

## An Extremely Rare Case of Glioblastoma Multiforme of the Spinal Cord

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**Key Words:** glioblastoma multiforme; spinal cord; intramedullary tumor; spinal magnetic resonance imaging.

**Summary.** Being the most common glial cell tumor of the adult brain, primary glioblastoma multiforme is an extremely rare but excessively devastating condition of the spinal cord. It presents with indistinctive magnetic resonance imaging findings, so the diagnosis is very complicated to make. A low-grade glioma may undergo a malignant transformation into glioblastoma multiforme in a very short period, critically impairing treatment possibilities and prognosis, so a correct and timely diagnosis is crucial. We report a case of intramedullary glioblastoma multiforme in a young man and describe the diagnostic difficulties and devastating progression of the entity.

### Introduction

Glioblastoma multiforme (GBM) of the spinal cord is an exceptionally rare but remarkably aggressive tumor of the spinal cord, comprising only about 2%–4% of all glial tumors of the central nervous system (CNS) (1). For the management of this destructive entity, it is very important to make the right and timely diagnosis with a proper radiological and clinical follow-up. The diagnosis is very complicated to make because intramedullary GBM, being extremely rare, presents with unspecific magnetic resonance imaging (MRI) findings. There are no pathognomonic clinical or radiological signs to help making a prompt and exact diagnosis without histological proof. GBM may originate de novo (primary) or following a transformation from a low-grade glioma (secondary) (2), so it is very important to suspect a spinal cord tumor in the context of other spinal cord pathologies as early as possible for a timely diagnosis to be made and the most effective treatment to be applied.

### Case Report

A 34-year-old man presented with progressing sensorimotor paraparesis and twitching of the left foot. Initially, a lumbar hernia was suspected, but computed tomography (CT) of the spinal cord examination revealed no pathological changes. No pathological changes were found during spinal angiography, CT of the abdomen and the pelvis, chest x-ray examinations, as well as MRI examination of the head.

The first MRI of the spinal cord (Fig. 1) revealed the widened conus medullaris with no postcontrast enhancement, just slightly pronounced surrounding veins. The main entities, included in the differential diagnosis, were myelitis, a dural fistula, and a neoplastic process (astrocytoma).

The control MRI of the spinal cord (Fig. 2), performed 1.5 months later, revealed expanded widening of the spinal cord at the level of Th9–L1, involving the conus medullaris, with a slightly T2W hyperintense focus in it. No pathological postcontrast enhancement was observed. Due to a presumed diagnosis of myelitis, the patient was treated with glucocorticoids. The patient's symptoms were stable while receiving prednisolone, but after withdrawal, his neurological status got significantly worse: weakness in the legs increased and the level of dysesthesias and pain in the back aggravated.

The second control MRI of the spinal cord (Fig. 3) was performed 4.5 months later. In comparison with the previous MRI, the widening of the spinal cord was enlarged, and delicate postcontrast enhancement at the Th6–Th7 level was observed. No pathological changes were found during the control MRI examination of the head and the neck. Inflammatory changes remained the first choice of diagnosis, and treatment – empiric with ceftriaxone and symptomatic with glucocorticoids – was prescribed. There was no improvement in the clinical symptoms after the treatment, i.e., the neurological status kept deteriorating: the pain in the back exacerbated and significant paraparesis and dysesthesia of the lower limbs, hypesthesia at the Th9–Th10 level, and partial pelvic organ dysfunction were observed.

The intramedullary glial tumor was suspected, and the biopsy was performed. A histological analysis of the biopsy sample confirmed GBM (WHO grade IV). The patient received aggressive treatment with chemotherapy and radiotherapy as well as corticosteroids.

The control MRI of the spine, performed after the third chemotherapy course (Fig. 4), revealed the amplification of changes due to the progression of the tumor as well as extensive postradiation and postbiopsy alterations of the spinal cord and the vertebrae.

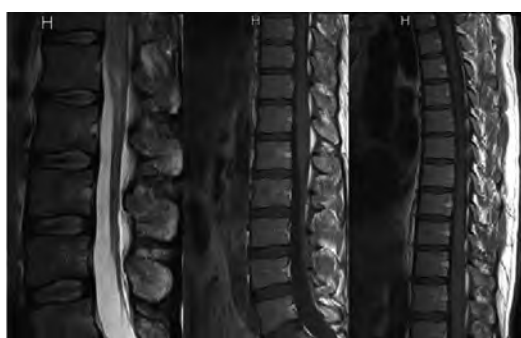
Two and a half months after the third chemo-

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*Fig. 1.* MRI findings: sagittal postcontrast T1-weighted images, T2- and T1-weighted images, and axial T1-weighted pre- and postcontrast images of the thoracolumbar part of the spinal cord

Enlarged conus medullaris; no postcontrast enhancement; slightly pronounced surrounding veins.



