

CASE REPORT

Open Access



A case report of rare location of ganglioglioma

Vikas Sharma^{1*} , S. Bhaskar², Sumit Ramdas Hire² and Arvind Ahuja³

Abstract

Background: Gangliogliomas are rare tumors of the central nervous system. They can occur anywhere in the central nervous system but are most commonly located in the temporal lobe and are mainly found in children. Anaplastic ganglioglioma can result from either de novo or transformation of a pre-existing lesion.

Case presentation: We report a case of de novo anaplastic ganglioglioma in the parieto occipital region, which is a rare location. A 34-year-old lady presented with features of raised intracranial pressure (ICP) with right side hemiparesis. Contrast-enhanced magnetic resonance imaging (CEMRI) of the brain showed well-defined intense heterogeneously enhancing solid cystic mass lesion 5.3 × 5.2 cm in the left parieto occipital region with mass effect and midline shift. Intraoperatively, a cystic mass lesion with reddish brown nodule was seen in the left occipital lobe. Complete tumor excision was done. Microscopic and IHC examination was suggestive of anaplastic ganglioglioma. The post-operative period was uneventful. The patient received 60-Gy radiotherapy with temozolamide as adjuvant therapy, and repeat imaging showed no tumor recurrence.

Conclusion: Anaplastic gangliogliomas are rare tumors with parieto occipital as rare location.

Keywords: Ganglioglioma, Tumor, Children, Radiotherapy

Background

Gangliogliomas are rare tumors, and they account for 1% (reported range of 0.4 to 7%) of all central nervous system neoplasms [1]. They are commonly seen in children and young adults with males affected more commonly than females [1]. These tumors occur throughout the CNS, but the temporal lobe is the commonest site [2]. The most common presentation is seizures. They are generally of WHO grades 1 and 2, and grade 3 (anaplastic ganglioglioma) is very rare. In 5% of cases, gangliogliomas show aggressive behavior and anaplastic histopathologic features characteristic of WHO grade 3 tumors [3]. These tumors can occur de novo or as a result of malignant transformation of a pre-existing lesion. We report here a rare case of de novo occipital anaplastic ganglioglioma. On searching the English literature, a total of 41 cases of de novo anaplastic ganglioglioma have been reported with only 4 cases in parieto occipital location.

The details of the previously reported cases have been summarized in Table 1.

Case presentation

A 34-year-old female presented with complaints of on/off headache with vomiting for 1 year, which aggravated for the last 5 days. Systemic examination was within normal limits. GCS was E4V5M6, pupils were bilateral normal size and reacting, on fundus examination-bilateral papilledema was present. On the motor system examination, right side hemiparesis was present (motor power 4/5). Contrast-enhanced magnetic resonance imaging (CEMRI) of the brain showed well-defined intense heterogeneously enhancing solid cystic mass lesion 5.3 × 5.2 cm in the left parieto occipital region. Solid component was isointense on T2/FLAIR images with peripheral cystic areas and isointense on T1W images. The cystic component was iso- to hyperintense on T2/FLAIR images and hypointense on T1W images. The lesion showed mass effect in form of compression and displacement of the occipital horn of the lateral ventricle and compression of

* Correspondence: vikassurg2015@gmail.com

¹Department of Neurosurgery, Bombay Hospital, Marine lines, Mumbai, India
Full list of author information is available at the end of the article

