

## GLIOSARCOMA WITH FEATURES OF A MALIGNANT FIBROUS HISTIOCYTOMA

(Received 23 July, 1991)

**A. Sav, M.D. \*\*/ F. Söylemezoğlu, M.D. \*\*\*\*\*/ S. Küllü, M.D. \*\*/ N. Pamir, M.D. \*  
F. Özer, M.D. \*\*\*/ M. Özek, M.D. \*\*\*\***

- \* Professor, Department of Neurosurgery, Faculty of Medicine, Marmara University, İstanbul, Türkiye.
- \*\* Associate Professor, Department of Pathology, Faculty of Medicine, Marmara University, İstanbul, Türkiye.
- \*\*\* Associate Professor, Department of Neurosurgery, Faculty of Medicine, Marmara University, İstanbul, Türkiye.
- \*\*\*\* Assistant Professor, Department of Neurosurgery, Faculty of Medicine, Marmara University, İstanbul, Türkiye.
- \*\*\*\*\* Research Assistant, Department of Pathology, Faculty of Medicine, Marmara University, İstanbul, Türkiye.

### SUMMARY

A rare case of gliosarcoma of an eleven-year-old girl is reported. Histologically, the tumor resembled that of a malignant fibrous histiocytoma (MFH). The monomorphic gliosarcomas (GS) may cause a diagnostic problem. In this case, the importance of using an immunoperoxidase preparation against the glial fibrillary acidic protein (GFAP) in diagnosing gliosarcoma is emphasized.

**Key Words :** Glial fibrillary acidic protein (GFAP), Gliosarcoma, Malignant fibrous histiocytoma

### INTRODUCTION

Gliosarcomas are relatively rare intracranial tumors that contain both neuroectodermal and mesenchymal elements (1-4). Gliosarcoma constitutes 1.7-2.3% of gliomas and 5% of astrocytomas (1,3,5,6). The histogenesis of gliosarcoma is still controversial (1,3,4). The cases with dominant sarcomatous component are reported, and called sarcogliomas (1). We report a case of gliosarcoma, histologically resembling MFH, which has been reported previously (2).

### CASE REPORT

An eleven, year-old-girl was admitted to the Department of Neurosurgery, Marmara University, School of Medicine, with the complaints of headache, vomiting, and epileptic seizures for 5-6 months. Nothing abnormal was detected on the neurological examination. A computed tomography scan showed a contrast-enhancing, space-occupying cystic lesion with a mural nodule in the right temporal region (Fig.1). During operation, a

firm subcortical tumor with a cystic component was totally excised. Tumor had no dural attachment.

The gross specimen consisted fragments of highly vascular grayish tissue with patchy yellowish areas. Histologic sections showed a monomorphic spindle cell sarcoma, with fascicles of pleomorphic elongated fibroblast-like cells intermingled with each other. Foci of necrosis and storiform pattern were identified (Fig.2). Typical and atypical mitoses were present. Multi and mononucleated cells with granular, foamy eosinophilic cytoplasm were also seen. Vascular endothelial proliferation was not seen. A biphasic pattern demarcating glial and mesenchymal components was absent.

The Gomori's reticulin stain showed an abundant pericellular reticulogenesis within the tumor. The Masson trichrome and van Gieson stains demonstrated presence of exclusively perivascular collagen fibers. The phosphotungstic acid hematoxylin (PTAH) stain showed neurofibrillogenesis within the tumor. There were scattered tumor cells with irregular nuclei stained positively for glial fibrillary acidic protein (GFAP) (Fig.3).

Oil red O (ORO) stain demonstrated intracytoplasmic granular staining in some of the tumor cells.