*Fig. 2.* MRI findings: sagittal T2-weighted image and post-contrast T1-weighted images of the thoracolumbar part of the spinal cord

Widening of the spinal cord at the Th9-L1 level, involving conus medullaris, with a slightly hyperintense focus on T2W in it. No pathological postcontrast enhancement.



*Fig. 3.* MRI findings: sagittal T2-weighted images, pre- and postcontrast T1-weighted images, and T2-weighted axial images of the thoracolumbar parts of the spinal cord

Enlarged width of the spinal cord at the Th10-L1 segment. A small T2 hyperintense focus at the Th5-L1 level; a delicate postcontrast enhancement at the Th6-Th7 level.



*Fig. 4.* MRI findings: sagittal T2-weighted and T1-weighted postcontrast images, as well as T1 and T2 pre- and T1 postcontrast weighted axial images of the thoracolumbar part of the spinal cord

Prominently widened spinal canal, nonhomogeneous postcontrast enhancement at the Th5-L1 level, reactive edematous spinal cord changes at the Th4-L1 level, and multiple degenerative cysts. Extradural cystic formation at the Th10-Th11 level and postbiopsy-radiation changes in the Th10 and Th11 vertebrae.

therapy course, the patient's symptoms were stable: paraplegia, complete pelvic organ dysfunction, and pain in the back remained, but no other new symptoms emerged. The spinal MRI examination, repeated at that time (Fig. 5), did not show major dynamics compared with the results of the previous MRI examination.

During the next 5 months, despite the aggres-

sive treatment, the patient's neurological status kept deteriorating, indicating the progression of intramedullary tumor: the severity of dysesthesias and hypesthesias increased, motor functions kept deteriorating, and the pelvic organ dysfunction developed further. A group of new symptoms pointing to the cerebral damage, i.e., visual and auditory hallucinations and swallowing dysfunction, emerged.



Fig. 5. MRI findings: sagittal T2-weighted and T1-weighted postcontrast images, as well as T1 and T2 pre- and T1 postcontrast weighted axial images of the thoracolumbar part of the spinal cord

Increased intramedullary cystic foci, significantly decreased extradural cystic formation. The width of the tumor at the Th10-Th11 level slightly enlarged; postcontrast enhancement more homogeneous but in a slightly shorter segment. The edematous changes of the same extent; local kyphosis increased.

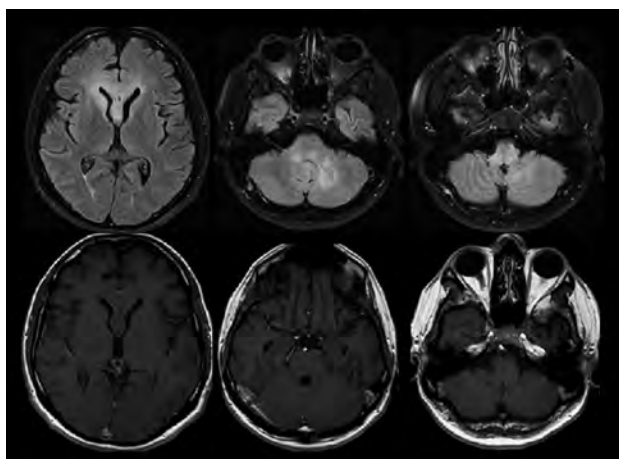


Fig. 6. Cerebral MRI findings: sagittal T2-weighted and T1-weighted images with contrast enhancement

Nonenhancing, T2 hyperintense zones in the heads of the caudate nucleus bilaterally, commissura anterior and the medulla oblongata around the fourth ventricle and in the left cerebellar hemisphere.

The cerebral MRI examination (Fig. 6), performed 5 months after the last spinal MRI, showed multiple nonenhancing foci in the brain, indicative of metastasis and compatible with the new neurological symptoms. A significant progression of the size and invasiveness of the tumor was observed during a simultaneously performed spinal MRI examination (Fig. 7).