Table 1 Previously published cases

S no	Authors/year	Age/sex	Location	Resection	Follow-up	RT	CT
1	Hall et al. 1986 [4]	6 months/F	Right frontal	Not mentioned	20 months	No	–
2	Lang et al. [4]	–	–	–	–	–	–
3	Hirose et al. 1992 [5]	12 years/F	Brain stem	Not mentioned	Not mentioned	Yes	Yes
5	Wolf et al. 1994 [6]	–	–	–	–	–	–
6		–	–	–	–	–	–
7	Prayson et al. 1995 [4]	–	–	–	–	–	–
8	Campos et al. 1994 [4]	10 years/M	Left temporal	–	6 months	–	–
9	Hakim et al. 1997 [4]	47 years/M	Frontal	Total	14 months	SRS	Yes
10	Nakajima et al. 1998 [4]	7 years/F	Right parieto occipital region	Subtotal	15 months	Yes	Yes
11	Araki et al. 1999 [4]	53 years/F	Frontal	Total	26 months	Yes	Yes
12	Danjoux et al. 2001 [4]	26 years/M	Temporal	–	2 months	Yes	–
13	Suzuki et al. 2002 [4]	59 years/M	Temporal	–	–	–	–
14	Nair et al. 2004 [4]	60 years/M	Right parietal	–	–	–	–
15	Lyken et al. 2004 [4]	–	–	–	11 years	Yes	Yes
16		–	–	–	–	Yes	Yes
17	Kang et al. 2007 [4]	45 years/M	Left frontal	Subtotal resection	35 months	Yes	Yes
18	Majores et al. 2008 [7]	45 years/M	Left temporo mesial	Total	11 months	Yes	Yes
19		M	–	–	–	Yes	Yes
20		M	–	–	–	Yes	Yes
21		M	–	–	–	Yes	Yes
22		F	–	–	–	Yes	Yes
23	Karremann et al. 2009 [4]	2 years/M	Left fronto basal	Total	31 months	No	No
24		3 years/M	Right frontal	Total	54 months	No	Yes
25		10 years/F	Right frontotemporoparietal	Total	42 months	Yes	Yes
26		10 years/M	Right parieto occipital	Partial	6 months	Yes	Yes
27		14 years/M	Pons and fourth ventricle	Subtotal	8 months	Yes	No
28		16 years/F	Right temporo mesial	Total	25 months	Yes	No
29	Kawataki et al. 2010 [4]	34 years/M	Left medial temporal	Subtotal	6 months	Yes	Yes
30	A.E. Rojas et al. 2013 [8]	22 years/M	Left frontotemporal lobes	–	11 months	Yes	–
31		24 years/F	Left frontotemporal lobes	–	3 months	Yes	–
32		25 years/M	Left frontotemporal lobes	–	6 months	Yes	–
33		34 years/M	Left frontal	–	9 months	Yes	–
34		9 years/M	Posterior fossa	–	6 months	Yes	–
35		9 years/M	B/L basal ganglia	–	2 months	Yes	Yes
36		37 years/M	Left frontotemporal region	–	2 months	Yes	–
37	Lucas et al. 2015 [9]	12 years/M	Left temporoparietal	Subtotal	23 months	Yes	Yes
38		11 years/M	Right temporal	Subtotal	22 months	Yes	Yes
39	Pikis et al. 2017 [10]	34 years/F	Left frontal	Subtotal	12 months	Yes	Yes
40	Lundar et al. 2018 [11]	15 years/M	Supratentorial	Total	14 years	–	–
41		19 years/M	Supratentorial	Total	12 months	–	–
42	Present study	34 years/F	Left parieto occipital	Total	26 months	Yes	Yes

the brain stem with midline shift and brain edema. These features were suggestive of hemangioblastoma/pilocytic astrocytoma (Fig. 1).

Left parieto occipital craniotomy was done which showed a cystic mass lesion with reddish brown solid nodule in the occipital region. Cyst was tapped followed

