International Surgery Journal
Syed AS et al. Int Surg J. 2024 Jun;11(6):1023-1026
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20241410

Case Report

Primary spinal glioblastoma multiforme: a case report

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Received: 22 April 2024
Accepted: 16 May 2024

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ABSTRACT

Spinal cord glioblastoma multiforme (GBM) is an uncommon disease within spinal tumours. According to our review of literature, less than 200 patients have been reported so far. Here we highlight the case of a 35 year female who complained of mild and intermittent lower back pain that radiates to bilateral thighs and was associated with tingling and numbness over both feet. The patient was evaluated and operated elsewhere with L1-L2 laminectomy and micro-surgical excision and it was reported as a case of myxopapillary ependymoma. However, immunohistochemical (IHC) stains were positive for GBM grade IV. A residual tumour was seen in postoperative magnetic resonance imaging (MRI) at our institute. Subsequently, the patient was subjected to adjuvant radiotherapy (RT) and regular Temozolomide chemotherapy following near-total resection of the mass. Aggressive management of this condition with timely chemo radiation is needed to enhance survival and assure a decent quality of life.

Keywords: Spinal cord, Glioblastoma, Temozolomide, RT, Surgery

INTRODUCTION

Spinal cord tumours in adults mostly occur at the extramedullary space.[1] Out of the intramedullary tumours, the most common is astrocytoma.[2] This is in contrast to that seen in children where ependymomas are much more prevalent.[2]

Spinal cord glioblastoma multiforme (GBM) is a unique tumour type which accounts for a small percentage of all tumours of the spinal cord.[3] Defined by world health organisation (WHO) as a grade IV astrocytoma, is a highly malignant central nervous system (CNS) tumour that is histologically, genetically, and clinically heterogeneous.[3]

According to our literature review, less than 200 cases of primary GBM have been reported. Based on these, the tumour is regarded more prevalent in young men and occurs more commonly in the cervical and thoracic area.[4] For establishing a diagnosis, MRI, IHC and HPE are indispensably required.[5] Clinical manifestations are dependent almost entirely upon the location and extent othethe tumour.[6]

Currently the most recommended management is similar to that of its intracranial counterpart, which consists of surgical excision along with adjuvant temozolamide and RT.[7] However, despite prompt intervention it has an aggressive course and an invariably dreadful prognosis such that the median survival is estimated at just ~11 months.[4]

CASE REPORT

A 35 year female, with no known comorbidities presented to our institute with a complaint of lower back pain for 1 month. The pain was insidious in onset, mild in

severity, intermittent in nature and radiated to bilateral thighs. It was also associated with tingling sensation and numbness over both feet (right>left). She also had a history of loss of bladder control. Around 2 months earlier, the patient was evaluated elsewhere and managed as a case of myxopapillary ependymoma, with laminectomy and micro-surgical excision revealing tumour at conus medullaris and surrounding the filum terminale.

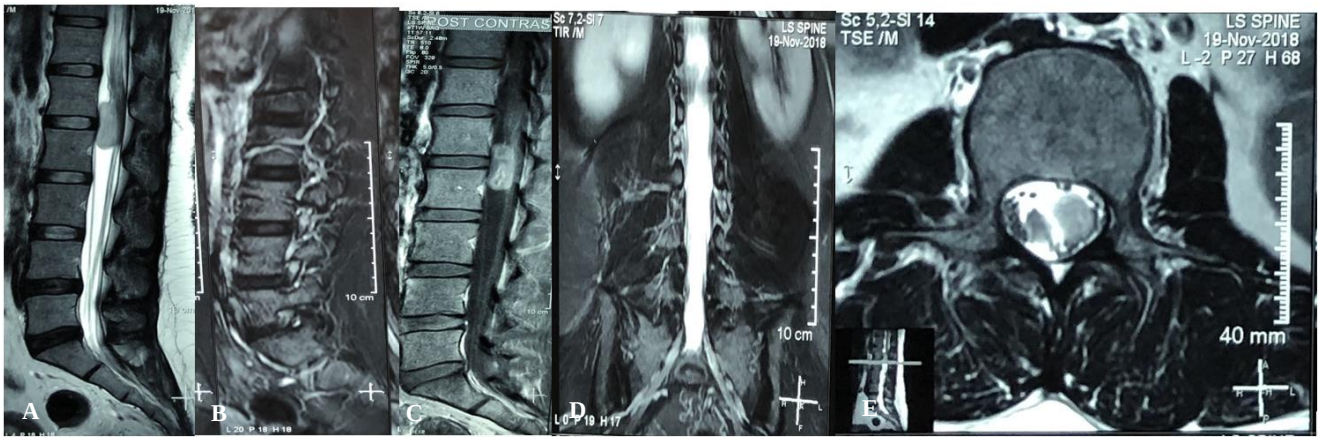

Figure 1 (A-E): MRI images before 1st surgery.

