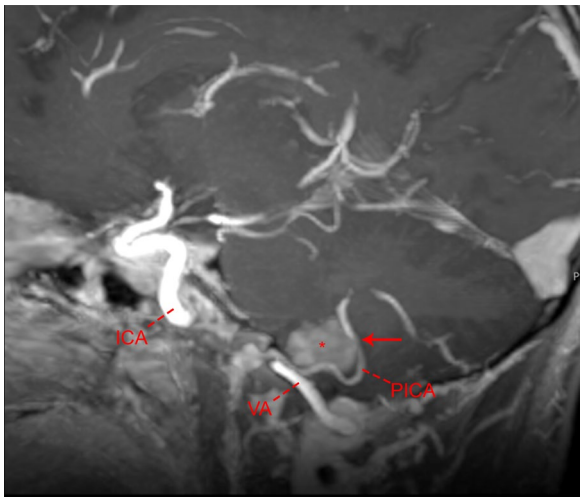

Fig. 2 Magnetic resonance angiography of the brain showing the right vertebral artery (VA), posterior inferior cerebellar artery (PICA), and internal carotid artery (ICA). The posterior inferior cerebellar artery encircles the tumor (*) and passes within the tumor at the location shown (arrow)

Operation

The telovelar approach was performed by using neuromonitorization and neuro-navigation during the surgery. Midline suboccipital craniotomy was performed. Access to the tumor was obtained with the dissection of the right cerebellar tonsil until the uvula. From there, the foramen of Luschka was accessed through the lateral recess of the fourth ventricle. The yellowish tumor, which was above the caudal loop of PICA, lateral to the telovelar segment, below the tonsil, and exophytic through the brain stem, was observed. It was stained with sodium fluorescein and visualized clearly by using the yellow-560 filter of the microscope. A biopsy was taken, and the tumor was debulked by using Cavitron Ultrasonic Surgical Aspirator. The frozen results indicated ganglioglioma. The

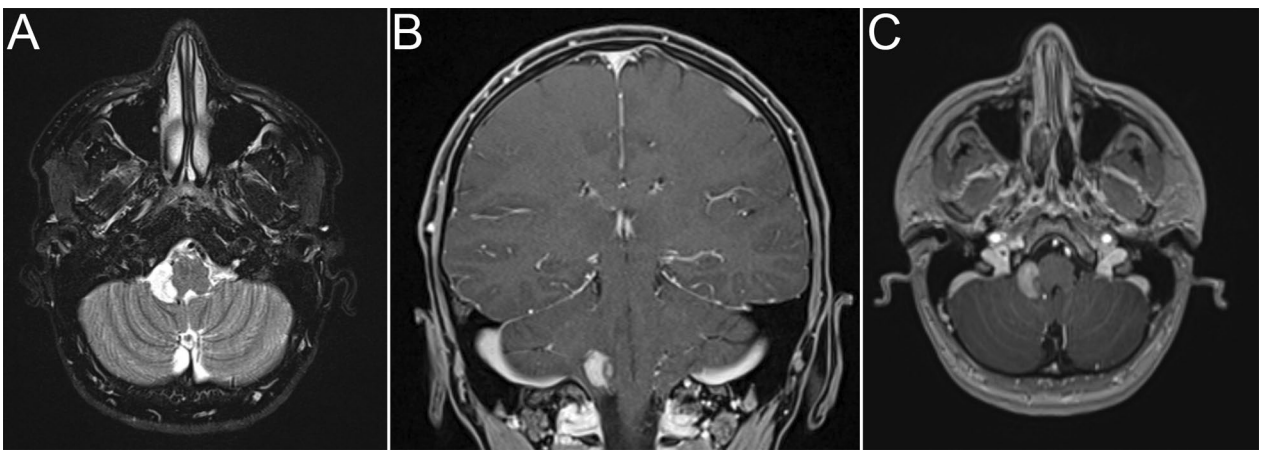

Fig. 1 Preoperative imaging: **A** Axial T2-weighted magnetic resonance imaging scan of the brain showing a hyperintense mass adjacent to the brain stem. **B** A coronal T1-weighted contrast-enhanced magnetic resonance imaging scan showing marked contrast enhancement of the mass. **C** An axial T1-weighted contrast-enhanced magnetic resonance imaging scan showing marked contrast enhancement and the extension of the mass to the foramen of Luschka. Arrows show the well-circumscribed tumor

tumor was dissected from cranial nerves (CNs) 9 and 10 laterally; CN 11 inferiorly; the PICA, and the medulla oblongata.