### DISCUSSION

Gliosarcomas are rare neoplasms which comprise 1.7-2.3% of all gliomas. They are composed of neoplastic glial and mesenchymal elements (1,3,4). They have a predilection for the temporal lobes with dural attachment (1,2). Although various CT findings of cerebral gliosarcoma have been described, more common pat-

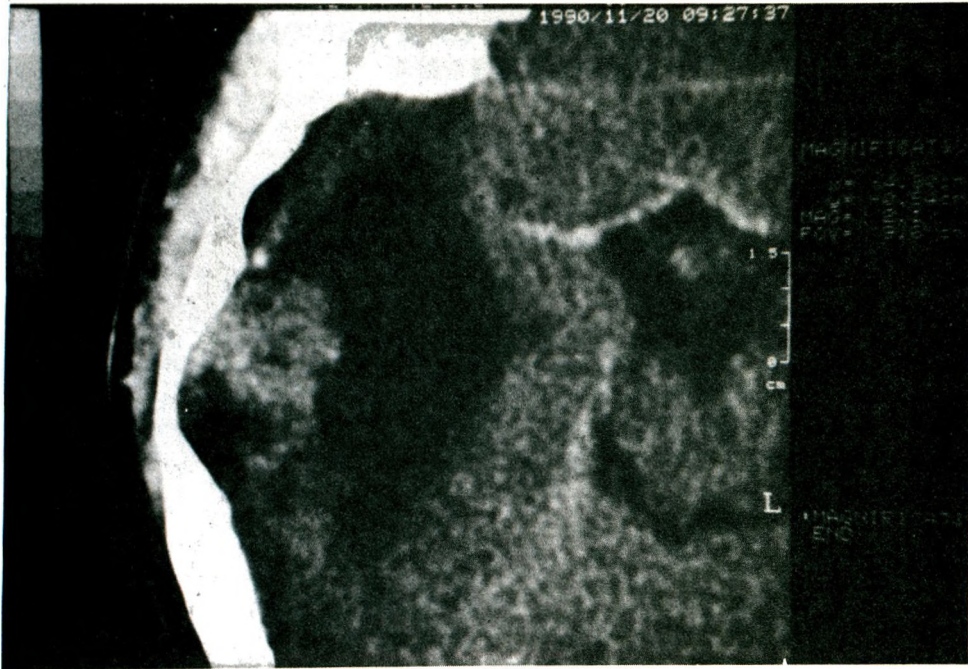


Fig. 1: CT scan showed a contrast-enhancing, space-occupying cystic lesion with a mural nodule in the right temporal region.

terns were hyperdense, well-defined mass mimicking a meningioma or a diffusely infiltrating tumor with hyper and hypodense enhancement suggesting glioblastoma (5). A case of gliosarcoma, affecting a child, that appeared as a large cystic mass on CT scan, just like in our case has also been described (7).

Although the histogenesis of GS is still unclear, there are two different hypotheses about the origin of the sarcomatous component of the tumor (1,3,4). Some authors agreed that sarcoma was secondary to glioma, and the origin of the sarcomatous component was neoplastic proliferation of vascular endothelial compartment (1,4). They suggested the presence of positive immunoreactivity against Ulex europaeus-I-lectin agglutinin (UEA I) and factor VIII related Ag (FVIII/RAg) within the sarcomatous compartment of the GS as an indirect biochemical indicator of mesenchymal origin of this curious entity (1,4). The presence of numerous Weibel-Palade-like bodies within some tumor cells was also demonstrated, that also could be regarded as the vascular origin of the tumor (4). On the other hand, some authors found that sarcoma cells of the GS to be negative for UEA I and FVIII/RAg and positive for fibronectin as well as for monohistiocytic markers such as lysozyme, x-1-antichymotrypsin, x-1-antitrypsin, and MAC 387, con-

sequently (2,4,6). Slowick et al suggested that the sarcomatous component of GS were of fibrohistiocytic origin (4).

The monomorphic GS may cause a diagnostic problem. The immunoreactivity against GFAP and reticulin stain could be of helpful tools in differential diagnosis. By the presence of GFAP positivity within the tumor, it is possible to differentiate monomorphic GS with MFH-like features from primary and metastatic MFH, and from monstrocellular sarcoma (1,2). The abundant pericellular reticulogenesis is absent in the lipidized form of glioblastoma multiforme (1). The presence of typical and/or atypical mitosis, and necrosis help to differentiate the tumor from pleomorphic xanthoastrocytoma which is common at that location and age group (1).

To keep in mind that a sarcoma of the brain is a gliosarcoma until proven otherwise. A meticulous emphasis should be given to differentiate this curious entity from other neoplastic lesions, because due to their histogenetic origin and biological behavior these tumors should be treated completely with different therapeutic modalities and so the life expectancy of patients would differ accordingly.

