Case Report

**Rare case of conus medullaris glioblastoma multiforme in a teenager**

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**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.1,2 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
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, neurosarcoïdosis, 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 MIB1/Ki-67 (g.g. high proliferative index 30% of nuclei reactive).

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).

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Conflicts of interest
There are no conflicts of interest.

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