Postoperative course

Histopathological examination of the tumor revealed astrocytic cells in the piloid morphology and dysmorphic ganglion cells. There were common eosinophilic granular bodies and focal Rosenthal fibrils (Fig. 3). There was not any vascular proliferation or necrosis. Mitosis was not seen, and the Ki-67 score of the tumor was less than 1%. Immunohistochemistry results were positive for synaptophysin and OLIG2 and negative for CD34, NFP, and H3K27M. Therefore, it was diagnosed as ganglioglioma (grade 1; World Health Organization [WHO] 2021). BRAFV600 mutation was negative. The postoperative period was uneventful, with a stable hemodynamic condition. Neurologic examination was normal, and the presenting symptom of flashing lights completely resolved. An early postoperative MRI scan showed GTR of the lesion. MRI at the 3rd, 9th, and 15th months showed stable punctuate contrast enhancement at the exit site of CNs 9, 10, and 11 from the brain stem (Fig. 4).

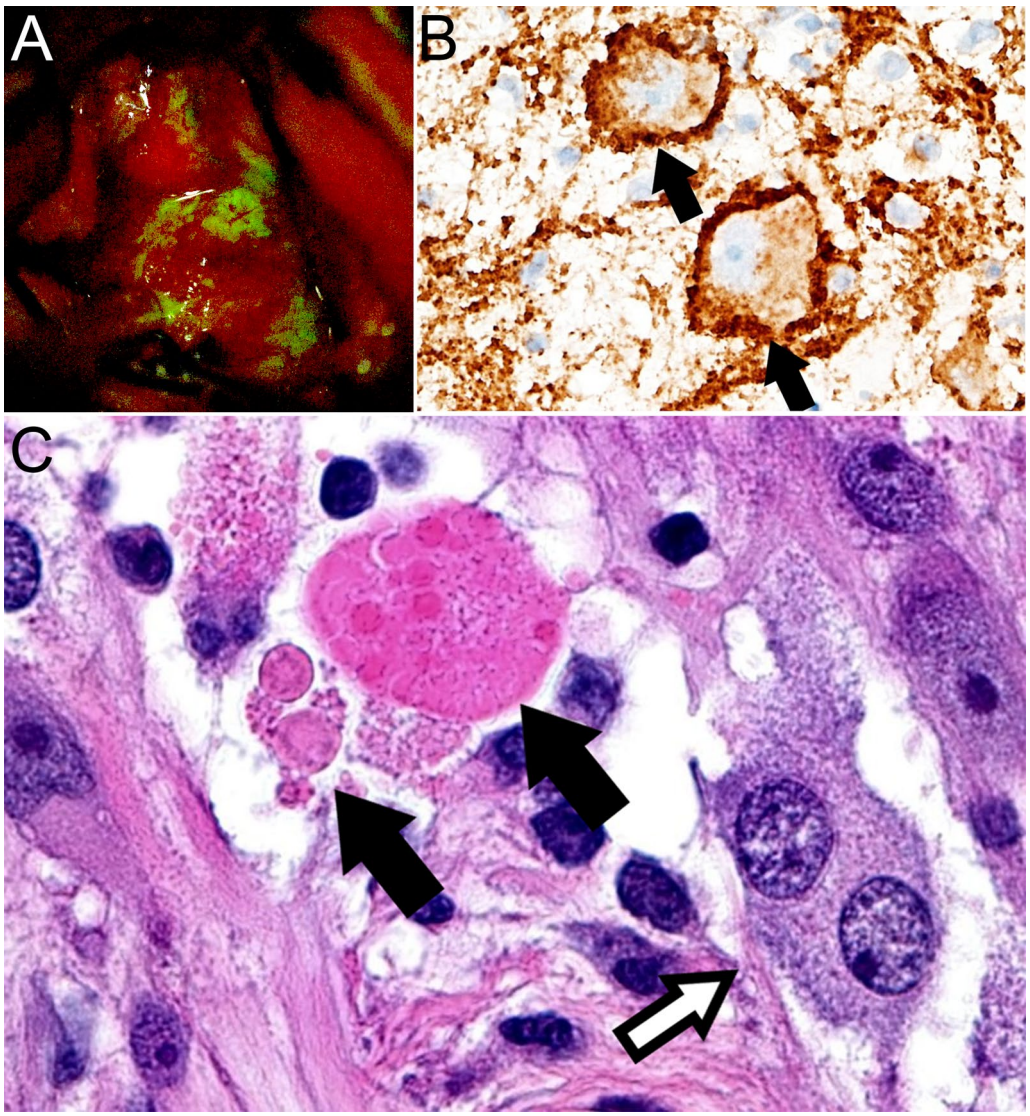

Fig. 3 **A** Intraoperative imaging shows that the tumor exhibits staining with sodium fluorescein. **B** The neoplastic cell's ganglionic feature was shown by synaptophysin positivity. (anti-synaptophysin, 400×). **C** The neoplastic cells were binucleated ganglionic cells (white arrow). There were many eosinophilic granular bodies (black arrow) (Hematoxylin–Eosin, 400×)

