Gliosarcoma with Prominent Leiomyosarcomatous Differentiation: Case Report

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Objective and importance: Gliosarcoma is a rare variant of glioblastoma multiforme that calls for the simultaneous presence of a gliomatous differentiation and a sarcomatous differentiation. A gliosarcoma with a sarcomatous differentiation that corresponds to a leiomyosarcoma has not previously been presented. This report presents an exceptional gliosarcoma with a prominent leiomyosarcomatous differentiation.

Clinical presentation: A 56 year-old man presented with weakness on the right arm and right leg that had progressed over one week. His physical examination revealed marked right-sided hemiparesis. A left-sided mass lesion was observed on magnetic resonance imaging that involved the surface of the parasagittal aspect of the left frontal lobe of the brain and that was associated with intense edema. Intervention: The patient underwent a craniotomy with resection of the mass lesion and was diagnosed as having a gliosarcoma with a prominent leiomyosarcomatous differentiation. Based on the imminent risk of relapse, the decision was made to proceed with radiation therapy. He suffered a relapse at nine months following the diagnosis, declined salvage treatment and died within one month.

Conclusion: Although an exceptional gliosarcoma with a prominent leiomyosarcomatous differentiation might be treated with surgery and radiation therapy, the outcome remains dismal.

Key words: Gliosarcoma, leiomyosarcomatous differentiation, surgery, radiation therapy

Leiomyosarkomatöz Diferansiyasyon Gösteren Gliosarkoma Olgusu


Anahtar kelimeler: Cerrahi, gliosarkom, leiomyosarkomatöz değişşim, radyoterapi

Gliosarcoma is a rare variant of glioblastoma multiforme that calls for the simultaneous presence of a gliomatous differentiation and a sarcomatous differentiation (1). The gliomatous differentiation characteristically corresponds to a glioblastoma multiforme whereas the sarcomatous differentiation usually corresponds to a fibrosarcoma, a chondrosarcoma or an osteosarcoma (2-3). A gliosarcoma with a sarcomatous differentiation that corresponds to...
a leiomyosarcoma has not previously been presented. This report presents an exceptional gliosarcoma with a prominent leiomyosarcomatous differentiation.

CASE REPORT

A 56 year-old man presented with weakness on the right arm and right leg that had progressed over one week. His physical examination revealed marked right-sided hemiparesis. A left-sided mass lesion at low signal intensity was observed on magnetic resonance imaging that involved the surface of the parasagittal aspect of the left frontal lobe of the brain and that abutted the meninges. The mass lesion appeared to compress the frontal horn of the left lateral ventricle and to traverse the corpus callosum and was associated with the necrotic foci and the intense edema leading to left-sided shift of the midline structures. Following the administration of the contrast medium, the mass lesion demonstrated notable circumferential enhancement (Figure 1). He underwent a craniotomy with resection of the demarcated mass lesion, that appeared to originate from the surface of the brain parenchyma, through an inter-hemispheric approach.

Macroscopical examination revealed a white-gray mass lesion that was rubbery in consist-
tency. On microscopical examination using hematoxylin and eosin staining, the mass lesion was composed of the atypical cells with a sarcomatous differentiation that were characterized by the spindle forming nuclei and the bipolar cytoplasms and the atypical cells with a gliomatous differentiation that were characterized by the hyperchromatic nuclei and the eosinophilic cytoplasms. The mass lesion was associated with the abundant mitotic figures and the vascular proliferation and the necrotic foci and was demarcated from the histiocytic proliferation and the demyelination in the surround. On microscopical examination using reticulin staining, the clusters of the atypical cells with the gliomatous differentiation that were devoid of the reticulin framework were interspersed among the fascicles of the atypical cells with the sarcomatous differentiation that were embedded in the reticulin framework (Figure 2).

Immunohistochemical analysis proved diffuse positive staining for vimentin, regional positive staining for alpha-smooth muscle actin protein and S100 proteins and focal positive staining for neurofilament proteins for the atypical cells with the sarcomatous differentiation, focal positive staining for glial fibrillary acidic protein and pancytokeratin for the atypical cells with the gliomatous differentiation and negative staining for desmin, epithelial membrane antigen, melanoma specific antigens, CD34 antigen, CD99 antigen, CD117 antigen, synaptophysin and collagen type IV. The proliferation index was estimated as 20% on staining for Ki-67 antigen (Figure 3).