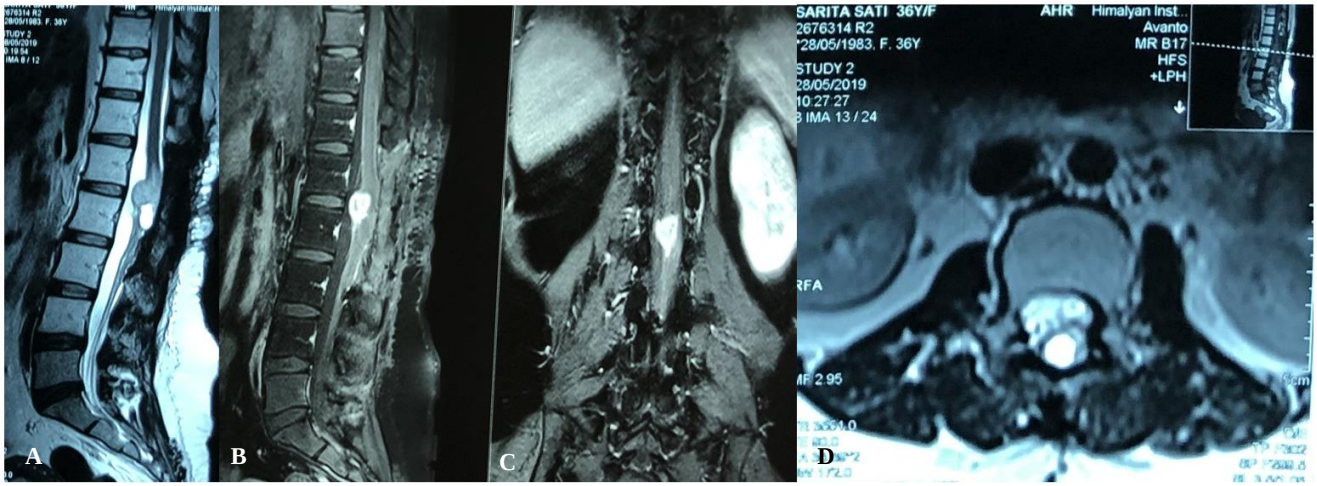

Figure 2 (A-D): MRI images at presentation.

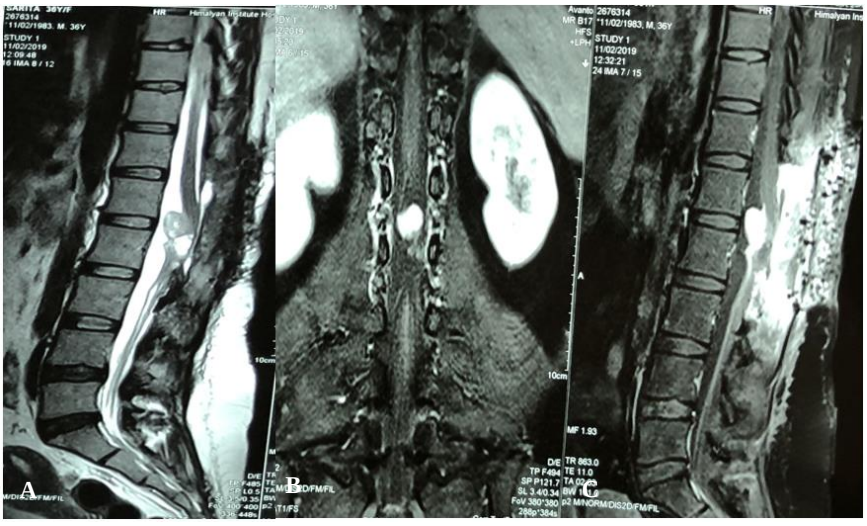

Figure 3 (A-C): MRI images after surgery.

At our institute, the patient was conscious, oriented and had a GCS score of 15. She had normal power across all joints in all 4 limbs. She had reduced sensory perception for all the modalities below L1 level. Her DTR was brisk in the lower limb. Upper limbs and cranial nerve examination was within normal limits. Routine blood investigations were unremarkable.

MRI lumbosacral spine with contrast revealed a well-defined rounded intradural hyperintense lesion at the level of L1-L2 vertebrae that was consistent with a residual tumour.