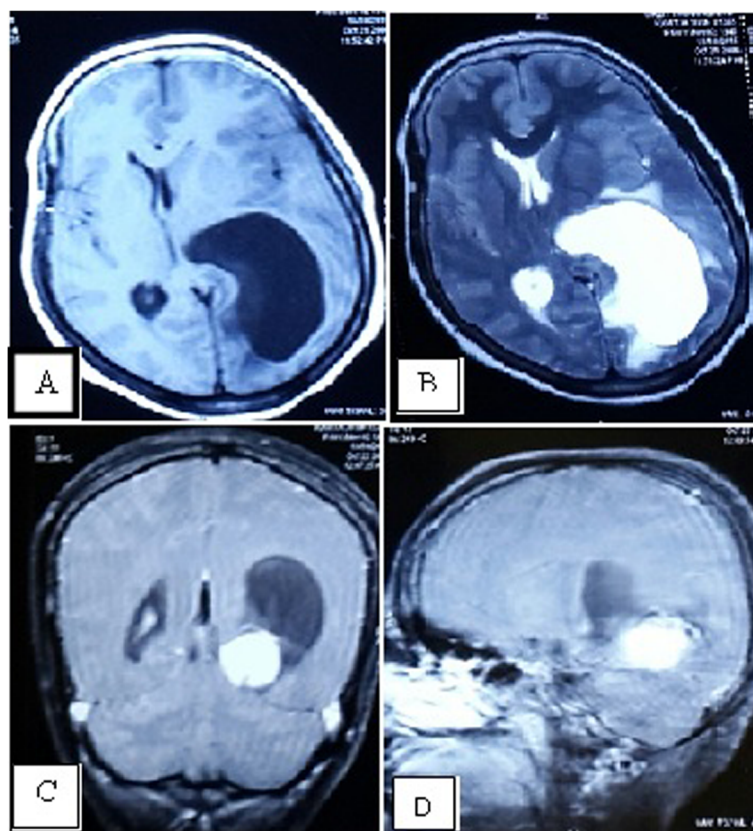


Fig. 1 CEMRI of the brain shows a well-defined solid cystic lesion in the left parieto occipital region. Solid component isointense and cystic component hypointense on T1 axial images (a). Solid component isointense and cystic component iso- to hyperintense on T2 axial images. Solid component shows intense enhancement on T1 contrast coronal and sagittal images (c, d), Solid component isointense and cystic component iso- to hyperintense on T2 axial images (b)

by nodule excision. The nodule was soft to firm in consistency and was mild to moderately vascular. Tumor was extending into the occipital horn and midline up to the falx; however, no attachment to the falx was seen gross total tumor excision was done.

Histopathology

Microscopic examination showed cellular glial tumor composed of malignant astrocytic cells and scattered ganglion cells. Astrocytic cells showed moderate nuclear pleomorphism with coarse chromatin with few mitosis. Immunohistochemistry (IHC) analysis showed focal positivity (ganglion cells) for CD56 and was negative for CD34, P53, IDH1, and synaptophysin. MIB1 index was 8% (Figs. 2 and 3). These features were suggestive of anaplastic ganglioglioma, WHO grade 3.

The immediate post-operative period was uneventful. The patient made gradual recovery over the and received 60-Gy radiotherapy with temozolamide as adjuvant therapy. Post-operative CEMRI of the brain showed craniotomy defect with gliosis in the left parieto occipital region with no contrast enhancement

(Fig. 4). CEMRI of the brain was done 14 and 24 months after surgery which showed post radiotherapy changes with gliosis with no recurrent lesion (Fig. 5). The patient is asymptomatic 26 months after surgery.

Discussion

Ganglioglioma is an infrequent tumor of the central nervous system, initially characterized by Perkins in 1926. The age of presentation of these tumors varies from 2 months to 70 years. The majority of gangliogliomas occur in the temporal lobe (>70%) [7]. The most common presenting signs and symptoms are seizures (temporal lobe and other supratentorial locations), followed by headache, dizziness, ataxia (posterior fossa), and progressive weakness (spinal cord). The typical of ganglioglioma is a benign, calcified tumor in the temporal lobe of a child with seizures [3]. On CT, the picture is of a circumscribed solid mass or cyst with a mural nodule. On MR imaging, gangliogliomas are isointense to hypointense on T1-weighted images, are hyperintense and heterogeneous on T2-weighted images, and can contain solid, cystic,

