Case Report

Gliosarcoma with long progression free survival: A case report and literature review

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Abstract

Background: Gliosarcoma (GS) is a primary rare malignant brain tumor that accounts 4% of all high-grade glial tumor of the brain.

Case Description: We present a 45-year-old female admitted to our center with progressive headache since 1 month ago concomitant with nausea and emesis and generalized weakness. Imaging revealed a large solid mass with well-defined margin and some cystic portions that enhanced brightly with contrast. We decided to operate the patient via right parietal craniotomy and we totally resected all visible portions of the mass, as en bloc resection. The histopathological report of the mass was GS. We are following the patient up to now, for about 50 months, and she is good without any complaint or neurologic deficit. All follow-up magnetic resonance imaging (MRI) did not show any tumor recurrence.

Conclusion: Aiming to achieve longer progression-free survival in cases of GS, we recommend resecting all portions of the mass as much as possible, so named en bloc resection, and then refer the patients for appropriate and timely chemoradiotherapy.

Key Words: En bloc resection, gliosarcoma, overall survival, progression-free survival

BACKGROUND

Gliosarcoma (GS) is a primary rare malignant brain tumor that contains both gliomatous and sarcomatous (mesenchymal) components and accounts 4% of all high-grade glial tumor of the brain. Based on the 2007 World Health Organization (WHO) classification, GS is a Grade 4 tumor same as glioblastoma multiform (GBM). In the literature, mean overall survival (OS) of GS in untreated patients is 4 months and with multidisciplinary tri-modal therapy, the mean OS is about 15 months. We present a patient with GS that we treated her and she has 50 months disease-free survival up to now.

CASE DESCRIPTION

We present a 45-year-old female admitted to our center with progressive headache since 1 month ago concomitant with nausea, emesis, and generalized weakness. She had minimal headache since 2 years ago and, on admission clinical examination, we did not find any neurologic deficits. Also, the patient had bilateral pupillary edema on fundoscopy. She had bilateral positive Hoffmann’s sign. On first brain computed tomography (CT), we saw a large heterogeneous mass with some vasogenic edema...
at right parieto-occipital region with 0.5 cm midline shift that compressed right occipital horn of lateral ventricle [Figure 1a]. The brain magnetic resonance imaging (MRI) with and without gadolinium (Gd) revealed a large solid mass with well-defined margin and some cystic portions that enhanced brightly with Gd [Figure 1b and c]. The vasogenic edema was less than our expectation.

According to very large mass effect, we decided to operate the patient via right parietal craniotomy. The tumor had a more firm consistency compared with adjacent normal brain tissue and it was well demarcated and we totally resected all visible portions of the mass, as en bloc resection. The histopathological report of the mass was GS [Figure 2a–d]. After surgical treatment, the patient’s treatment continued by chemotherapy with temozolamide and a course of radiotherapy. We are following the patient up to now, for about 50 months, and she is good without any complaint or neurologic deficit. All follow-up MRI did not show any tumor recurrence [Figure 1d].

DISCUSSION

The GS, as known Feigin tumor, is a rare and malignant glioma.\[3\] With best existing tri-modal therapy, contained surgical gross total resection and then chemoradiotherapy, this tumor has very poor prognosis.\[10\] For GS, histopathological exam shows both gliomatous and sarcomatous components. At the surgery compared with GBM, the borders of tumor are more distinct because of presence of sarcomatous component, but on the other hand, the patients have less response to adjuvant treatment contained chemotherapy and radiotherapy during postoperative period.

In the literature, we found only eight reports of GS with OS >2 years [Table 1]. Considering the term of survival, we have to pay attention to this subject that the “overall survival” is different with “progression-free survival” (PFS). The patients with longer OS have not necessarily longer PFS and they may have multiple recurrence period needed complete treatment courses that patients suffered from them. Longer PSF can predict more excellent quality of life (QOL). Compared with our case, only four reports of GS had a longer OS concomitant with long PFS.\[1,4,9,10\]

Although in almost all cases, infiltrative nature of gliomatous tumors hinder to remove all portions of them, because of sarcomatous components of GS, differences in consistency of tumoral tissue compared with brain tissue assist to define a relatively well-demarcated plane between them during surgery. In addition, en bloc resection with preservation of tissue planes during dissection prevents missing any tumor component intermingled with brain parenchyma at the time of dissection. Finally, location of the lesion in non-eloquent area of brain lets intraoperative manipulations and more feasible en bloc tumor removal. We think all these factors together in association with appropriate and timely adjuvant therapy helps surgeon to give the most chance of PFS to the patient with GS.

