Temporoparietal Gliosarcoma: A Case Report with Literature Review

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ABSTRACT

Gliosarcoma (GSM) is a primary tumor of the central nervous system composed of both malignant glial and sarcomatous elements. GSM is classified as grade IV according to World Health Organization (WHO) and regarded as aggressive tumor and often requires both surgery and radiotherapy. The incidence of GSM is between 1.8%-2.8% of all malignant gliomas and thus represents an exceptionally rare neoplasm. Consequently, our knowledge about this entity is limited to small retrospective case series and case reports. Here, we describe a case of a 46-year-old male with progressive right extremity weakness, accompanied with aphasia. Imaging studies showed a tumor mass in the left temporoparietal region. Surgery was performed and gross total removal was achieved. Histopathology finding established a diagnosis of GSM. The extremity weakness was improved postoperatively within a week.

Keywords: Gliosarcoma, Rare case, Temporoparietal


INTRODUCTION

Gliosarcoma (GSM), from histopathology point of view, is a rare variant of Glioblastoma Multiforme (GBM) and is the fastest growing central nervous tissue tumors with biphasic tissue patterns.1,2,3 GSM were described for the first time in 1898 as an unusual variant with sarcomatous elements.3 GSM recognized in World Health Organization (WHO) classification as WHO grade IV that display an aggressive behavior, including neoplastic glial cells in association with spindle cell sarcomatous elements.4 It usually affects 4th to 6th decade of life with slight male preponderance; most of them had involvement of temporal lobe.1,2,5,6,7 These highly malignant and rare primary tumor represent 1.8%-2.8% of all malignant gliomas. It has a propensity for distant metastasis and usual site of involvement is lung followed by liver and bone.8,9 Here we present a case report of 46 years old male who presented with space occupying lesion in brain which turned out to be GSM after surgical resection and histopathological evaluation.

CASE REPORT

A 46-year-old male suffered from progressive right extremity weakness followed by aphasia in the past 2 days and worsened one day prior to admission. He also had progressive intermittent headaches for the last 1 week. Neurological examination indicated a left cranial nerve (CN) 7 paralysis. Other cranial nerves were normal with right hemiparesis. Routine laboratory blood- and urine-examinations showed normal findings. Imaging study using computed tomography (CT) scanning showed a tumor mass in the left temporoparietal region. Brain CT scan images showed an isohyperdense lesion of the left temporoparietal region that enhanced in-homogeneously with contrast, with peri-tumoral edema, and compressed sulcus and gyrus (Figure 1). Based on the clinical features, pre-operative differential diagnosis of tuberculoma was suspected. A craniotomy tumor removal from a supine position was performed and gross total removal was achieved. Intra-operatively, we

Figure 1  Head CT scan with contrast (above) and non-contrast (below) showing a heterogeneous lesion in the right temporal lobe with significant peri-tumoral edema, mass effect and midline shift.
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found intratumoral bleeding as expected. The mass itself was tan-pink, semi-firm, easily aspirated and heavily vascularized. Adequate tumor decompression with controlled tumor bleeding was performed and the solid component was biopsied. Multiple section studies showed that the lesion was composed of round, oval and spindle shaped cells arranged in fascicles. In some foci, they were arranged irregularly. The tumor cells had oval, elongated, and vesicular nuclei with prominent nucleoli as well as eosinophilic cytoplasm. Mono- and multi-nucleated tumor giant cells was also observed. Numerous atypical mitotic figures were also noted. Occasional foci showed necrosis and pseudo-pallisading pattern of arrangement of the tumor cells (Figure 2). Glial with malignant mesenchymal differentiation component was seen in some foci (Figure 3). The final histopathology report established the diagnosis of GSM. The extremity weakness was improved postoperatively within a week.

