



Unexpected Prolonged Survival in a Case of Cerebellar GBM: An Interesting Case with Literature Review

Maruti Nandan¹ Ashish Patnaik² Rabi Narayan Sahu² Priyadarshi Dikshit¹ Ved P. Maurya¹
Kuntal K. Das¹ Raj Kumar¹

¹ Department of Neurosurgery, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, Uttar Pradesh, India

² Department of Neurosurgery, All India Institute of Medical Sciences, Bhubaneswar, Orissa, India

Address for correspondence Ved P. Maurya, MCh, Department of Neurosurgery, Sanjay Gandhi Post Graduate Institute of Medical Sciences, First Floor, C-Block (Hospital Building), Raebareli Road, Lucknow, Uttar Pradesh 226014, India (e-mail: vpmsurgery@gmail.com).

Indian J Neurosurg

Abstract

Keywords

- ▶ glioblastoma
- ▶ cerebellum
- ▶ radiotherapy
- ▶ metastatic

Glioblastoma multiforme (GBM) of cerebellar hemisphere is a rare entity and constitutes less than 1% of all the GBMs. The rarity of occurrence leads to significant challenge in differentiating morphologically from other subtypes of glioma in the posterior fossa. Previous studies have suggested that cerebellar GBM occurs in the younger age group as compared with the supratentorial counterpart.

Here, we report a case of cerebellar GBM in a young adult and discuss the pathogenesis including radiological and pathological aspects involved in the treatment of cerebellar GBM.

Introduction

Cerebellar glioblastoma is a rare tumor accounting for less than 1% of all cases of GBM.¹ Due to its rarity in this unusual location, the pathogenesis and prognosis are not well understood. Glioblastoma multiforme (GBM) is the most common primary brain tumor in adults comprising ~50% of all primary intracranial tumors. They generally occur in the fifth and sixth decades of life.²⁻⁵

We report here a case of 36-year-old man who was diagnosed to have a recurrent left cerebellar GBM after 5 years of gross total excision and was followed by adjuvant radiotherapy. He was again subjected to surgery and gross total resection was achieved following which he remains symptom free.

Case Report

A 36-year-old adult man operated somewhere 5 years ago for posterior fossa GBM and received radiotherapy as well,

presented to the casualty with history of gradual-onset mild-to-moderate intermittent throbbing headache associated with multiple episodes of projectile vomiting for 1 month, and instability and difficulty in walking for 15 days with Glasgow coma score of 15 (→Fig. 1). A neurologic examination showed cerebellar signs, including cerebellar ataxia, dysmetria, and dysidiadochokinesia. The funduscopy revealed bilateral papilledema. A computed tomogram and magnetic resonance imaging (MRI) of the brain revealed the recurrence of posterior fossa solid cystic tumor with a diameter of 50 mm × 35 mm × 30 mm (→Fig. 2). Gross total excision of the tumor and duraplasty with G patch was done using the left retromastoid suboccipital approach. The tumor was reddish brown, firm, nonsuckable, moderately vascular, and was adherent to the tentorium cerebelli with cystic component having straw colored fluid. He developed wound infection and there was frank pus discharge, culture of which revealed Methicillin resistant staphylococcus aureus (MRSA) sensitive to colistin, and managed conservatively. Gradually,

DOI <https://doi.org/10.1055/s-0042-1744246>.
ISSN 2277-954X.

© 2022. Neurological Surgeons' Society of India. All rights reserved. This is an open access article published by Thieme under the terms of the Creative Commons Attribution-NonDerivative-NonCommercial-License, permitting copying and reproduction so long as the original work is given appropriate credit. Contents may not be used for commercial purposes, or adapted, remixed, transformed or built upon. (<https://creativecommons.org/licenses/by-nc-nd/4.0/>)

Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India

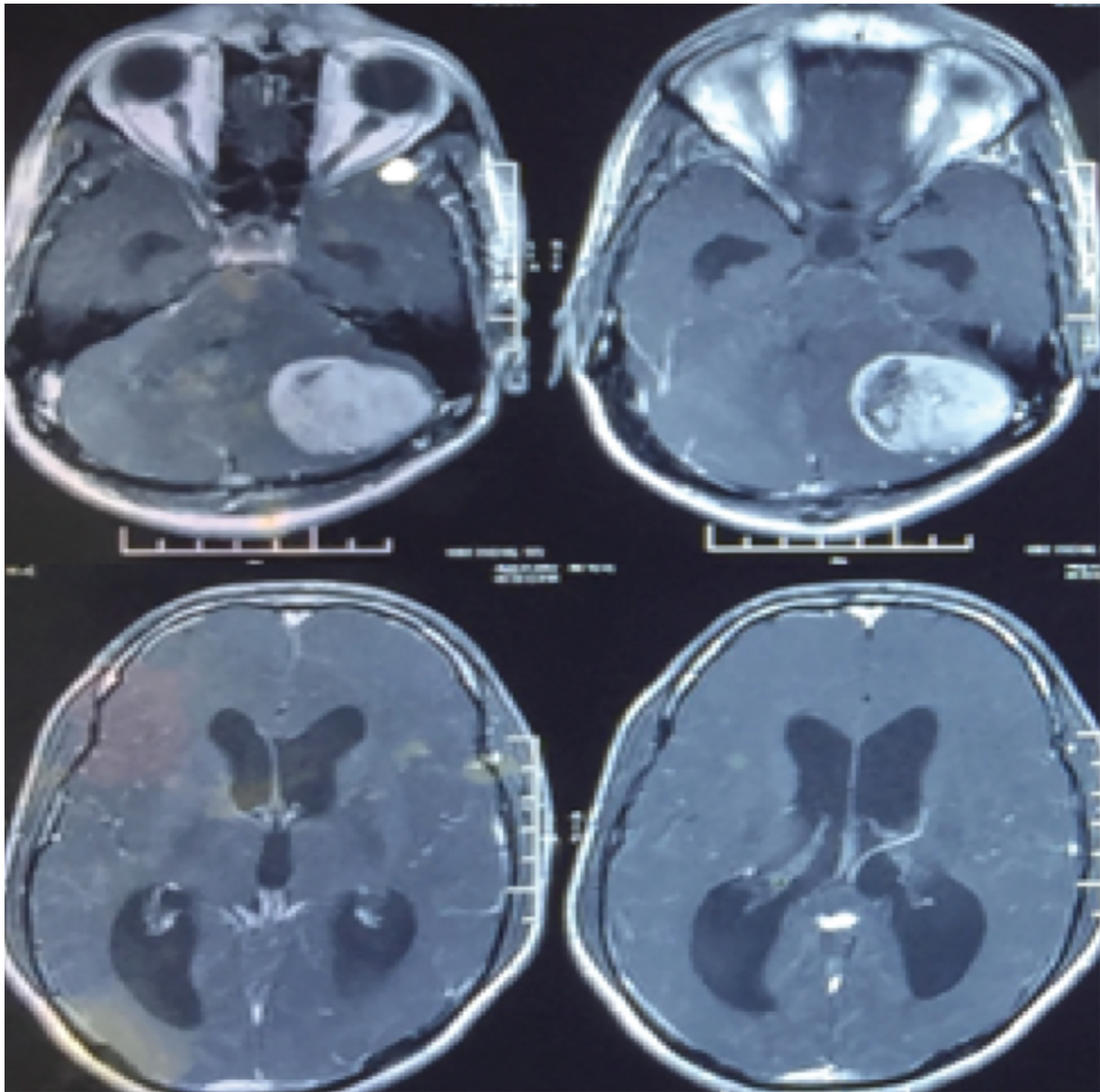


Fig. 1 Preoperative magnetic resonance imaging (MRI) scan shows homogenous contrast-enhancing space-occupying lesion in the left cerebellar hemisphere with effacement of fourth ventricle causing obstructive hydrocephalus.

the amount of pus discharge reduced but cerebrospinal fluid (CSF) leak was persistent. Also, serial scan revealed hydrocephalus. He was again operated and right parietal Medium pressure ventriculoperitoneal shunt (MPVP) shunt was done, wound was explored, and infected G patch was removed, and secondary suturing was done. Gradually, the patient recovered over a long period of time and there was delayed healing of the wound, might be due to radiation changes and scarring.

Histopathology was suggestive of marked pleomorphism with numerous tumor giant cells with extensive areas of coagulation necrosis with endothelial proliferation. Marked pleomorphism with a high mitotic index was noticed. Strong immunopositivity for Glial fibrillary acidic protein (GFAP) and p53 and negative for Isocitrate dehydrogenase (IDH) mutant-specific antibody. Alpha thalassemia mental

retardation X linked syndrome (ATRX) was retained in the tumor cells with impression of glioblastoma grade 4 IDH and ATRX wild-type.

