Case Report

Primary intramedullary spinal gliosarcoma: An unusual presentation

ABSTRACT

Gliosarcoma is a biphasic central nervous system malignancy composed of glial and mesenchymal components. It is recognized as a rare variant of glioblastoma with unique histology, immunostaining properties, and natural history. Although usually described to primarily involve the brain, a search of the published literature revealed four reported cases of gliosarcoma arising in the spinal cord. This report describes the case of a 35-year-old woman with progressive back pain with loss of sensation and power of both lower limbs. Magnetic resonance imaging showed an intramedullary unifocal space-occupying lesion in her thoracolumbar spinal cord. Subtotal resection and subsequent histopathological and immunohistochemical studies confirmed the diagnosis of gliosarcoma of the spinal cord. This report further establishes the clinical entity of primary spinal gliosarcoma and proposes the need to consider this possibility among the differential diagnoses of a space-occupying spinal lesion.

KEY WORDS: Glioblastoma multiforme, gliosarcoma, intramedullary, spinal cord

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INTRODUCTION

Gliosarcoma, a variant of glioblastoma multiforme (GBM), is a very rare and aggressive central nervous system (CNS) tumor arising from the glial cells. It is histologically characterized by two distinct components, one resembling GBM and the other sarcoma. The clinical features of the intracranial primary component of the gliosarcoma are the same as that of GBM. However, gliosarcoma is unique in its tendency to spread via the bloodstream, and it is almost exclusively supratentorial site of origin. Gliosarcoma originating in the spinal cord is extremely rare, and only four previously reported cases were found. In this report, we describe primary intramedullary spinal gliosarcoma in a 35-year-old female patient.

CASE REPORT

A 35-year-old woman with no previous history of serious illnesses or comorbidities presented with gradually progressive back pain of insidious onset for 1 year and decreasing sensation and power of both lower limbs for 5 months. Multiple consultations during this period led to no diagnosis or symptom relief. She finally visited a district hospital in December 2018 with urinary retention and inability to stand or walk. She was referred and admitted to a teaching hospital. Physical examination revealed decreased sensation to crude and fine touch, pain, and temperature on front and back of the lower trunk and bilateral lower limbs almost symmetrical in distribution. Power of both lower limbs was 3/5. Contrast-enhanced magnetic resonance imaging of the CNS showed a single contrast-enhancing intradural intramedullary soft tissue lesion of the spinal cord occupying the D9–L1 vertebral levels [Figure 1]. The lesion appeared heterogeneously hyperintense on T2 sequence with associated perilumbar edema. There was syrinx formation superiorly, and posteriorly, there was evidence of extension of the lesion. Contrast-enhanced computed tomography scan of the thorax and abdomen did not reveal any abnormality.

A subsequent polymerase chain reaction test for *Mycobacterium tuberculosis* from the cerebrospinal fluid (CSF) was negative.

Other CSF studies were within normal limits.

Subtotal resection of the mass was done under general anesthesia but without relief of pain or neurodeficits.

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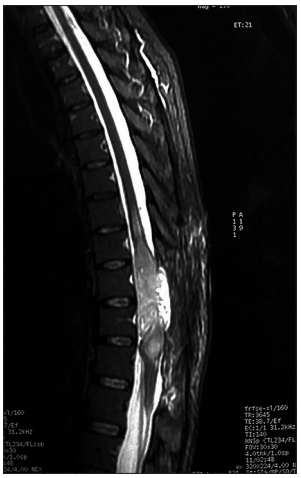

Figure 1: T2-weighted magnetic resonance imaging of the spine showing a heterogeneously hyperintense soft tissue lesion of the spinal cord occupying the D9–L1 vertebral levels with associated perilumbar edema and syrinx formation cranially (original image courtesy author no. 3)