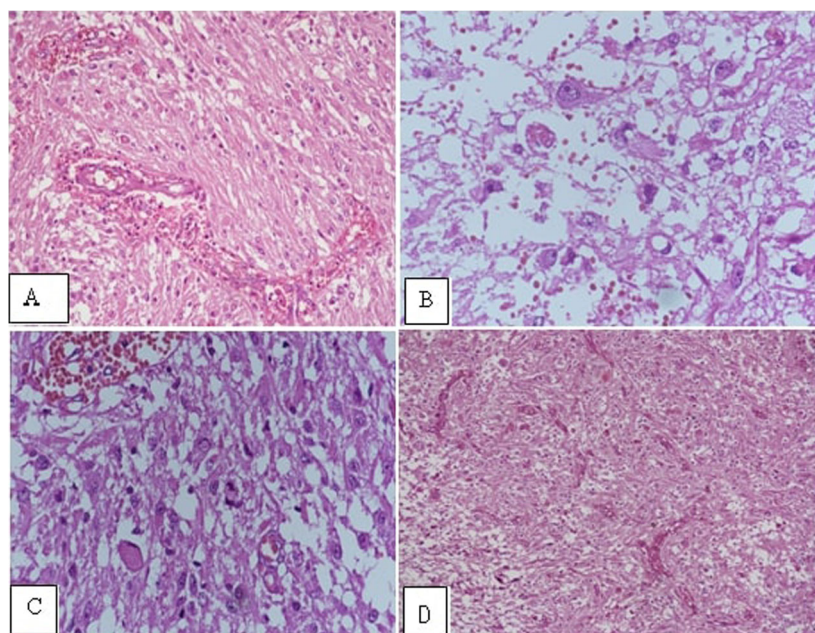


Fig. 2 Photomicrograph showing **a** branching capillaries and eosinophilic granular bodies seen in a fibrillary glial background. H&E, $\times 100$. **b** Ganglion cells with vesicular chromatin, prominent nucleoli, and moderate to abundant amount of eosinophilic cytoplasm. H&E, $\times 400$. **c** Photomicrograph showing a mitotic figure. H&E, $\times 100$. **d** Cellular glial tumor composed of malignant astrocytic cells with scattered ganglion cells and marked vascular proliferation. H&E, $\times 100$

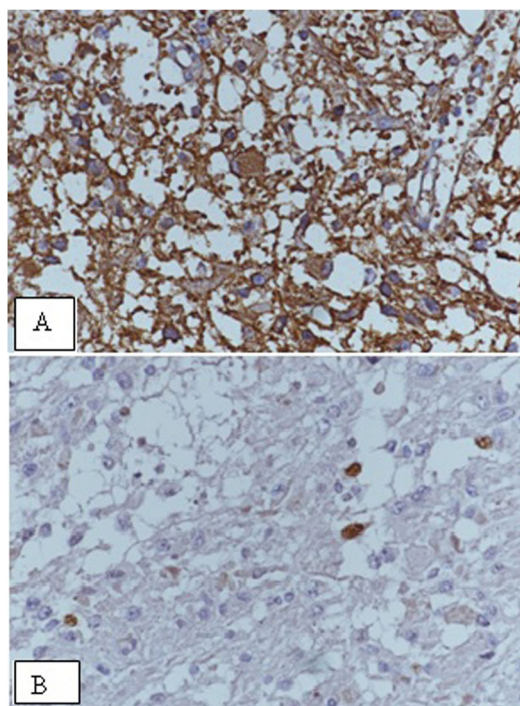


Fig. 3 HC **a** ganglion cells showing cytoplasmic positivity for CD 56. **b** Ki67/MIB1 positivity

and calcified components. Enhancement after administration of gadolinium has also been found to vary from no enhancement to marked, heterogeneous enhancement [2, 12].