Discussion

Glioblastoma multiforme (GBM) is the most common primary intracranial tumor in adults. Although it presents at all ages, a peak incidence is seen around the sixth decade and a second peak in the first decade.^{6,7} The male to female ratio is 2:1.⁶ The cerebellum acts as the primary site for GBM and accounts for only 0.24% to 1.00% of all the intracranial occurrences.⁸ Symptoms associated with cerebellar glioblastoma are very nonspecific such as headache, gait disturbance, vertigo, nausea, vomiting, and ataxia. And a few others such as, dizziness, neck pain, and mental confusion

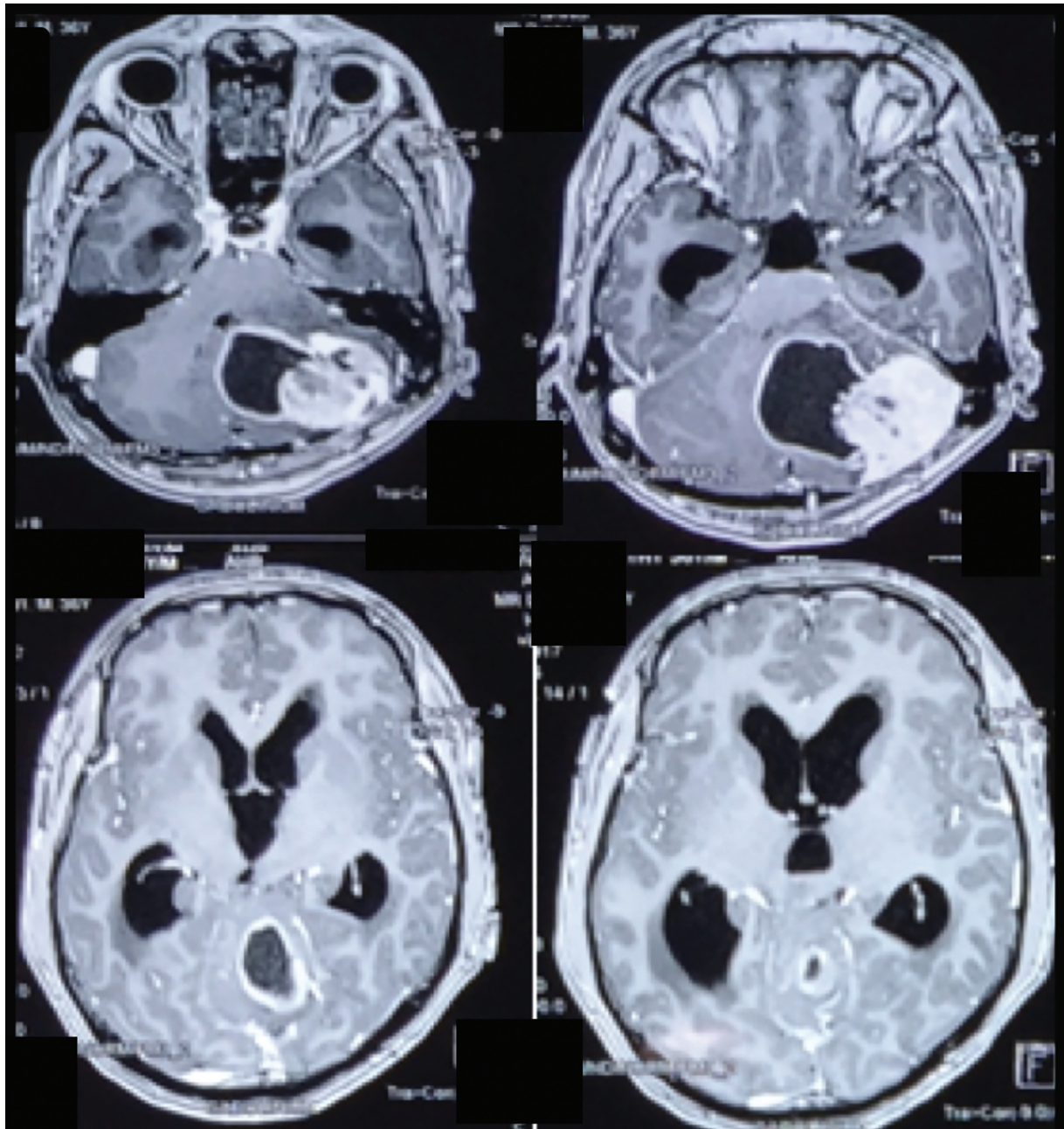


Fig. 2 MRI scan with FLAIR sequence suggests recurrent homogenous contrast-enhancing SOL having a solid and cystic component with effacement of the fourth ventricle causing obstructive hydrocephalus and infiltrating the tentorium cerebelli.