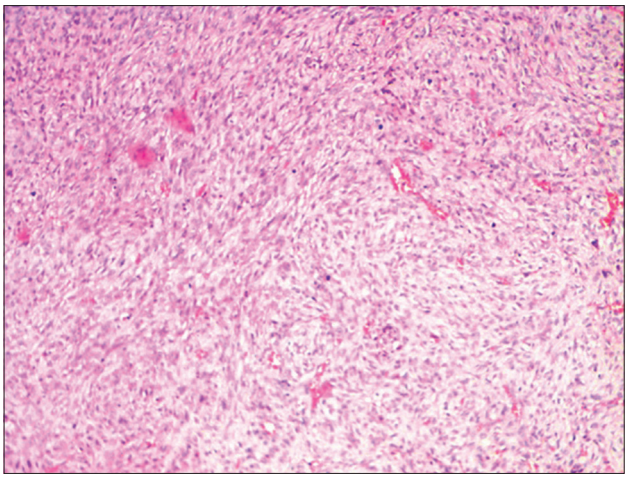

Figure 2: Scanning magnification showing a biphasic tumor in a hematoxylin and eosin-stained slide. The left side of the picture showing haphazardly arranged spindle-shaped cells, and the right and lower parts of the image showing stellate-to-plump cells in a fibrillary background(original image courtesy author no. 2)

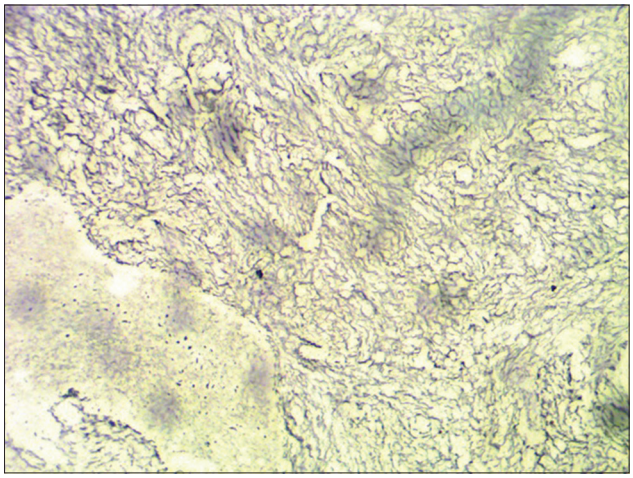

Figure 3: A reticulin stain preparation showing reticulin meshwork in the sarcomatous area on the right side and the glial component of the tumor devoid of reticulin on the left (original image courtesy author no 2)

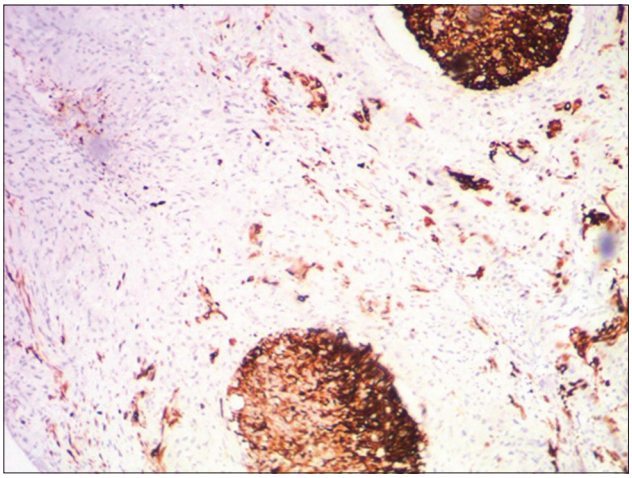

Figure 4: Slide stained for glial fibrillary acidic protein (GFAP) showing two islands (top right and bottom centre) of GFAP positive glial tissue surrounded by sarcomatous area negative for GFAP (original image courtesy author no 2

Histopathological examination with hematoxylin and eosin staining revealed areas of high cellularity and microvascular proliferation resembling high-grade astrocytoma [Figure 2]. These areas contained stellate to plump cells in a fibrillary background. These regions stained with antibodies against glial fibrillary acidic protein (GFAP) but did not take up reticulin stain [Figures 3 and 4]. There were also regions of haphazardly arranged spindle-shaped cells, characteristic of sarcoma surrounding the areas containing the high-grade astrocytoma. These sarcomatous areas showed reticulin meshwork in the stroma.