Given this, the patient underwent re-surgery under general anaesthesia. L1-L2 re-exploration and near total excision of the mass was performed. On opening dura, a fibrous band (probably tumorous) was seen entering into the subarachnoid space, which was continuous with the residual tumour. The Tumour was soft, partly fibrous, which was amenable to cavitron ultrasonic surgical aspirator (CUSA). It had a very poor plane with adjacent nerve roots and conus medullaris. Near-total resection could be achieved with bits of tumorous tissue left at the upper pole where it was indistinguishable from conus medullaris.

HPE and IHC were used and the diagnosis of GBM Grade IV was formulated in accordance with the WHO criteria. IHC showed positivity for ATRX, p53 and negativity for EMA, D240. Malignancy also exhibited an elevated MIB-1 labelling index ranging ~8-10%.

Postoperatively, Karnofsky performance status (KPS) score was 80%. Preoperative complaints showed improvement. An MRI (plain and enhanced) of the head disclosed no abnormalities. Lumbar puncture was performed, which was again normal. The cerebral spinal fluid (CSF) showed no malignant cells. Subsequently, patient received conformal external beam RT to post-operative site on 6 MV LINAC for 50.4 Gy in 28 fractions with concurrent temozolamide followed by 6 cycles of adjuvant temozolamide.

MRI lumbosacral spine showed a heterogeneously enhancing intradural lesion at L1-L2. The patient came for follow-up 12 months after surgery, with headache secondary to increased intracranial pressure (ICP). Computed tomography (CT) of head showed blunting of sulcal subarachnoid spaces and brain surface. Her fundus examination revealed gross papilledema and diagnosis of benign intracranial hypertension was made. In view of this, she underwent right ventriculo-peritoneal shunting. Despite ventriculo-peritoneal shunting patient had only transient improvement for a few days and the patient had progressive worsening of vision and drop in sensorium.

Given the disease progression, further treatment was not deemed appropriate and the patient succumbed to the illness a few months later.

DISCUSSION

Spinal cord GBM is a unique tumour type which accounts for a small percentage of all tumours of the spinal cord.[3] Less than ~5.5% of these originate in the conus medullaris making our case even more unique.[8] However, some authors claim that the tumour location has no impact on the overall survival rate.[9] At the time of diagnosis, the majority of spinal GBM cases exhibit severe disability.[10] Consequently, an early diagnosis becomes essential for increasing the survival rate.[17]

The gold standard for establishing a spinal GBM diagnosis is MRI.[11] Most lesions arise as expansile and infiltrative masses with high T2 signal and demonstrate heterogeneous enhancement on post-contrast T1-weighted sequences.[12] Nevertheless, distinguishing spinal GBM from other spinal tumours is challenging. For tumours with vague MRI traits, F-18 fluoride deoxyglucose positron emission tomography (FDG PET) can be done.[4] A newer modality that may assist in the diagnosis is diffusion tensor imaging and perfusion which shows decreased fractional anisotropy and increased relative cerebral blood volume for its cranial counterpart.[13]

Surgical intervention continues to deliver a definitive diagnosis via HPE, and at the same time is also therapeutic.[5] The histological characteristics of this tumour are comparable to cerebral GBM, illustrating marked hypercellularity, nuclear atypia, microvascular proliferation and necrosis.[14]

IHC staining of most tumours shows a prominent expression of the p53 gene, positivity for GFAP and negativity for EMA.[15] Some tumours also demonstrate overexpression of EGFR protein.[15]

Pathology

WHO grading of astrocytoma[19-] Grade I: Pilocytic Astrocytoma, grade II: Low-grade Diffuse Astrocytomas, grade III: Anaplastic Astrocytomas and grade IV: Glioblastoma

Treatment

The conventional treatment for spinal GBM includes maximal resection, followed by RT given concomitantly with temozolamide followed by adjuvant temozolamide.[7] Gross-total resection (no residual enhancement) is preferred, but near-total resection (thin rim of enhancement) is reasonable when margin between tumour and normal tissue is not precise.[17] The most radical surgical technique is cordectomy which is reserved for lumbar or sacral tumours.[18]

Lately, targeted drug therapy is also being used to gain maximum survival benefit. Bevacizumab (BEV) a humanised monoclonal antibody against vascular

endothelial growth factor A (VEGF-A) has potential benefits.[19] Some authors have proposed an intrathecal regime of interferon-β via an ommaya reservoir.[13]

Prognosis is determined by CSF involvement, intracranial seeding and the use of adjuvant RT and temozolamide.[20] Thus, regular follow up is advocated.

