Pakistan Journal of Neurological Sciences (PJNS)

Volume 4 | Issue 1 Article 6

4-2009

Gliosarcoma

Rafael Cincu
Miguel Servet University Hospital

Juan F. Martín Lázaro
Quiron Clinic, Madrid, Spain

José Luis Capablo Liesa
Miguel Servet University Hospital

Jose Eiras
Miguel Servet University Hospital

GLIOSARCOMA: AN UNCOMMON BRAIN TUMOR

Rafael Cincu,[1] Juan F. Martín Lázaro,[2] José Luis Capablo Liesa,[3] and **Jose Eiras[1]**

Departments of [1]Neurosurgery and [3]Neurology, Miguel Servet University Hospital, Zaragoza, Spain; [2]Department of Medical Biochemistry, Quiron Clinic, Madrid, Spain

Correspondence to: Dr. Cincu, Consultant Neurosurgeon, Department of Neurosurgery, Miguel Servet University Hospital, Zaragoza, Spain. Email: rafael.cincu@gmail.com

Pak J Neurol Sci 2009; 4(1): 20-22

ABSTRACT

Gliosarcomas are rare bimorphic intraaxial malignant neoplasms of the central nervous system. A 69-year-old gentleman presented with a history of right-sided weakness and slurred speech. Brain imaging (CT scan and MRI) was suggestive of a left fronto-parietal mass lesion with contrast enhancement at the periphery. Neuropathological examination revealed a malignant brain tumour presenting a biphasic tissue pattern with gliomatous and mesenchymal components suggestive of gliosarcoma that was further evaluated immuno¬histochemically. Although the treatment of gliosarcomas is almost similar to glioblastomas (surgical resection and, depending on clinical status, radiotherapy and/or chemotherapy) the prognosis of gliosarcomas remains poor.

Gliosarcomas are rare bimorphic intraaxial malignant neoplasms of the central nervous system and comprise approximately 2% of all glioblastomas.[1,2,3,4] We present a case of gliosarcoma and review the literature of this uncommon clinical entity.

CASE REPORT

A 69-year-old gentleman presented with a history of right-sided weakness and slurring of speech. On examination, he had right hemiparesis grade 4/5 (lower limb weaker than upper limb) and slurred speech. General and systemic examination were unremarkable. CT scan showed a left fronto-parietal isodense lesion with marked peripheral edema in the left frontal region; peripheral enhancement was noted after administration of contrast (Figure 1). On MRI the lesion was hypointense on T1- and hyperintense on T2-weighted images, and was irregularly enhancing with contrast. Total surgical excision was undertaken.

Neuropathological examination revealed a malignant brain tumour presenting a biphasic tissue pattern with gliomatous and mesenchymal components suggestive of gliosarcoma. The glial component was similar to a glioblastoma, with nuclear pleomorphism, high mitotic index, marked vascular proliferation and foci of necrosis, either focal or geographical. Gliomatous areas showed glial-fibrillary acidic protein expression. The mesenchymal component consisted of areas with densely packed long bundles of spindle cells in a storiform pattern. Immunohistochemically the cells expressed vimentin and were surrounded by a dense network of reticulin fibres as

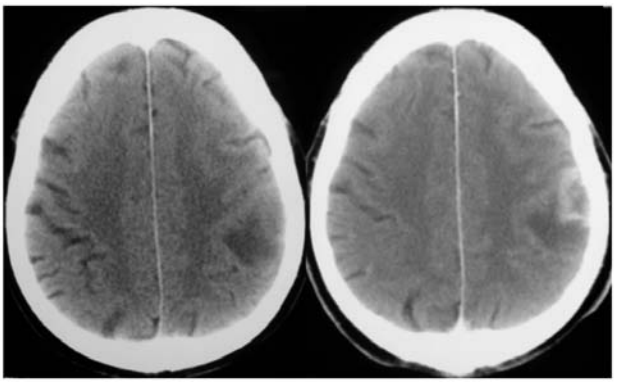

Figure 1 *CT scan displaying an isodense lesion with marked peripheral edema in the left frontal region (left). Peripheral enhancement of the lesion was observed after contrast medium injection (right)*

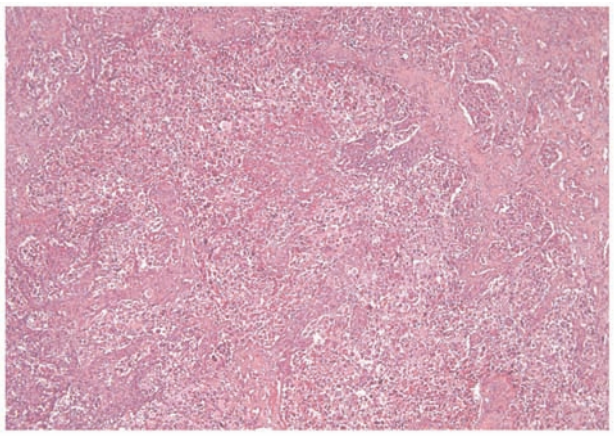

Figure 2 *Photomicrograph displaying the biphasic architecture of gliosarcoma (H&E 40X)*