CONCLUSION

We present a patient with GS that has a long PFS after surgical treatment. Aiming to achieve longer PFS in cases of GS and regarding to more distinct consistency, we recommend resecting all portions of the mass, as much as possible, to reach en bloc resection and then refer the patients for appropriate and timely chemoradiotherapy.
Table 1: A literature review for all reported cases with GS and longer OS

| Author/year | Patient’s age (years old)/sex | Presenting signs and symptoms | Site of tumor | OS          | PFS         | Number of recurrences | Presence of metastasis |
|-------------|-------------------------------|-------------------------------|---------------|-------------|-------------|------------------------|------------------------|
| Winkler et al. [10] | 61/female | Headache | Left parietal | 22 years | 20 years | 1 | No |
| Chen et al. [2] | 31/female | Dizziness, headache, vomiting | Right temporal | 92 months | 46 months | 2 | Yes |
| Linhares et al. [7] | 51/female | N/A | Left hemi-body paresthesia, seizure, dysarthria | 36 months | 21 months | 1 | No |
| Huo et al. [4] | 47/male | Headache, right leg weakness | Left lateral ventricle (frontal horn), corpus callosum | N/A | 130 months | No | No |
| Huo et al. [4] | 63/female | Headache, nausea | Right temporal | N/A | 48 months | No | No |
| Burzynski et al. [1] | 9/male | Seizure, vomiting | Right temporal, pons | N/A | 13 years | 1 | Yes |
| Kalita et al. [3] | 23/female | Headache, nausea, vomiting, left arm weakness | Right frontal, right lateral ventricle | 31 months (alive until the time of article publication) | 28 months | 1 | No |
| Wang et al. [3] | 21/male | Headache | Right frontal | Alive until the time of article publication | 111 months | 1 as a meningeal sarcoma | No |
| Tabibkhooei et al. (2018) current study | 45/female | Headache, nausea, vomiting, generalized weakness | Right parieto-occipital | Alive until the time of article publication | 50 months | No | No |

N/A=Not available

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.

REFERENCES
1. Burzynski SR, Janicki TJ, Burzynski GS, Marszalek A. Long-term survival (>13 years) in a child with recurrent diffuse pontine gliosarcoma: A case report. J Pediatr Hematol Oncol 2014;36:e433-9.
2. Chen L, Xiao H, Xu L, Zou Y, Zhang Y, Xu M. A case study of a patient with gliosarcoma with an extended survival and spinal cord metastases. Cell Biochem Biophys 2012;62:391-5.
3. Feigin IH, Gross SW. Sarcoma arising in glioblastoma of the brain. Am J Pathol 1955;31:633-53.
4. Huo Z, Yang D, Shen J, Li Y, Wu H, Meng Y, et al. Primary gliosarcoma with long-survival: Report of two cases and review of literature. Int J Clin Exp Pathol 2014;7:6323-32.
5. Kalita O, Zlevorova M, Megova M, Vaverka M, Trojanec R, Tuckova L. A patient with primary intraventricular gliosarcoma and long-term survival-A case report. Klin Onkol 2016;29:454-9.
6. Karsy M, Gelbman M, Shah P, Balumbu O, Moy F, Arslan E. Established and emerging variants of glioblastoma multiforme: Review of morphological and molecular features. Folia Neuropathol 2012;50:301-21.
7. Linhares P, Martinho O, Carvalho B, Castro L, Lopes JM, Vaz R, et al. Analysis of a synchronous gliosarcoma and meningioma with long survival: A case report and review of the literature. Surg Neurol Int 2013;4:151.
8. Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, Burger PC, Jouvet A, et al. The 2007 WHO classification of tumours of the central nervous system. Acta Neuropathol 2007;114:97-109.
9. Wang Z, Kong QT, Wu XH, Zhu XX. Long-term survival in gliosarcoma with radiation-induced meningeal sarcomas: Case report and molecular features. J Cancer Res Ther 2015;11:651.
10. Winkler PA, Buttner A, Tomezzoli A, Weis S. Histologically repeatedly confirmed gliosarcoma with long survival: Review of the literature and report of a case. Acta Neurochir (Wien) 2000;142:91-5.