CONCLUSION

Spinal cord GBM is an extremely unique tumour with a dismal prognosis. Aggressive management is required which includes maximal resection, followed by RT given concomitantly with temozolamide followed by adjuvant temozolamide. Regular follow up of CSF cytology and spinal MRI is advised. Further, a central registry and treatment trials are required to interpret this disease and improve survival.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Mori K, Imai S, Shimizu J, Taga T, Ishida M, Matsusue Y. Spinal glioblastoma multiforme of the conus medullaris with holocordal and intracranial spread in a child: a case report and review of the literature. Spine J. 2022;12(1):e1-6.
2. Balériaux DL. Spinal cord tumors. Eur Radiol. 1999;9(7):1252-8.
3. Cacchione A, Mastronuzzi A, Cefalo MG, Colafati GS, Diomedi-Camassei F, Rizzi M, et al. Pediatric spinal glioblastoma of the conus medullaris: a case report of long survival. Chin J Cancer. 2016;35(1):44.
4. Shen C-X, Wu J-F, Zhao W, Cai Z-W, Cai R-Z, Chen C-M. Primary spinal glioblastoma multiforme: A case report and review of the literature. Medicine (Baltimore). 2017;96(16):e6634.
5. Koeller KK, Rosenblum RS, Morrison AL. Neoplasms of the spinal cord and filum terminale: radiologic-pathologic correlation. Radiographics. 2000;20(6):1721-49.
6. Farzin M, Hajiabadi M, Rahmani M, Kolahdouzan K. Mixed malignant glioblastoma and schwannoma in spinal cord with metachronous ependymoma: A case report. Clin Case Rep. 2021;9(6):e04162.
7. Kotecha R, Mehta MP, Chang EL, Brown PD, Suh JH, Lo SS, et al. Updates in the management of intradural spinal cord tumors: a radiation oncology focus. Neuro Oncol. 2019;21(6):707-18.
8. Ononiwu C, Mehta V, Bettegowda C, Jallo G. Pediatric spinal glioblastoma multiforme: current treatment strategies and possible predictors of survival. Childs Nerv Syst. 2012;28(5):715-20.
9. Konar SK, Maiti TK, Bir SC, Kalakoti P, Bollam P, Nanda A. Predictive factors determining the overall outcome of primary spinal glioblastoma multiforme: An integrative survival analysis. World Neurosurg. 2016;86:341-8.e1-3.
10. Behmanesh B, Setzer M, Konczalla J, Harter P, Quick-Weller J, Imoehl L, et al. Management of patients with primary intramedullary spinal cord glioblastoma. World Neurosurg. 2017;98:198-202.
11. Bonde V, Balasubramaniam S, Goel A. Glioblastoma multiforme of the conus medullaris with holocordal spread. J Clin Neurosci. 2008;15(5):601-3.
12. Kim WH, Yoon SH, Kim C-Y, Kim K-J, Lee MM, Choe G, et al. Temozolomide for malignant primary spinal cord glioma: an experience of six cases and a literature review. J Neurooncol. 2011;101(2):247-54.
13. Jagtap V, Shashank B. Spinal glioblastoma multiforme with brain and spinal seeding: treatment approach from various view points | Int J Curr Res. 20179(5):50025-8.
14. Stupp R, Mason WP, van den Bent MJ, Weller M, Fisher B, Taphoorn MJB, et al. Radiotherapy plus concomitant and adjuvant temozolomide for glioblastoma. N Engl J Med. 2005;352(10):987-96.
15. Chaurasia A, Park S-H, Seo J-W, Park C-K. Immunohistochemical analysis of ATRX, IDH1 and p53 in glioblastoma and their correlations with patient survival. J Korean Med Sci. 2016;31(8):1208-14.
16. Louis DN, Perry A, Reifenberger G, von Deimling A, Figarella-Branger D, Cavenee WK, et al. The 2016 world health organization classification of tumors of the central nervous system: A summary. Acta Neuropathol. 2016;131(6):803-20.
17. Young RM, Jamshidi A, Davis G, Sherman JH. Current trends in the surgical management and treatment of adult glioblastoma. Ann Transl Med. 2015;3(9):121.
18. Mayer RR, Warmouth GM, Troxell M, Adesina AM, Kass JS. Glioblastoma multiforme of the conus medullaris in a 28-year-old female: a case report and review of the literature. Clin Neurol Neurosurg. 2012;114(3):275-7.
19. Kaley TJ, Mondesire-Crump I, Gavrilovic IT. Temozolomide or bevacizumab for spinal cord high-grade gliomas. J Neurooncol. 2012;109(2):385-9.
20. Strik HM, Effenberger O, Schäfer O, Risch U, Wickboldt J, Meyermann R. J Neurooncol. 2000;50(3):239-43.

Cite this article as: Syed AS, Sehgal S, Pandey SK, Tiwari BK, Kumar R, Gupta M. Primary spinal glioblastoma multiforme: a case report. Int Surg J 2024;11:1023-6.