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SURGICAL NEUROLOGY INTERNATIONAL
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Editor: Nancy E. Epstein, MD Winthrop Hospital, Mineola, NY, USA

Case Report

Rare case of conus medullaris glioblastoma multiforme in a teenager

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Received: 14 January 17 Accepted: 28 July 17 Published: 26 September 17

Abstract

Background: Primary intramedullary spinal glioblastoma multiforme (GBM) lesions are very rare comprising only 1.5% of all spinal cord tumors. These lesions typically result in rapid neurological deterioration and are associated with a very poor prognosis.

Case Description: A 16-year-old male exhibited a slowly progressive paraparesis with urinary incontinence, ultimately resulting in paraplegia. On magnetic resonance (MR), he was diagnosed with an intramedullary GBM of the spinal cord extending from the T9 level through the conus medullaris. Ten months following decompression/partial surgical resection of the intramedullary mass, he remained paraplegic.

Conclusion: GBM of the spinal cord are rare in the pediatric age group. A 16-year-old male presented with a flaccid paraplegia attributed to an MR-documented GBM of the distal thoracic cord extending from the T9 level through the conus medullaris. Despite partial intramedullary tumor resection, 10 months postoperatively, he remained paraplegic.

Key Words: Conus medullaris, glioblastoma multiforme, intramedullary tumor, poor prognosis, radiation therapy, review of GBM, spinal cord

Access this article online
Website: www.surgicalneurologyint.com
DOI: 10.4103/sni.sni_21_17
Quick Response Code:

INTRODUCTION

Glioblastoma multiforme (GBM) rarely involves the spinal cord (1–5% of all GBM cases), and accounts for only 1.5% of all spinal cord tumors.[8,9] Spinal GBM are highly malignant neoplasms that typically produce devastating neurological deficits. Nevertheless, histologically and genetically pediatric GBM differ from adult intramedullary spinal GBM tumors, requiring different treatment protocols.

At present, younger patients are optimally treated with maximal tumor removal to minimize neurological morbidity. Here, we present the case of a 16-year-old male with flaccid paraplegia attributed to magnetic resonance (MR) documented intramedullary T9-conus likely GBM. Despite partial tumor resection followed by aggressive adjunctive treatment. he remained paraplegic 10 months later.

CASE REPORT

Clinical presentation

A 16-year-old male presented with a 3-week history of low back pain and a rapidly progressive sensorimotor flaccid paraplegia with urinary incontinence. Motor examination showed 1/5 strength in all distributions in the lower extremities, with a relative loss of pin

How to cite this article: Fayçal L, Mouna B, Najia EA. Rare case of conus medullaris glioblastoma multiforme in a teenager. Surg Neurol Int 2017;8:234.
http://surgicalneurologyint.com/Rare-case-of-conus-medullaris-glioblastoma-multiforme-in-a-teenager/

appreciation below the Th10-Th11 levels. The thoracic MR showed an intramedullary, fusiform, expansile lesion extending from T9-conus. It was iso- to hypointense on T1- and hyperintense on T2-weighted MR images and heterogeneously enhanced with gadolinium [Figure 1a-c]. Differential diagnostic considerations included GBM, astrocytoma, ependymoma, demyelinating disease, transverse myelitis, or a dural fistula. The surgery included a T9–L1 laminectomy for maximal cord decompression; a midline myelotomy allowed partial intramedullary tumor resection. The lesion was poorly demarcated, infiltrative, and contained necrotic areas with thrombosed vessels.

Histopathology

Histopathologically, the lesion was a GBM (WHO grade IV). It was characterized by hyperchromatism, pleomorphism (hematoxylin and eosin stains), atypical cells with high cellularity, vascular proliferation, and necrosis. Immunohistochemistry revealed glial fibrillary acidic protein (GFAP) and S100 protein, along with a high MIB1/Ki-67 labelling index.

Outcome

Despite surgery, conventional radiation therapy (40–50 Gy in 20–25 fractions), chemotherapy [temozolomide (TMZ)], and corticosteroids, the patient remained paraplegic at 10 postoperative months. Of interest, there has been no further tumor progression.

