Gliomatosis cerebri - A case report
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Abstract: Gliomatosis cerebri is a rare malignant astrocytic tumor characterized by diffuse, neoplastic glial cell infiltration of the brain involving more than 2 cerebral lobes. We report a case of Gliomatosis cerebri in a 20 year old young male patient who was evaluated for complaints of headache and double vision. He was treated with Radiotherapy and 6 cycles of adjuvant temozolamide. Patient is alive and disease free at 1 year of follow up. This case is being presented on account of its rarity.

Keyword: glial tumors, gliomatosis cerebri

INTRODUCTION
The term Gliomatosis Cerebri (GC) was first proposed by Nevin in 1938[7]. Just over 300 cases of gliomatosis cerebri have been reported in the medical literature since 1938[3]. Gliomatosis cerebri, otherwise known as diffuse infiltrative astrocytosis is a rare form of malignant astrocytic tumor which results from widespread infiltration of neoplastic astrocytes involving more than two cerebral lobes. It often extends bilaterally and into infratentorial structures. It corresponds to WHO grade III astrocytoma. The brain architecture is commonly preserved and the mass effect is minimal [5,6]. It can occur at any age, ranging from neonates to elderly, most common being third to fifth decade [1]. There is no sex predilection. The tumor may appear de novo (primary GC) or result from the spreading of a focal glioma (secondary GC). Patients commonly present with headache, seizures, cranial nerve deficits, papilledema, corticospinal tract deficits, increased intracranial tension and spinocerebellar deficits.

CASE REPORT
20 year old male patient was evaluated for complaints of headache and double vision for 3 months duration. There was no focal neurologic deficit on examination. MRI brain showed large areas of altered signal intensity in right frontal, temporal lobes and capsuloganglionic area, hyperintensity in bilateral hippocampal region, left frontal lobe, white matter genu of corpus callosum, septum pellucidum, right cerebral peduncle, right corona radiata, left medial temporal region, left insular region and white matter tracts of brain on right side. MRS showed elevated choline and decreased NAA (N Acetyl Aspartate) in some of the voxels on right frontal lobe and capsuloganglionic region. Lactate peak was seen in few voxels. Possibility of gliomatosis cerebri was suggested. Biopsy was performed from the frontal lesion, histopathologic analysis of which showed increased cellularity and atypia, findings consistent with grade III glioma. Together with the imaging findings, the diagnosis of gliomatosis cerebri was made.

Fig1. Histological picture showing predominantly glial tissue with focal areas showing hypercellularity and atypia

Fig2. T2 weighted MRI showing altered signal intensity lesions in right frontal and temporal lobes

Fig3. T2 weighted MRI after 60gy RT and 4 cycles Temozolomide showing excellent response

Patient was treated with focal radiotherapy by right and left opposing lateral portals to a total dose of 60 gy followed by 6 cycles of adjuvant temozolomide 250mg/m2. Patient is alive and symptom free at 1 year of follow up.

DISCUSSION
Diagnosis and management of GC are difficult [2]. The continuity of cellular infiltration and a lack of clear distinction from adjacent normal brain tissue helps to distinguish gliomatosis cerebri from multifocal gliomas [1]. Its cell of origin and pathogenesis remain unclear, probably due in part to its rarity. There are two types of Gliomatosis cerebri. Type I is the classical form which is characterized by diffuse infiltration of neoplastic cells without a focal mass. Type I may give rise to Type II where in addition to diffuse infiltration there is a focal mass which is usually a high
Grade glioma. According to autopsy studies, the areas that are invaded include (in decreasing order of frequency) the cerebral hemispheres, midbrain, pons, thalamus, basal ganglia, cerebellum, medulla oblongata, and, in less than 10% of cases each, the hypothalamus, optic nerve and chiasm, and spinal cord [4]. The corpus callosum may also be involved in up to 50% of cases [1].

WHO 2007 has included GC as a distinct nosological entity among neuroepithelial tumors. Histologic analysis shows elongated glial cells that typically resemble astrocytes [8]. Infiltrating cells often form parallel rows among white matter tracts. GFAP (Gliaal Fibrillary Acidic Protein) and S-100 are occasionally expressed. The histologic features of gliomatosis cerebri show variations from patient to patient and even within the same lesion [9]. Features of high grade glioma like microvascular proliferation and necrosis are not seen [10]. The largest case series was by Taillibert et al. who analyzed 90 cases from the French neuro-oncology network between 1993 and 2004 and more than 206 cases described in the literature between 1938 and 2004 [11]. According to this series, the day 1; and vincristine (1.4 mg/m2 on days 8 and 29) or temozolomide (TMZ; 150 to 200 mg/m2 for 5 days every 4 weeks) has been tried and has been found to be useful in some patients with GC. Temozolomide is well tolerated and seems to be a good substitute for procarbazine – lomustine (CCNU) – vincristine, mainly for cases of slow growth and low grade gliomatosis cerebri [18].

**REFERENCES:**