Histopathologically, gangliogliomas are benign, well-differentiated neuroepithelial tumors. The typical feature is a combination of neuronal and glial cell elements, which may exhibit marked heterogeneity [13]. On IHC, the glial component is positive for GFAP, S-100 protein, and vimentin, whereas the neuronal component is reactive for synaptophysin, MAP 2, NeuN, and neurofilaments [14, 15]. Histopathologic differential diagnoses comprise both high-grade and low-grade neoplasms, such as diffuse astrocytomas, oligodendrogliomas, dysembryoplastic neuroepithelial tumors, pilocytic astrocytomas (PAs), and pleomorphic xanthoastrocytomas (PXAs) [7]. Mitotic figures in ganglioglioma are rare, and MIB index varies from 1.1 to 2.7%. CD 34 is present 70–80% in the neuronal component of ganglioglioma but is less common in anaplastic variants [15]. IDH1 and P53 should be done to rule out diffuse glioma.

Anaplastic gangliogliomas typically demonstrate malignant transformation of the glial component with hypercellularity, vascular proliferation, and necrosis high mitotic labeling indices (Ki-67) [16–18]. Anaplastic transformation is more common in the pediatric population and has been associated with previous

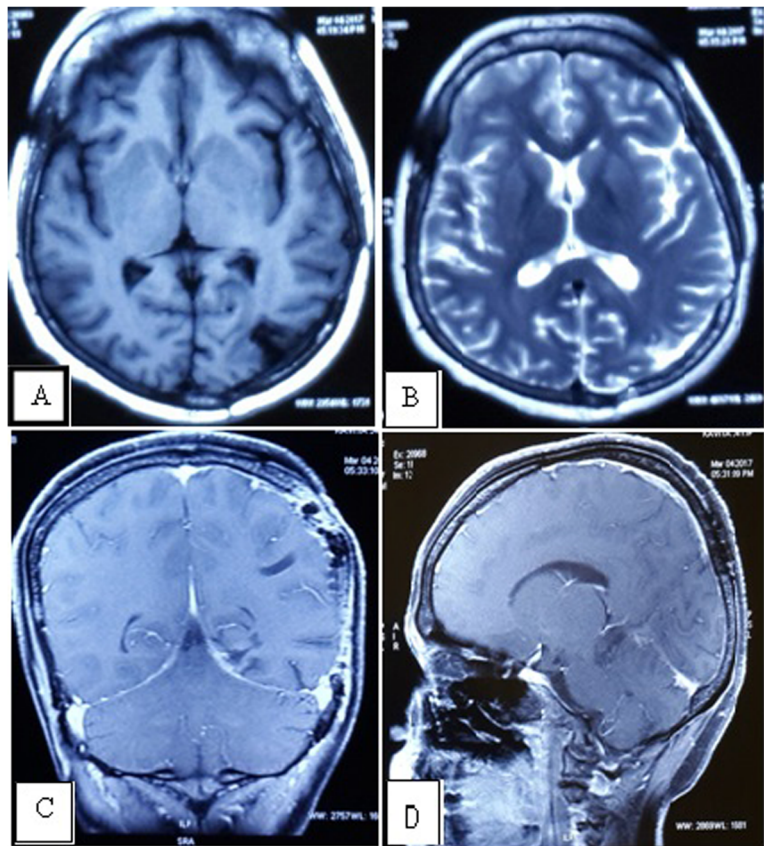


Fig. 4 Post-op CEMRI of the brain shows post-operative changes with no contrast enhancement. T1 and T2 axial (a, b) and T1 with contrast coronal and sagittal (c, d)