The patient was diagnosed as having a gliosarcoma with a prominent leiomyosarcomatous differentiation. He experienced prompt improvement and his physical examination was unremarkable apart from obscure weakness on the right arm and right leg at one week following surgery. Based on the imminent risk of relapse, the decision was made to proceed with radiation therapy. He suffered a relapse at nine months following the diagnosis, declined salvage treatment and died within one month.
DISCUSSION

In accordance with the World Health Organization classification for the tumors of the central nervous system, a gliosarcoma is a rare variant of a glioblastoma multiforme that shares the clinical course and the genetic features of a glioblastoma multiforme, yet the simultaneous presence of a gliomatous differentiation and a sarcomatous differentiation constituting the disparity (1). The gliomatous differentiation corresponds characteristically to a glioblastoma multiforme and uncharacteristically to an oligodendroglia or an ependymoma whereas the sarcomatous differentiation usually corresponds to a fibrosarcoma, a chondrosarcoma or an osteosarcoma and unusually to an angiosarcoma, a liposarcoma or a rhabdomyosarcoma (2-3). In similarity to a glioblastoma multiforme, a gliosarcoma tends to affect the aged men and tends to involve the frontal lobe or the temporal lobe of the brain (4). On neuroimaging, a gliosarcoma typically appears as a mass lesion that is demarcated from the surround and is accompanied by intense edema, necrotic foci and notable circumferential contrast enhancement (5). Although the mass lesion typically originates from the surface of the brain parenchyma, the presumptive diagnosis on neuroimaging might include a meningioma based on the fact that the mass lesion frequently abuts the meninges (6). In a study by Dwyer and colleagues that involved the evaluation of the magnetic resonance imaging features of six gliosarcomas, the mass lesion was demarcated from the surround, was at intermediate signal intensity, was accompanied by intense edema and notable circumferential contrast enhancement and abutted the meninges in all gliosarcomas (7).

On microscopical examination, the diagnosis of a gliosarcoma is based on the observation of the atypical cells with a sarcomatous differentiation that are arranged in fascicles and interspersed among the atypical cells with a gliomatous differentiation (2). The atypical cells with a gliomatous differentiation are characterized by the abundant mitotic figures, the necrotic foci and the vascular proliferation in the surround whereas the atypical cells with a sarcomatous differentiation are characterized by the spindle forming nuclei and the bipolar cytoplasms (3). In a study by Sreenan and Prayson that involved the microscopical examination of the tissue samples of thirteen gliosarcomas using hematoxylin and eosin staining, the atypical cells with a gliomatous differentiation comprised more than 75% of the tissue samples in five gliosarcomas as opposed to less than 25% of the tissue sample in one gliosarcoma whereas the atypical cells with a sarcomatous differentiation corresponded to a fibrosarcoma in twelve gliosarcomas (8). Immunohistochemical analysis highlights the distinction between the sarcomatous differentiation and the gliomatous differentiation and enables the categorization of the sarcomatous differentiation. The mainstay of the immunohistochemical analysis involves staining for glial fibrillary acidic protein regarding the gliomatous differentiation and staining for vimentin regarding the sarcomatous differentiation (9). In a study by Machuca and colleagues that involved the immunohistochemical analysis of the tissue samples of four gliosarcomas, the atypical cells with a gliomatous differentiation proved diffuse positive staining for glial fibrillary acidic protein, focal positive staining for vimentin and negative staining for CD34 antigen whereas the atypical cells with a sarcomatous differentiation proved diffuse positive staining for vimentin and negative staining for glial fibrillary acidic protein and CD34 antigen in all gliosarcomas (10).

A gliosarcoma characteristically follows an aggressive clinical course and the outcome remains far from satisfactory (4). In the absence of the randomized data comparing the treatment options that include surgery and radiation therapy, the treatment decisions are based on the non-randomized data and the institutional policies.
Surgery alone might be appropriate for an undersized gliosarcoma, for which complete resection is most likely to be performed whereas radiation therapy should follow surgery for a sizable gliosarcoma for which complete resection is not likely to be performed (11). In a study by Lutterbach and colleagues that involved the evaluation of the treatment and the outcome of twelve gliosarcomas, a near complete resection was performed in nine gliosarcomas as opposed to an incomplete resection in three gliosarcomas and radiation therapy followed surgery in twelve gliosarcomas whereas relapse resulted in death at 2 months to 19 months following the diagnosis in all gliosarcomas (12).

CONCLUSION

Although an exceptional gliosarcoma with a prominent leiomyosarcomatous differentiation might be treated with surgery and radiation therapy, the outcome remains dismal.

ACKNOWLEDGMENT

The authors are indebted to Bernd Scheithauer, M.D. from the Department of Anatomic Pathology at Mayo Clinic College of Medicine for kindly reviewing the tissue samples.

REFERENCES