The patient was thereafter referred to us for external beam radiation. The treatment planned for the patient was postoperative radiotherapy 36 Gy in 20 fractions, with further planned dose increment according to tolerability up to 54 Gy in the second phase. The patient defaulted after eight fractions of 1.8 Gy each due to a combination of reasons unrelated to the treatment or its toxicities and opted for palliative management at her home. Telephonic follow-up revealed that she passed away in August 2019 due to respiratory complications. Her back pain and lower body neurodeficits had remained gradually progressive during this period, with the former being partially controlled on oral opioids.

DISCUSSION

A brain malignancy comprising glial and mesenchymal components was first described and named as gliosarcoma by Stroebe in 1895.[1] Gliosarcoma is characterized by its biphasic growth pattern: a glial component fulfilling the histologic

criteria of glioblastoma including immunoreactivity for GFAP and a mesenchymal component with dense extracellular matrix deposition evident on reticulin staining, which may have fibroblastic, cartilaginous, osseous, adipose, or striated or smooth muscle differentiation. The sarcomatous and glial components have been found to harbor similar genetic abnormalities, thus suggesting a monoclonal origin and subsequent focal mesenchymal metaplasia of the primary glioma.[2] The recent WHO classification of CNS tumors classifies gliosarcoma as a variant under GBM,[3] comprising about 2% of all GBM.

Gliosarcoma resembles GBM in presenting features, age distribution, and male preponderance. Imaging and operative findings are similar to GBM on some occasions. However, on other occasions, gliosarcoma may mimic meningioma, having a well-defined margin and homogeneous contrast enhancement on imaging, and being firm and well demarcated on gross appearance. Gliosarcoma is also unique in its predilection for the temporal, and to some extent, frontal lobes. It is rarely found in the infratentorial location.[4] Carstens *et al.* described the first report of spinal gliosarcoma arising in the thoracolumbar region in a patient previously treated for cerebellar oligodendroglioma with resection and adjuvant radiation.[5] Subsequently, a patient of multifocal primary gliosarcoma of the spinal cord was described by Kumar and Finn,[6] who was treated with postoperative radiotherapy to cervical, thoracic, and lumbar spine with concurrent temozolomide followed by avastin on further progression. Thereafter, unifocal gliosarcoma of the thoracic spine reported by Wu *et al.*[7] was also treated with adjuvant radiotherapy with concurrent and adjuvant temozolomide. Soon afterward, another cervical spinal gliosarcoma with rhabdomyoblastic differentiation described in a 6-year-old girl by Yao *et al*.[8] treated only with low-dose radiation. Another report by Barbagallo *et al*.[9] described a unique case of high cervical meningeal sarcoma with no connection to the spinal cord harboring GFAP reactive components. Unlike GBM, gliosarcoma may metastasize via the bloodstream to extracranial sites, a feature thought to be mainly contributed to by the sarcomatous component. Treatment of gliosarcoma has mirrored that of GBM, given the paucity of evidence regarding optimal therapy. Surgery should aim at gross total resection. The use of postoperative radiation is supported by observational and cohort studies.[4] Published case series have used varying doses of radiotherapy and various chemotherapeutic agents, with very little data supporting the superiority of any one over the others. Prognosis is uniformly poor, with a median survival ranging from 8 to 11 months in most series, and differences in survival with GBM have been nonsignificant.[4]

To conclude, gliosarcoma clearly needs to be considered as a possible differential diagnosis in case of a suspicious spinal cord space-occupying lesion, with the additional implication that CSF and hematogenous metastases should be excluded. Treatment results should also be documented to contribute to the sparse evidence available for this disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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