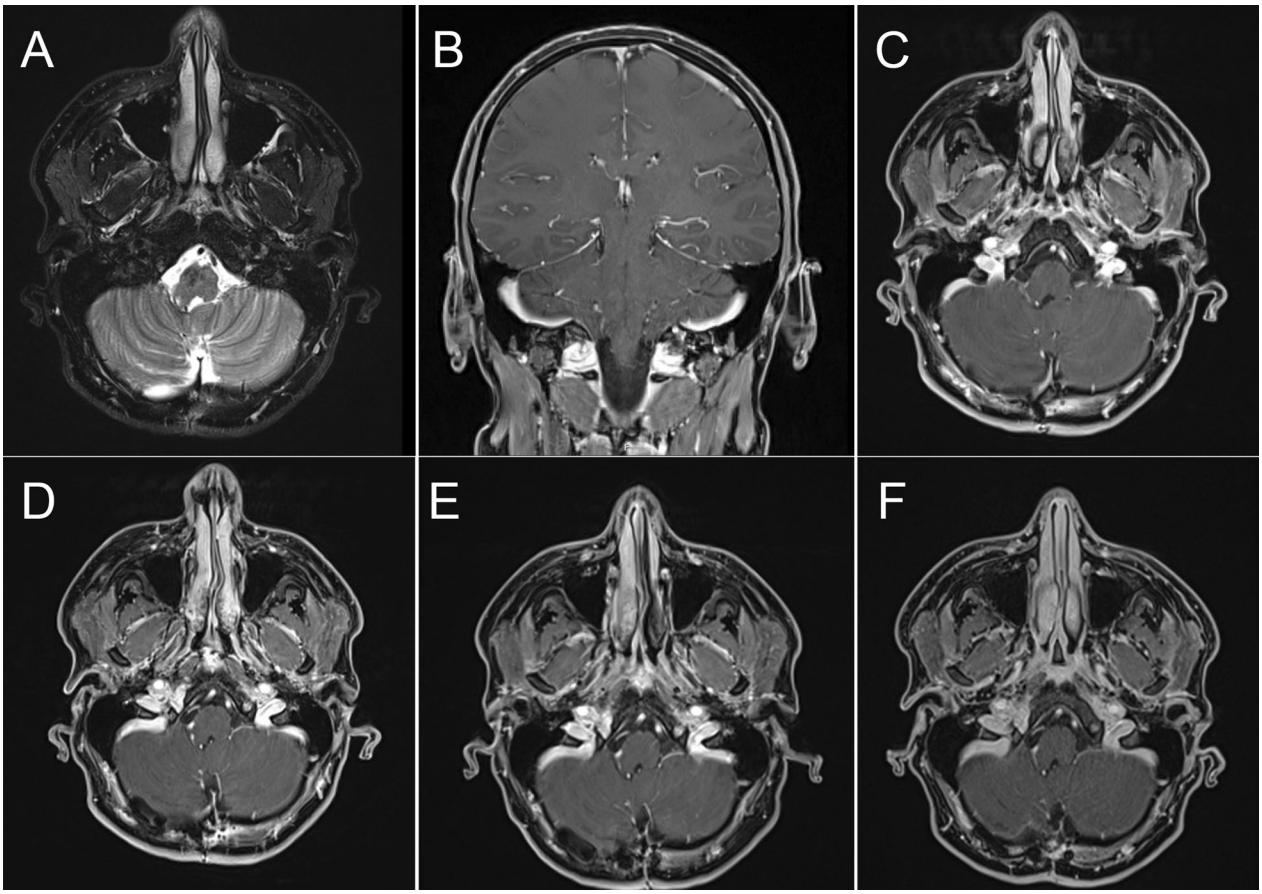

Fig. 4 Postoperative magnetic resonance imaging scan. An early postoperative (**A**) axial T2-weighted magnetic resonance imaging scan shows nonspecific hyperintensities, while (**B**) coronal and (**C**) axial T1-weighted contrast-enhanced magnetic resonance imaging scans not showing any obvious contrast enhancement. Postoperative (**D**) 3rd-month, (**E**) 9th-month, and (**F**) 15th-month axial T1-weighted contrast-enhanced magnetic resonance imaging scans display stable punctuate contrast enhancement at the exit site of cranial nerves 9, 10, and 11 from the brainstem

Discussion

This case report presents an exceptionally rare case of low-grade ganglioglioma, which exhibits an exophytic growth pattern and centralization within the foramen of Luschka. Previous analyses from the Surveillance, Epidemiology, and End Results (SEER) database by Dudley *et al*. revealed that. between 2004 and 2010, there were 348 gangliogliomas, only 13 (3.45%) of which were localized to the brain stem. Similarly, the rarity of these brain stem gangliogliomas has been emphasized by different groups [3, 7, 8]. Most of the brain stem gangliogliomas have exophytic portions in the fourth ventricle or cerebellopontine angle (CPA), referred to as the "transitional form" [9, 10]. These tumors mostly allow for resection of their exophytic component, whereas radical excisions of the brain stem component result in severe morbidity. In the present case, the tumor, unlike the transitional forms, was exophytic and extended to the foramen of Luschka (Fig. 1), allowing for GTR.

The most common presentation of gangliogliomas is epilepsy (85%) at the supratentorial compartment. However, at the infratentorial compartment, patients present with various findings, such as CN deficits, ataxia, hydrocephalus, and motor or sensory deficits [3]. In our case, the patient's only symptom was paroxysmal lights in the right peripheral visual field.

Gangliogliomas are mixed glioneuronal tumors of the CNS comprising mature dysmorphic ganglion cells and various types of glial cells. Atypical glial component and higher Ki-67 values are seen in grade 2 tumors. Anaplasia, hypervascularity, and necrosis are responsible for grade 3 gangliogliomas [11]. In our case, the tumor was composed of dysmorphic ganglion cells and astrocytes in piloid morphology (Fig. 3). Hypervascularity or necrosis was not seen, and the Ki-67 value was less than 1%. Therefore, it was diagnosed as a grade 1 ganglioglioma. Given the benign nature of grade 1 gangliogliomas, adjuvant therapy was not recommended.