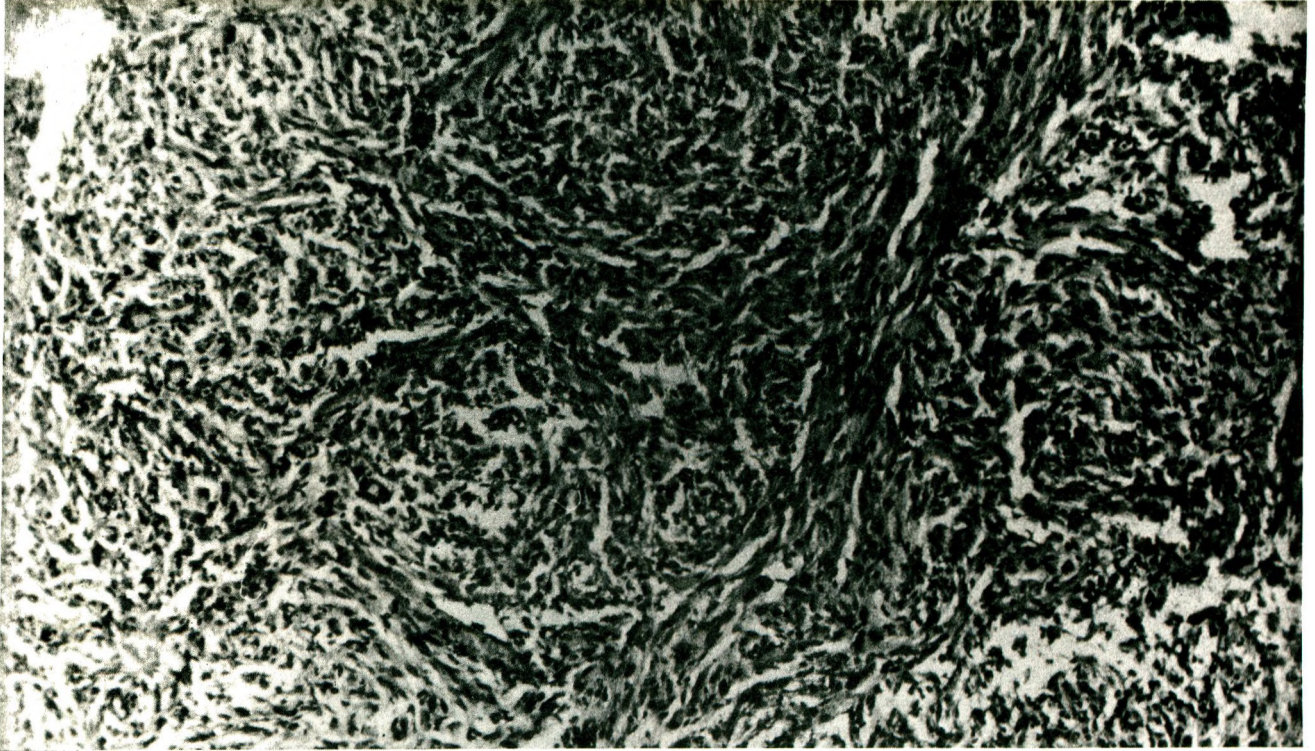


Fig. 2: Bundles of spindle-shaped cells arranged in a storiform pattern (H & E, original magnification x 100).