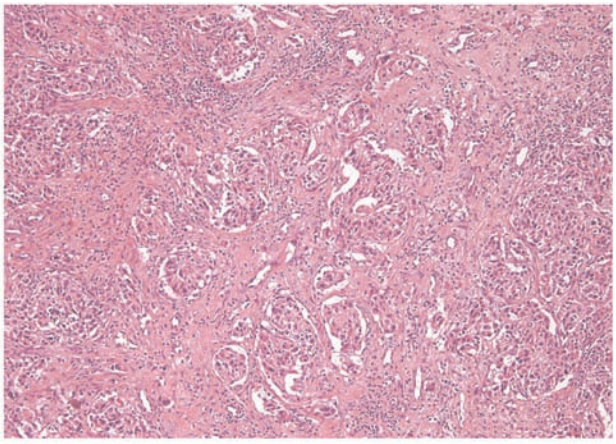

Figure 3 *Photomicrograph displaying the biphasic architecture of gliosarcoma (H&E 400X)*

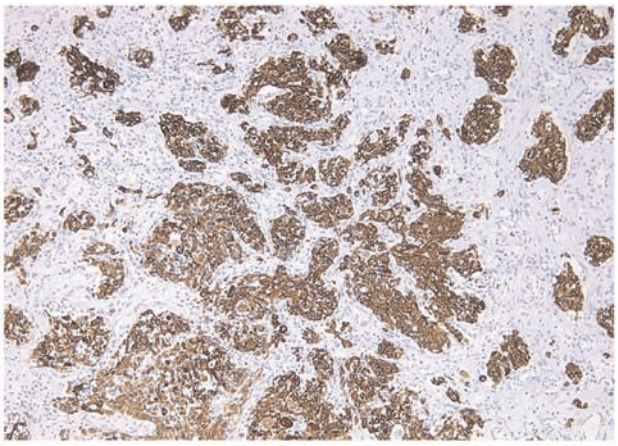

Figure 4 *Photomicrograph displaying immunopositivity for PGA (polyglycolic acid)*

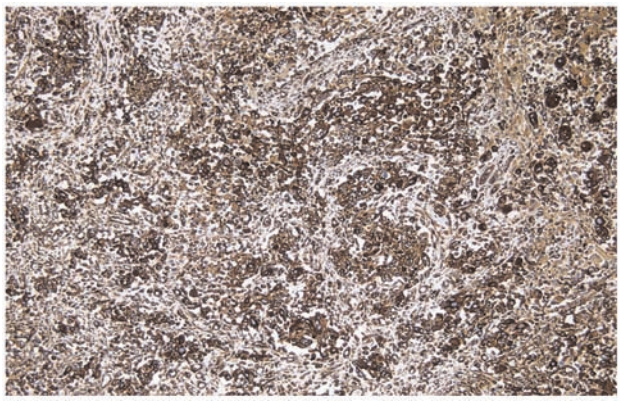

Figure 5 *Photomicrograph displaying immunopositivity for vimentin*

typical features of sarcomatous cells (Figures 2-6). Additionally, the patient received 60 Gy of radiation therapy.

DISCUSSION

As in the present case, gliosarcomas typically develop in older patients. They are slightly more common in males than females.[5,6,7] Clinically, primary glioblastomas and gliosarcomas are indistinguishable, as both variants present with a short clinical course, low median survival and similar peak of incidence.[4,5,7] Gliosarcomas (GSa) are characterized by a biphasic histological pattern displaying both glial and sarcomatous components.[8,9,10]

The histogenesis of GSa has been the subject of debate. Feigin and Gross, who first described GSa, suggest that the cell of origin would arise from neoplastic transformation of blood vessels in a preexisting glioblastoma.[8] However, immunohistochemical studies have failed to detect endothelial markers in the sarcomatous component.[11,12,13] Some studies have shown expression of monohistiocytic markers, suggesting that gliosarcomas develop from histiocytes, whereas others suggest an origin from fibroblasts, pluripotent mesenchymal cells of the perivascular adventitia, or perivascular spaces.[12,14]