### Discussion

The most common classification of spinal cord tumors is anatomically based. It divides them into epidural and intradural and the latter into extramedullary (~84%) and intramedullary (~16%) tumors (3). Spinal intramedullary neoplasms account for about 4%–10% of all CNS tumors and only for



Fig. 7. Sagittal T2-weighted and T1-weighted pre- and postcontrast images of the thoracolumbar part of the spinal cord. Postradiation changes of the vertebrae at the Th4-S3 level, postoperation changes at the Th10-Th11 level, compression fracture of the body of the Th11 vertebra. Significant widening of the spinal cord at the Th5-L1 level. Nonhomogeneously enhancing tumor masses at the Th5-Th12 level with remarkable cystic zones. Edematous changes at the Th2-Th12 level.

2%–4% of all glial tumors (1). Keeping in mind that GBM is the most common primary brain neoplasm in adults, there is a presumption that a relatively low proportion (about 1 to 10) (4) of the absolute number of neuroglial cells in the spinal cord, as compared with the brain, probably determines the rarity of these intramedullary neoplasms (1, 4).

The most important factor affecting survival and treatment possibilities of patients with spinal cord tumors is the histological grade of the tumor (5). Patients with GBM have a very poor prognosis with the survival time after onset being only 6 to 16 months in most cases (4). If a clinical history is longer, it is usually associated with a low-grade diffuse astrocytoma, undergoing a malignant transformation into GBM.

In our reported case, the survival time from the onset of the symptoms was longer than 2 years. It leads us to a presumption that the initial tumor was the astrocytoma of a lower grade than GBM. The premise is reinforced by the apparent changes in the MRI features during the time the patient was observed. A retrospective review of the images showed that the MRI features of the tumor during the initial examinations were more compatible with a low-grade diffuse astrocytoma than with GBM. The emergence of contrast material uptake by the tumor, which was absent in the initial MRI, in the later course of the disease gives us a strong presumption and sustains the theory that the transformation of the malignant tumor into the tumor of a higher grade took place in this case.

MRI is the imaging modality of choice in the evaluation of spinal cord masses. The main 3 tumor features that must be evaluated are cord expansion, postcontrast enhancement, which in some cases may be absent, and cysts, which may be tumoral and nontumoral (6). Unfortunately, there are no pathognomonic features for the exact differentiation of intramedullary tumors and the malignancy degree by MRI alone (6), and although there are some relevant patterns of appearance of intramedullary tumors (7), a histopathological evaluation of a biopsy specimen is necessary.

Although MRI does not provide the exact diagnosis, the findings of MRI together with the medical history of the illness and detailed clinical examination are helpful in the prediction of tumor histology (7).

The clinical presentation of intramedullary GBM depends on the site and the size of the tumor. The main symptoms include back pain, motor disturbance, and bowel and urinary incontinence (8). Exacerbation and worsening of the neurological status are often related to the progression and the growth rate of the tumor.

In the context of intramedullary pathology, the most important tasks for a radiologist are to es-

tablish a right differential diagnosis, initially differentiating the neoplastic process from the non-neoplastic one (1). This is the most difficult task to perform because there are a lot of entities having similar MRI features and mimicking a tumor, and vice versa. The spinal tumor should be differentiated from demyelinating disease, neurosarcoidosis, vascular malformations, ischemia, pseudotumor, chronic arachnoiditis, transverse myelitis, and many others. Intramedullary spinal neoplasms have limited distinctive features on MRI, i.e., there are no pathognomonic imaging findings for differentiation between histologically different tumors (1).

The optimal management of intramedullary GBM remains controversial: radiotherapy has become the standard of care, but the relationship between the extent of surgical resection and survival remains uncertain (9). The majority of authors recommend aggressive multimodal therapy (4, 10) consisting of surgical treatment, chemotherapy, and radiotherapy, which was applied in the presented case.

### Conclusions

Glioblastoma multiforme of the spinal cord is a very rare tumor with no specific imaging features. Careful neurological and radiological evaluations should be made when patient's symptoms indicate an intramedullary lesion. An intramedullary low-grade astrocytoma may undergo a malignant transformation to glioblastoma multiforme in a short period, so an early diagnosis is essential for timely treatment and a better survival prognosis. An exact diagnosis of the histological grade of the tumor may be obtained only through a histopathological examination of a biopsy sample. A close clinical and radiological follow-up is very important for the management of the tumor. Aggressive multimodal treatment may result in prolonged survival.

### Statement of Conflict of Interest

The authors state no conflict of interest.

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