can be present.^{8,9} Our patient did have hydrocephalus as also seen in four out of nine patients in the study by Kuroiwa et al.⁸ The pathogenesis of cerebellar glioblastoma is still partially understood even after years of research confirming its rarity. Two subsets of cerebellar GBM have been reported by Grahovac et al.¹⁰ They can occur de novo as in the primary type or from previous low-grade astrocytoma as in the secondary type.¹¹ Primary glioblastomas affect mostly elderly patients and generally are characterized by the loss of heterozygosity 10 q (LOH 10q) (70%), epidermal growth factor receptor amplification (36%), and *TP53* mutation at a frequency of lower than 30%,¹¹ whereas secondary glioblastomas are seen in younger patients and often contain *TP53* mutation (65%).

The diagnosis of cerebellar GBM is always uncertain preoperatively. Computed tomography (CT) and MRI are helpful in asserting toward the diagnosis. Cerebellar metastases, abscess, and cerebellar infarct is similar with GBM on imaging; therefore, differential diagnosis from these diseases is difficult.^{5,8,12}

Metastatic tumor is the most common differential diagnosis in adults. Solid tumors with contrast enhancement or those with central hypodensity suggesting necrosis can be mistaken for cerebellar GBM. Occhiogrosso et al found little peritumoral edema in patients with cerebellar GBM, and Zito et al stated that CT was helpful in differentiating GBM from metastasis of the cerebellum via peritumoral edema or mass effect.^{6,13} For

Table 1 Review of cerebellar glioblastoma published in the literature

Author	No. of patients	Mean age	Location	Treatment	CT/RT	Median overall survival
Babu et al ¹⁵ (1975–2009)	247	59	Cerebellum	Resection(203) Biopsy (37) Unknown (7)	UK	7 months
Demir et al ⁹ (2005)	2	22.5	Cerebellum	GTR 1 STR 1	Yes	7 months
Weber et al ¹⁶ (2006)	45	50.3	Cerebellum	GTR (9) STR (29) Biopsy (5)	Yes	9.9 months
Jindal et al ¹⁷ (2006)	1	15	Cerebellum + CP angle	GTR	Yes	3 months
Hur et al ¹⁸ (2008)	1	69	Cerebellum	GTR	Yes	No recurrence up to 1 year
Gopalakrishnan et al ¹⁹ (2011)	5	50	Cerebellum	GTR	Yes	9 months
Hong et al ²⁰ (2018)	8	36 ± 26	Cerebellum	GTR 4 STR 4	Yes	5.5 months (7 patient) Still alive after 74 months (1 patient)
Kluska ²¹ (2020)	1	46	Cerebellum	Biopsy	Yes	Unknown
Present case (2021)	1	34	Cerebellum	GTR	Yes	72 months

good tissue resolution and omitting major bone artifacts in differentiating the lesions, MRI is a very precise diagnostic tool. Differences in vascularity and metabolite levels in the periphery of a tumor have been helpful in differentiation of metastases and GBMs. Metastasis may demonstrate cystic components or show hypointensity on T2-weighted images, suggesting intratumoral hemorrhage, calcification, or mucinous components as well as that is rare with GBM.

On CT, cerebellar GBM shows normal-appearing T2-weighted images were as those with abscess are well-defined, thin-walled enhancing rim on contrast-enhanced T1-weighted images.¹⁴ Data of cerebellar GBM worldwide are very scarce and so is true for India.^{9,15–21} Previously, one case report of cerebellar GBM with cerebellopontine angle extension reported by Jindal et al and a case series of five patients from different centers reported by Gopalakrishnan et al in 2011 from India. ► **Table 1** provides a review of the literature for the cerebellar GBM with an average survival period following surgical intervention. Adams et al analyzed the SEER (Surveillance, Epidemiology and End results) database from 1973 to 2009 and concluded that patients with cerebellar GBM had prolonged survival as compared with supratentorial GBM. Factors associated with prolonged survival in these patients are younger age at presentation, radiation therapy and “Asian or Pacific Islander” race.²² These findings were also observed in our case report as the age of first presentation was 31 years, received radiotherapy, and he belonged to the Asian island.

Conclusion

Cerebellar GBM is rare and should be considered in the differential diagnosis of a cerebellar mass lesion. MRI and

contrast enhancement are two most important tools for its characterization from its differentials. Although the median survival described in the literature is ~7 months, the overall survival in our case is 6 years after recurrence.

Funding

None.

Conflicts of Interests

None declared.

