Case Report

A Rare Brain Tumour-Gliosarcoma

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Abstract

A 57-year-old gentleman presented with a history of leftsided weakness and loss of appetite. Brain MRI was suggestive of right frontal and thalamic mass lesion with contrast enhancement at the periphery. Open biopsy examination revealed a malignant brain tumour presenting a biphasic tissue pattern with gliomatous and mesenchymal components suggestive of gliosarcoma. 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.

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Introduction

Gliosarcoma is a rare primary malignancy of the central nervous system, classified by the World Health Organization (WHO) as a high-grade glioma and a variant of glioblastoma multiforme¹. Gliosarcoma accounts for <2% of all gliomas and 5% of all astrocytomas. The tumour has a clinical presentation, natural history and radiological profile similar to glioblastoma multiforme.

Case Report

57 years old right handed gentleman admitted in Apollo Hospital Dhaka with the complains of difficulty in moving left lower limb for 1 month and loss of appetite for 2 months duration. He gave the history of fall down 2 months back. He was hypertensive, but non diabetic. No history of loss of consciousness or convulsion. At admission, his GCS scores were E4V5M6, muscle power was 3/5 on left side, 5/5 on right side. Investigations at admission were normal for blood CBC, S. Creatinine, S. electrolytes, liver function tests and lipid profile. He underwent MRI of brain which revealed 3.3 x 3.1 x 3.9 cm well defined lesion in right thalamic region with mild perifocal edema. The mass is hypointense on T1 WI and hyperintense on T2 WI with hypointense rim (fig I a,b). After contrast, which showed irregular rim enhancement and an enhancing thin septation (fig I c,d). The mass causes mass effect as evidenced by compression on body of right lateral and 3rd ventricles wit mild obstructive triventriculomegaly.



Figure- I: (a) T1 WI Saggital (b) T2 WI Axial and (c) FLAIR Axial and (d) T1 Coronal post contrast sequences showing two similar rounded lesion in right thalamic region and frontal region. The mass is hypointense on T1WI and hyperintense on T2WI and FLAIR sequence. After contrast, it shows rim enhancement and enhancing thin septa within.

Another smaller similar lesion was noted in right frontal lobe. In DW and ADC, cystic area was similar to CSF signal intensity, which represent its malignant behavior (fig II-a,b).

The conclusion of MRI examination was drawn as multi focal gliomas, however, other possibility of metastases could not be ruled out. Patient underwent CT scan of abdomen to search for any primary cause or metastases, but revealed no abnormality. His chest X ray was unremarkable. PSA was within normal limit (1.3 ng/ml). For uncertain diagnostic dilemma, patient underwent right fronto-parietal craniotomy and open biopsy of the right thalamic lesion was taken under GA. The material was grey brown. At biopsy, the section showed proliferation of anaplastic tumor cells displaying gliomatous as well as sarcomatous differentiation along with transition area. The spindle cells were forming fascicles and at places adenoid arrangement. The cells showed hyperchromatic nuclei with frequent mitoses and focal areas of necrosis. Gliosarcoma, WHO grade - IV, was diagnosed. The patient was discharged 10th postoperative day with hemodynamic stable condition and advised to consult with Radiotherapist as patient needs radiotherapy as no radiotherapic facility was available in our hospital.



Figure-II: (a) DW and (b) ADC sequences showing cystic area having similar signal intensity as CSF.

Discussion

According to the new WHO classification, gliosarcoma is defined as a glioblastoma variant, characterised by a biphasic tissue pattern with alternating areas displaying glial and mesenchymal differentiation¹. Gliosarcoma is a relatively rare malignant neoplasm accounting for approximately 2% of glioblastomas². Gliosarcoma occurs most commonly in adults in the fourth to sixth decades of life, and men are more commonly affected than women (ratio 1.8:1). Anatomically, gliosarcomas are usually located in the cerebral cortex and involve the temporal, frontal, parietal, and occipital lobes in decreasing frequency³. The presenting symptoms depend on the location of the tumour. The aetiology of gliosarcoma remains uncertain, although it is recognised that gliomas can induce sarcomatous transformation in the supporting mesenchymal elements and irradiation of the central nervous system can induce malignant transformation of the brain parenchyma and the meninges, predominantly to fibrosarcoma⁴. The appearance of gross gliosarcoma may be a poorly delineated peripheral grevish tumour mass, with central yellowish necrosis stippled with red and brown from haemorrhage, and with the sarcomatous component producing а firm discrete mass. Histologically, the diagnosis of gliosarcoma is based on a biphasic tissue pattern comprising 2 distinct malignant cell populations, one component being gliomatous (fulfilling the criteria for glioblastoma) and the other with malignant mesenchymal differentiation (fulfilling the criteria for sarcoma). The glioblastoma part of the tumour forms heterogeneous infiltrative areas with haemorrhage and necrosis. The sarcomatous portion produces a firm discrete mass. At microscopy, the glial portions show the typical features of glioblastoma multiforme. The sarcomatous areas can resemble either a fibrosarcoma, with bundles of spindle cells in a herringbone pattern, or a malignant fibrous histiocytoma, with giant cells. Other mesenchymal tissues that can be seen are cartilage, bone, and smooth and striated

muscle. Epithelial differentiation has also been noted. The sarcomatous portion demonstrates reticulin. The glial portion stains for GFAP. Many gliosarcomas stain for factor VIII-related antigen and ulex europaeus. Interestingly, the sarcomatous component can have varied histological features, ranging from the herring bone pattern of fibrosarcoma to the malignant bone of an osteosarcoma and cartilaginous differentiate on of chondrosarcoma⁵. Single institutional data from Germany published in 2009 revealed only 16 patients treated over a period of 10 years(1997-2006). ⁶Another audit from an institute of India published in 2008 presents only 24 patients of gliosarcoma as compared to 251 cases of GBM treated over a period of 15 years (1990-2004). Local data revealed 16 cases of gliosarcoma reported from pathology department of Aga Khan University hospital in 2004⁸. Recently another case has been reported in local journal of Pakistan, there are fewer than 20 reported cases of extracranial metastases of gliosarcoma with majority of them reflecting a tendency for haematogenous dissemination⁹.

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