The telovelar approach and transvermian approach are the two approaches performed the most for accessing the fourth ventricle. The transvermian approach is widely used for lesions that are in the rostral fourth ventricle and do not spread into the lateral recesses. In contrast, the telovelar approach provides access to lateral recess [12] and foramen of Luschka. Cerebellar mutism can be observed and is usually related to the transvermian approach and damage to the dentate nucleus [13]. In the present case, the tumor was extending to the right foramen of Luschka, so we performed a telovelar approach without cerebellar retraction to have a better view and avoid cerebellar mutism and other cerebellogenic complications.

Gangliogliomas have an excellent prognosis, with the calculated 7.5-year recurrence-free survival rate being 97% [1]. Compton *et al.* also report favorable prognostic outcomes with 94% 15-year survival in their 30 years of experience at the Mayo Clinic [6]. However, when considering the location of gangliogliomas, Dudley *et al.* reports that brain stem gangliogliomas have significantly worse 5-year survival compared with the remaining locations, with rates of 96.6% versus 80.6% [2]. The grade of the tumor is a significant factor affecting the prognosis. Majores *et al.* reported 5-year survival rates of 99%, 79%, and 53% for WHO grades 1, 2, and 3, respectively [14].

One more interesting point of this case report is that the first author, who is a student in his last year of medical school, is also the patient who was operated on. This unique perspective provides valuable insights from the standpoint of both a medical professional and a medical student.

Conclusion

Brain stem gangliogliomas are exceptionally rare CNS neoplasms. Here, to the best of our knowledge, we present a unique case of an exophytic brain stem ganglioglioma that extends to the foramen of Luschka. Gangliogliomas are rarely considered in the differential diagnosis of tumors in this specific location. The telovelar approach provides a safe and effective approach for the excision of the tumors in this region.