DISCUSSION

GSMs are usually occurred in the cerebral hemispheres involving the temporal, frontal, parietal and occipital lobes in decreasing order of frequency, respectively. It is commonly found in adults males at the fourth to sixth decades of life with mean peak incidence age at 52 years old and quite similar to that of GBM. However, some rare case may occur in children. Rarely, GSM may occur in the posterior fossa and spinal cord. An unusual location of a GSM developing from an ependymal has been identified in some cases as well as multifocal occurrence of GSM. Clinically, most of the patients present with symptoms of increased intracranial pressure due to an expanding intracranial mass such as headache, hemiparesis, seizures and cognitive decline. In this case, the chief complaint was progressive right extremities weakness although it was accompanied with intermittent headache. GSM was originally defined as a GBM in which sarcomatous component was the consequence of malignant transformation of proliferating tumor blood vessel; recently, there was a cytogenetic- and molecular-evidence for a monoclonal origin of the both the glial and sarcomatous component originating via aberrant mesenchymal differentiation.

The mixture of gliomatous and sarcomatous component of GSM result in striking biphasic tissue pattern. The glial portion is usually astrocytic in nature and anaplastic, mostly showing the typical features of a GBM. The sarcomatous component by definition shows signs of malignant transformation such as nuclear atypia, mitotic activity, necrosis and often demonstrates the typical pattern of fibrosarcoma with densely packed long bundles of spindle cells. The high content of connective tissue contributes to the gross appearance of GSM as a firm, well circumscribed mass that often be

![Figure 2](A) Spindle shaped tumor cells with one foci showing necrosis and pseudo-pallisading pattern (H&E, x20). (B) Tumor tissue with spindle shaped tumor cells, multi nucleated tumor giant cells and congested blood vessels (H&E, x200). (C) Tumor tissue with mesenchymal type and cartilage differentiation (H&E, x200). (D) Tumor tissue with mesenchymal type and muscle differentiation (H&E, x100)

![Figure 3](A) Spindle shaped tumor cells positive for SMA (x100). (B) Tumor cells positive for Reticulin (x100). (C) Tumor cells positive for Vimentin (x100). (D) Tumor cells positive for GFAP (x100)
A subset of cases may show additional lines of mesenchymal differentiations, e.g. the formation of cartilage, bone, osteoid-chondral tissue, smooth and striated muscle and even lipomatous features as in our case (Figure 2C and 2D).³⁸,¹⁹ According to the histopathology evaluation, we differentiated the diagnosis with intracranial leiomyosarcomas since it’s correspond histologically to their soft tissue counter parts, expressing smooth muscle actin (Figure 3A). The reticulin and vimentin staining showed abundant connective tissue fibers (Figure 3B and 3C). This component did not express GFAP, which, on the contrary, was observed in the glial part (Figure 3D). The demonstration of a clearly malignant mesenchymal GFAP-negative component is important to distinguish true GSM from GBM with a florid fibroblastic proliferation (desmoplasia) elicited by meningial invasion; 11% GSM contains PTEN mutations (38-45%), p16INK4-alpha deletions (38%) and TP53 mutations (23-24%), but shows infrequent EFGR amplifications (0-8%); suggesting that they have a distinct profile like primary GBM, except for the infrequent EFGR amplification.⁵,⁹,¹⁰,¹¹,¹⁵,¹⁶ Comparative genomic hybridization in 20 GSM revealed that chromosomal imbalances commonly detected were gains on chromosomes 7 (75%), X (20%), 9p and 20q (15% each) as well as losses of chromosomes 10, 9p (35% each), 13q (15%).¹¹ Metastases has been reported in up to one third of cases, mainly to the lungs, pleura, lymph nodes, bone marrow, liver, spinal cord, kidney and peripancreatic areas.⁵,⁹ Management of GSM includes maximum surgical decompression with post-operative radiotherapy.⁵,⁹ The possible role of chemotherapy in GSM is still undefined and still need to be investigated.⁵,⁹,¹⁰,¹¹,¹⁵

CONCLUSION

A case of GSM in a 46-year-old male presented with a rare temporoparietal Glioblastoma which was established histopathologically and successfully treated by craniotomy. Although GSM is considered as a rare case, its management is still need to be developed especially regarding the role of chemotherapy.

REFERENCES

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