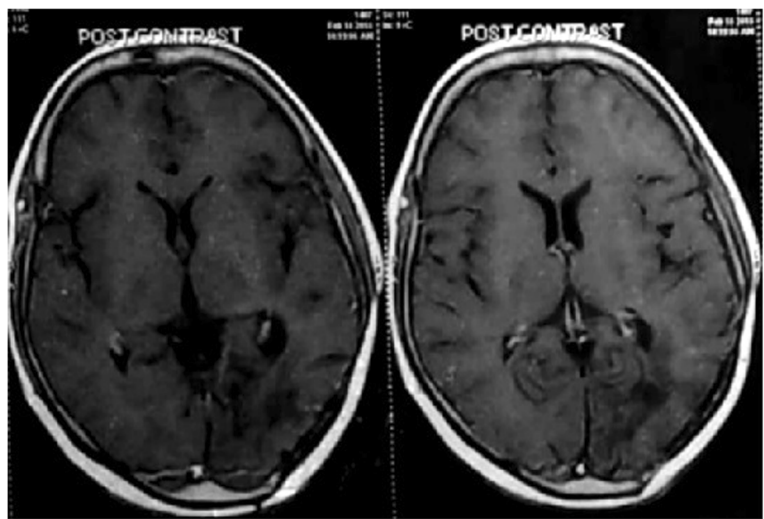


Fig. 5 CEMRI brain T1 contrast image done after 14 months of surgery showing post radiotherapy changes with no enhancement

subtotal tumor resection and radiotherapy [19]. Gross total resection should be attempted whenever possible with the preservation of neurological function. Some studies have advocated the role of gross total resection along with adjuvant radiochemotherapy in achieving good survival rates [20, 21].

Conclusion

Anaplastic gangliogliomas are rare tumors with parieto occipital as rare location. Imaging may vary, and histology along with IHC is needed for the diagnosis. Gross total resection along with adjuvant therapy improves outcome.

Abbreviations

CEMRI: Contrast-enhanced magnetic resonance imaging; ICP: Intracranial pressure; IHC: Immunohistochemistry; PAs: Pilocytic astrocytomas; PXAs: Pleomorphic xanthoastrocytomas

Acknowledgements

Not applicable.

Authors' contributions

VS and SB contributed to the data collection and writing of the manuscript. AH contributed to the histopathology and IHC analysis part. VS and SRH contributed to the cross-checking and editing of the manuscript. All authors read and approved the final manuscript.

Funding

There is no external funding for the publication of this article.

Availability of data and materials

Please contact Dr. Vikas Sharma for data requests. All identifiable data (name of patients, birthdate) have been removed out of the figure scans.

Ethics approval and consent to participate

As this is a case report, ethical clearance is not required.

Consent for publication

Consent for publication was taken from the patient prior to reporting of the case.

Competing interests

The authors declare that they have no competing interests.

Author details

¹Department of Neurosurgery, Bombay Hospital, Marine lines, Mumbai, India.

²Department of Neurosurgery, PGIMER & Dr RML Hospital, New Delhi, India.

³Department of Pathology, PGIMER & Dr RML Hospital, New Delhi, India.