DISCUSSION

Clinical, diagnostic, and pathological presentation of pediatric glioblastoma multiforme

GBM involves the spinal cord in only 1–5% of all GBM cases, accounting for only 1.5% of all spinal cord tumors.[9] They rarely occur in patients in their second or third decades of life, and are typically found in the cervical followed by the thoracic cord; they only rarely involve the conus.[1,5] On MR studies, they must be differentiated from demyelinating disease, neurosarcoidosis, vascular malformations, ischemia, pseudotumor, chronic arachnoiditis, transverse myelitis, and other lesions. Pathologically, GBM typically demonstrate packed tumor cells with microvascular proliferation. GFAP staining is usually positive as is staining with M1B1/Ki67 (g.g. high proliferative index 30% of nuclei reactive).

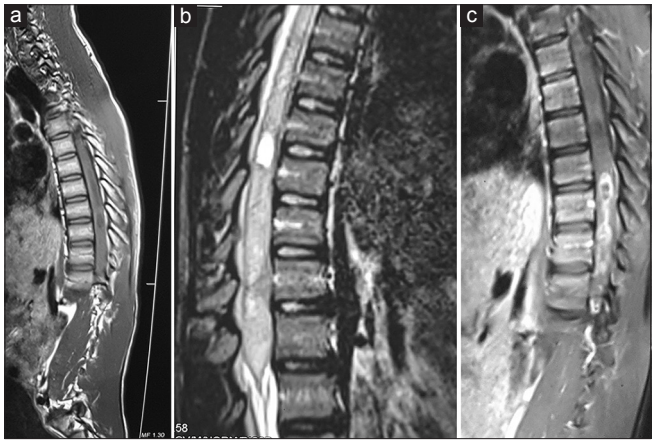

Figure 1: MRI findings: sagittal T1-weighted (a) and T2-weighted (b) and T1 post contrast (c) weighted sagittal images of a dorsal intramedullary GBM from Th9-L1. Note then large conus medullaris in a prominently widened spinal canal. Diffuse, inhomogeneous tumor enhancement and reactive cord edema were seen on the enhanced MR (c)

Treatment, surgery, and chemotherapy/radiation therapy

Although surgical decompression/excision of intramedullary GBM is the treatment of choice, it is often accompanied by significantly increased morbidity and does not necessarily increase survival; therefore, some recommend biopsy alone. Although radiotherapy and adjuvant TMZ are widely used, effective results have not been documented in children whose average survival barely exceeds 6 to 16 months, with a mean survival of 12 months.[2-4,6,7,10]

CONCLUSION

Intramedullary GBM of the spinal cord (T9-conus) are extremely rare in the pediatric age group. Despite aggressive intramedullary tumor resection, followed by adjuvant chemotherapy and radiation therapy, the patient's ultimate long-term outcomes are minimally impacted (e.g. estimated survival at 6–16 month; mean survival of 12 months).

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Bonde V, Balasubramaniam S, Goel A. Glioblastoma multiforme of the conus medullaris with holocordal spread. J Clin Neurosci 2008;15:6013.
2. Chiagozie O, Vivek M, Chetan B, George J. Pediatric spinal glioblastoma multiforme: Current treatment strategies and possible predictors of survival. Childs Nerv Syst 2012;28:715-20.
3. Cohen AR, Wisoff JH, Allen JC, Epstein F. Malignant astrocytomas of the spinal cord. J Neurosurg 1989;70:50-4.
4. Cohen KJ, Pollack IF, Zhou T. Temozolomide in the treatment of high-grade gliomas in children: A report from the Children's Oncology Group. Neuro Oncol 2011;13:317-23.
5. Morais N, Mascarenhas L, Soares-Fernandes JP, Silva A. Primary spinal glioblastoma: A case report and review of the literature. Oncol Lett 2013;5:992-6.
6. 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 2012;12:e1-6.
7. Perkins SM, Rubin JB, Leonard JR, Smyth MD, El Naqa I, Michalski JM, *et al*. Glioblastoma in children: A single-institution experience. Int J Radiat Oncol Biol Phys 2011;80:1117-21.
8. Scarrow AM, Rajendran P, Welch WC. Glioblastoma multiforme of the

conus medullaris. Clin Neurol Neurosurg 2000;102:166-7.
9. Singh PK, Singh VK, Tomar J, Azam A, Gupta S, Kumar S. Spinal glioblastoma multiforme: Unusual cause of post-traumatic tetraparesis. J Spinal Cord Med 2009;32:583-6.
10. Stupp R, Hegi ME, Mason WP. Effects of radiotherapy with concomitant and adjuvant temozolomide versus radiotherapy alone on survival in glioblastoma in a randomised phase III study: 5-year analysis of the EORTC-NCIC trial. Lancet Oncol 2009;10:459-66.