References

- Hernandez-Gonzalez G, Marchione P, De Angelis F, Giannone C, Kouleridou A, Spallone A. Long-term survival in cerebellar glioblastoma multiforme. Case report. *J Neurosurg Sci* 2012;56(04):379–381
- Aun RA, Stavale JN, Silva Junior D. Glioblastoma multiforme no cerebelo: registro de um caso. *Arquivos de Neuro-Psiquiatria* 1981;39(03):350–354
- Hegedüs K, Molnár P. Primary cerebellar glioblastoma multiforme with an unusually long survival. Case report. *J Neurosurg* 1983;58(04):589–592
- Kopelson G. Cerebellar glioblastoma. *Cancer* 1982;50(02):308–311
- Miller EM, Mani RL, Townsend JJ. Cerebellar glioblastoma multiforme in an adult. *Surg Neurol* 1976;5(06):341–343
- Occhiogrosso M, Spada A, Merlicco G, Vailati G, De Benedictis G. Malignant cerebellar astrocytoma. Report of five cases. *J Neurosurg Sci* 1985;29(01):43–50
- Rosenfeld J, Rossi ML, Briggs M. Glioblastoma multiforme of the cerebellum in an elderly man. A case report. *Tumori* 1989;75(06):626–629
- Kuroiwa T, Numaguchi Y, Rothman MI, et al. Posterior fossa glioblastoma multiforme: MR findings. *AJNR Am J Neuroradiol* 1995;16(03):583–589

- 9 Demir MK, Hakan T, Akinci O, Berkman Z. Primary cerebellar glioblastoma multiforme. *Diagn Interv Radiol* 2005;11(02):83–86
- 10 Grahovac G, Tomac D, Lambasa S, Zoric A, Habek M. Cerebellar glioblastomas: pathophysiology, clinical presentation and management. *Acta Neurochir (Wien)* 2009;151(06):653–657
- 11 Ohgaki H, Kleihues P. Genetic pathways to primary and secondary glioblastoma. *Am J Pathol* 2007;170(05):1445–1453
- 12 Tamaki T, Eguchi T, Sakamoto M, Teramoto A. Glioblastoma multiforme of the cerebellum in an elderly man. *J Chin Med Assoc* 2004;67(06):301–304
- 13 Zito JL, Siva A, Smith TW, Leeds M, Davidson R. Glioblastoma of the cerebellum. Computed tomographic and pathologic considerations. *Surg Neurol* 1983;19(04):373–378
- 14 Lai PH, Ho JT, Chen WL, et al. Brain abscess and necrotic brain tumor: discrimination with proton MR spectroscopy and diffusion-weighted imaging. *AJNR Am J Neuroradiol* 2002;23(08):1369–1377
- 15 Babu R, Sharma R, Karikari IO, Owens TR, Friedman AH, Adamson C. Outcome and prognostic factors in adult cerebellar glioblastoma. *J Clin Neurosci* 2013;20(08):1117–1121
- 16 Weber DC, Miller RC, Villà S, et al. Outcome and prognostic factors in cerebellar glioblastoma multiforme in adults: a retrospective study from the Rare Cancer Network. *Int J Radiat Oncol Biol Phys* 2006;66(01):179–186
- 17 Jindal A, Jaiswal AK, Jaiswal S, Pandey P, Mahapatra AK, Sharma MC. Cerebellar glioblastoma multiforme presenting as a cerebellopontine angle mass. *J Pediatr Neurosci* 2006;1(02):21–23
- 18 Hur H, Jung S, Jung TY, Kim IY. Cerebellar glioblastoma multiforme in an adult. *J Korean Neurosurg Soc* 2008;43(04):194–197
- 19 Gopalakrishnan CV, Dhakoji A, Nair S, Menon G, Neelima R. A retrospective study of primary cerebellar glioblastoma multiforme in adults. *J Clin Neurosci* 2012;19(12):1684–1688
- 20 Hong B, Banan R, Christians A, et al. Cerebellar glioblastoma: a clinical series with contemporary molecular analysis. *Acta Neurochir (Wien)* 2018;160(11):2237–2248
- 21 Kluska A, Tracz N, Loga K, Papis U, Anna, Fijuth J, et al. *Medycyna Paliatywna. Poznan* 2020;12(01):36–39
- 22 Adams H, Chaichana KL, Avendaño J, Liu B, Raza SM, Quiñones-Hinojosa A. Adult cerebellar glioblastoma: understanding survival and prognostic factors using a population-based database from 1973 to 2009. *World Neurosurg* 2013;80(06):e237–e243