The presence of identical p53 mutations and similar

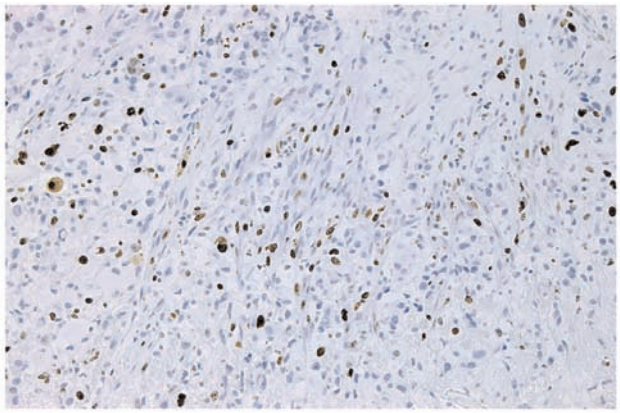

Figure 6 *MIB-1 staining showing high mitotic activity*

chromosomal imbalances and cytogenetic alterations in both gliomatous and sarcomatous components strongly supports the concept of a monoclonal origin of gliosarcomas.[5,7,10,15]

The treatment of gliosarcomas is almost similar to glioblastomas, consisting of surgical resection and, depending on clinical status, radiotherapy and/or chemotherapy.[1,4,11,16,17] Despite aggressive management, prognosis of gliosarcomas remains poor. However, a multidisciplinary approach (surgery, radiotherapy and chemotherapy) seems to be associated with slightly prolonged survival times.[16,17]

REFERENCES

1. Sade B, Prayson RA, Lee JH. Gliosarcoma with infratemporal fossa extension. Case report. *J Neurosurg* 2006 ;**105(6)**:904-7.
2. Beaumont TL, Kupsky WJ, Barger GR, Sloan AE. Gliosarcoma with multiple extracranial metastases: case report and review of the literature. *J Neurooncol* 2007;**83(1)**:39-46.
3. Burger PC, Scheithauer BW. Tumors of the Central Nervous System. Washington, Armed Forces Institute of Pathology, 1994:265-66
4. Meis JM, Martz KL, Nelson JS. Mixed glioblastoma multiforme and sarcoma. A clinicopathologic study of 26 radiation therapy oncology group cases. *Cancer* 1991, **67**:2342-2349
5. Reis RM, Konu -Leblebicioglu D, Lopes JM, Kleihues P, Ohgaki H. Genetic Profile of Gliosarcomas. *Am J Pathol* 2000, **156**:425- 432.
6. Biernat W, Hegi M, Aguzzi A, Kleihues P: Gliosarcoma: Pathology and Genetics of Tumours of the Nervous System. Kleihues P, Cavenee WK, editors. Lyon, International Agency for Research on Cancer, 1997;27-28
7. Ohgaki H, Biernat W, Reis R, Hegi M, Kleihues P. Gliosarcoma. In Kleihues P, Cavenee WK, editors. Pathology and genetics of tumors of the nervous system. Lyon: IARC Press 2000:42-44.
8. Feigin IM, Allen LB, Lipkin L,Gross SW. The endothelial hyperplasia of the cerebral blood vessels and its sarcomatous transformation. *Cancer* 1958;**11**:264-277.
9. Kleihues P, Burger PC, Scheithauer BW: The new WHO classification of brain tumours. *Brain Pathol* 1993, **3**:255-268
10. Biernat W, Aguzzi A, Sure U, Grant JW, Kleihues P, Hegi ME. Identical mutations of the p53 tumor supressor gene in the gliomatous and the sarcomatous components suggests a common origin from glial cells. *J Neuropathol Exp Neurol* 1995;**54**:651-656.
11. Perry JR, Ang LC, Bilbao JM, Muller PJ. Clinicopathologic features of primary and postradiation cerebral gliosarcoma. *Cancer* 1995;**75**:2910-2918.
12. Grant JW, Steart PV, Aguzzi A, Jones DB, Gallagher PJ: Gliosarcoma: an immunohistochemical study. *Acta Neuropathol (Berl)* 1989, **79**: 305-309
13. Sreenan JJ, Prayson RA. Gliosarcoma: a study of 13 tumors, including p53 and CD34 immunohistochemistry. *Arch Pathol Lab Med* 1997; **121**:129-133.
14. Kochi N, Budka H: Contribution of histiocytic cells to sarcomatous development of the gliosarcoma. An immunohistochemical study. *Acta Neuropathol* (Berl) 1987, **73**:124 -130
15. Machuca TN, Prevedello DM, Pope LZB, Haratz SS, Araújo JC, Torres LFB. Gliosarcoma: Report of four cases with immunohistochemical findings. *Arq Neuropsiquiatr* 2004;**62(3-A)**:608-612
16. Pakos EE, Goussia AC, Pitouli EJ, Tsekeris PG. Primary cerebral gliosarcoma: a case presentation with review of the literature. *J BUON*. 2004;**9(4)**:481-4.
17. Salvati M, Caroli E, Raco A, Giangaspero F, Delfini R, Ferrante L. Gliosarcomas: analysis of 11 cases do two subtypes exist? *J Neurooncol* 2005; **74(1)**:59-63