Acknowledgements
Not applicable.

Author contributions
EAU, UO, and TK designed the case report. EAU and UO collected the data, prepared all the figures, and wrote the manuscript. EAU, UO, and TK analyzed and interpreted the data. TK made the critical revision and supervised the case report, and all the authors reviewed the manuscript.

Funding
No funding was received for this case report.

Availability of data and materials
Not applicable.

Declarations

Ethics approval and consent to participate
The case report has been performed in accordance with the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards. Consent to participate was obtained from the patient.

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declare that they have no competing interests.

Received: 8 November 2024 Accepted: 6 February 2025
Published online: 01 March 2025

References

1. Luyken C, Blümcke I, Fimmers R, Urbach H, Wiestler OD, Schramm J. Supratentorial gangliogliomas: histopathologic grading and tumor recurrence in 184 patients with a median follow-up of 8 years. Cancer. 2004;101(1):146–55. https://doi.org/10.1002/cncr.20332.
2. Dudley RWR, Torok MR, Gallegos DR, *et al.* Pediatric low-grade ganglioglioma: epidemiology, treatments, and outcome analysis on 348 children from the surveillance, epidemiology, and end results database. Neurosurgery. 2015;76(3):313–9. https://doi.org/10.1227/NEU.0000000000000619.
3. Im S-H, Chung CK, Cho B-K, *et al.* Intracranial ganglioglioma: preoperative characteristics and oncologic outcome after surgery. J Neurooncol. 2002;59:173–83.
4. Kwon JW, Kim I-O, Cheon J-E, *et al.* Cerebellopontine angle ganglioglioma: MR findings. AJNR Am J Neuroradiol. 2001;22:1377–9.
5. Shin JH, Lee HK, Khang SK, *et al.* Neuronal tumors of the central nervous system: radiologic findings and pathologic correlation. Radiographics. 2002;22(5):1177–89. https://doi.org/10.1148/radiographics.22.5.g02se051177.
6. Compton JJ, Laack NNI, Eckel LJ, Schomas DA, Giannini C, Meyer FB. Long-term outcomes for low-grade intracranial ganglioglioma: 30-year experience from the Mayo Clinic. J Neurosurg. 2012;117(5):825–30. https://doi.org/10.3171/2012.7.JNS111260.
7. Lin X, Huang R, Zhang P, *et al.* Low-grade gangliogliomas in adults: a population-based study. Cancer Med. 2021;10(1):416–23. https://doi.org/10.1002/cam4.3577.
8. Zhang S, Wang X, Liu X, Ju Y, Hui X. Brainstem gangliogliomas: a retrospective series. J Neurosurg. 2013;118(4):884–8. https://doi.org/10.3171/2013.1.JNS121323.
9. Baussard B, Di Rocco F, Garnett MR, *et al.* Pediatric infratentorial gangliogliomas: a retrospective series. J Neurosurg Pediatr. 2007;107(4):286–91. https://doi.org/10.3171/PED-07/10/286.
10. Mpairamidis E, Alexiou GA, Stefanaki K, Sfakianos G, Prodromou N. Brainstem ganglioglioma. J Child Neurol. 2008;23(12):1481–3. https://doi.org/10.1177/0883073808319316.
11. Lisievici AC, Pasov D, Georgescu T-A, Lisievici MG, Sajin M. A novel histopathological grading system for ganglioglioma. J Med Life. 2021;14(2):170–5. https://doi.org/10.25122/jml-2021-0054.
12. Tanriover N, Ulm AJ, Rhoton AL Jr, Yasuda A. Comparison of the transvermian and telovelar approaches to the fourth ventricle. J Neurosurg. 2004;101(3):484–98. https://doi.org/10.3171/jns.2004.101.3.0484.
13. Ghali MGZ. Telovelar surgical approach. Neurosurg Rev. 2021;44(1):61–76. https://doi.org/10.1007/s10143-019-01190-5.
14. Majores M, von Lehe M, Fassunke J, Schramm J, Becker AJ, Simon M. Tumor recurrence and malignant progression of gangliogliomas. Cancer. 2008;113(12):3355–63. https://doi.org/10.1002/cncr.23965.

Publisher's Note