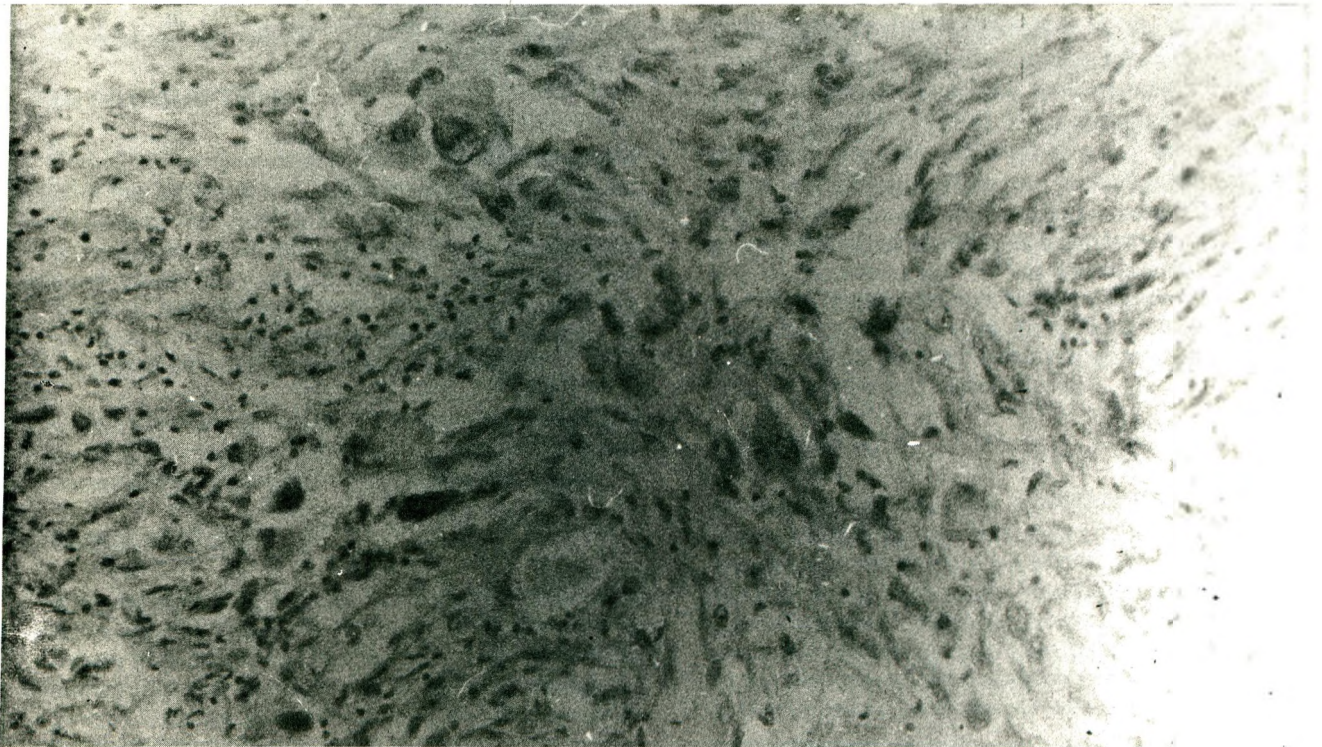


Fig. 3: Positive staining for GFAP is seen within the cytoplasm of some tumor cells (PAP method, original magnification x 200).

**REFERENCES**

1. Russell DS, Rubinstein LJ. Tumours of the Nervous System. In: Russell DS, Rubinstein LJ, eds. *Pathology of Tumours of the Nervous System*. London: Edward Arnold, 1989:83-350.
2. Ng HK, Poon WS. Gliosarcoma of the posterior fossa with features of a malignant fibrous histiocytoma. *Cancer* 1990; 65:1161-1166.
3. Grant JW, Steart PV, Aguzzi A, Jones DB, Gallagher PJ. Gliosarcoma: An immunohistochemical study. *Acta Neuropathol (Berl)* 1989; 79:305-309.
4. Slowik F, Jellinger K, Gaszo L, Fischer J. Gliosarcoma: histological, immunohistochemical, ultrastructural, and tissue culture studies. *Acta Neuropathol (Berl)* 1985; 67:201-210.
5. Maiuri F, Stella L, Benvenuti D, Giamundo A, Pettinato G. Cerebral Gliosarcomas: Correlation of computed tomographic findings, surgical aspect, pathological features, and prognosis. *Neurosurgery* 1990;26(2):261-267.
6. Banerjee AK, Sharma BS, Kak VK, Ghatak NR. Gliosarcoma with cartilage formation. *Cancer* 1989;63:518-523.
7. Goldstein SJ, Young B, Markesbery WR. Congenital malignant gliosarcoma. *Am J Neuroradiol* 1981;2:475-476.