Received: 7 May 2019 Accepted: 16 October 2019

Published online: 13 November 2019

References

1. Zhang D, Henning TD, Zou LG, Hu LB, Wen L, Feng XY, Dai SH, Wang WX, Sun QR, Zhang ZG. Intracranial ganglioglioma: clinicopathological and MRI findings in 16 patients. *Clin Radiol*. 2008;63(1):80–91.
2. Zentner J, Wolf HK, Ostertun B, Hufnagel A, Campos MG, Solymosi L, Schramm J. Gangliogliomas: clinical, radiological, and histopathological findings in 51 patients. *J Neurol Neurosurg Psychiatry*. 1994;57(12):1497–502.
3. Rumana CS, Valadka AB. Radiation therapy and malignant degeneration of benign supratentorial gangliogliomas. *Neurosurgery*. 1998;42:1038–43.
4. Demarchi R, Abu-Abed S, Munoz D, Loch Macdonald R. Malignant ganglioglioma: case report and review of literature. *J Neurooncol*. 2011;101:311–8.
5. Hirose T, Kannuki S, Nishida K, Matsumoto K, Sano T, Hizawa K. Anaplastic ganglioglioma of the brain stem demonstrating active neurosecretory features of neoplastic neuronal cells. *Acta Neuropathol*. 1992;83:365–70.
6. Wolf HK, Muller MB, Spanle M, Zentner J, Schramm J, Wiestler OD. Ganglioglioma: a detailed histopathological and immunohistochemical analysis of 61 cases. *Acta Neuropathol*. 1994;88:166–73.
7. Majores M, von Lehe M, Fassunke J, Schramm J, Becker AJ, Simon M. Tumor recurrence and malignant progression of gangliogliomas. *Cancer*. 2008;113:3355–63.
8. Romero-Rojas AE, Diaz-Perez JA, Chinchilla-Olaya SI, Amaro D, Lozano-Castillo A, Ligia I. Restrepo-Escobar histopathological and immunohistochemical profile in anaplastic gangliogliomas. *neurocirugia*. 2013;24:237–43.
9. Lucas JT Jr, Huang AJ, Mott RT, Lesser GJ, Tatter SB, Chan MD. Anaplastic ganglioglioma: a report of three cases and review of the literature. *J Neuro-oncol*. 2015;123:171–7.
10. Pikis S, Petrosyan T, Diamantopoulou K, Kelesis C. Anaplastic ganglioglioma becoming symptomatic in the third trimester of pregnancy. *Int J Reprod Contracept Obstet Gynecol*. 2017;6:1158–60.
11. Lunder T, Due-Tønnessen BJ, Fric R, Egge A, Krossnes B, Due-Tønnessen P, Stensvold E, Brandal P. *Acta Neurochir (Wien)*. 2018;160(6):1207–14.
12. Johannsson JH, Rekeate HL, Roessmann U. Gangliogliomas: pathological and clinical correlation. *J Neurosurg*. 1981;54:58–63.
13. Hirose T, Scheithauer BW, Lopes MB, et al. Ganglioglioma: an ultrastructural and immunohistochemical study. *Cancer*. 1997;79:989–1003.
14. Blumcke I, Giencke K, Wardelmann E, Beyenburg S, Kral T, Sarioglu N, Pietsch T, Wolf HK, Schramm J, Elger CE, Wiestler OD. The CD34 epitope is expressed in neoplastic and malformative lesions associated with chronic, focal epilepsies. *Acta Neuropathol (Berl)*. 1999;1997:481–90.
15. Blumcke I, Muller S, Buslei R, Riederer BM, Wiestler OD. Microtubule-associated protein-2 immunoreactivity: a useful tool in the differential diagnosis of low-grade neuroepithelial tumors. *Acta Neuropathol*. 2004;108:89–96.
16. Kawataki T, Sato E, Sato T, Kinouchi H. Anaplastic ganglioglioma with malignant features in both neuronal and glial components-case report. *Neurol Med Chir*. 2010;50:228–31.
17. Karabekir HS, Balci C, Tokyol C. Primary spinal anaplastic ganglioglioma. *Pediatr Neurosurg*. 2006;42:374–8.
18. Nakajima M, Kidooka M, Nakasu S. Anaplastic ganglioglioma with dissemination to the spinal cord: a case report. *Surg Neurol*. 1998;49:445–8.
19. Selvanathan SK, Hammouche S, Salminen HJ, Jenkinson MD. Outcome and prognostic features in anaplastic ganglioglioma: analysis of cases from the SEER database. *J Neurooncol*. 2001;105:539–45.
20. Karremann M, Pietsch T, Janssen G, Kramm CM, Wolff JE. Anaplastic ganglioglioma in children. *J Neurooncol*. 2009;92:157–63.
21. Terrier, L. M., Bauchet, L., Rigau, V., Amelot, A., Zouaoui, S., Filipiak, I., ... Club de Neuro-Oncologie of the Société Française de Neurochirurgie. Natural course and prognosis of anaplastic gangliogliomas: a multicenter retrospective study of 43 cases from the French Brain Tumor Database. *Neuro-Oncol*. 19(5): 678–688, 2016

Publisher's Note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.