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# Gliomatosis Cerebri: a rare Malignant Tumour of Brain

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## Abstract

Gliomatosis cerebri is a rare form of brain glioma with incidence of less than 1% of total brain tumors .Gliomatosis cerebri is an infiltrative astrocytic tumor which has acquired the status of a distinct clinical, radiological and pathological entity where the diffuse neoplasm involves two or more lobes of cerebrum.

**Key Words:** Brian ,gliomatosis cerebri,MRI.

## INTRODUCTION

Gliomatosis cerebri is a rare form of brain glioma with incidence of less than 1% of total brain tumors.[1-3] Nevin et al described the entity way back in 1938. Till date, not more than 100 to 120 cases have been reported in the literature.[4-6] Gliomatosis cerebri has been given the status of a clinic pathological entity by the WHO classification of brain tumors and diffuse infiltrative tumor process which involves more then 2 lobes of the brain by malignant glioma (high or low) grade that permeates the brain extensively without destroying neuro architecture.[7-8] We report such a rare of case.

## CASE PRESENTATION

A 20-year old young female, student, presented with history of progressively increasing sub occipital headache, vomiting, blurring of vision, and diplopia of 15 days duration. No history of loss of consciousness, seizure, motor and sensory complaints. On clinical examination, she had right sided upper motor neuron type of hemi paresis with left rectus muscle palsy and right sided upper motor neuron type of facial palsy. Visual acuity was 6/18 in right eye and 6/12 in left eye and had bilateral papilloedema. Field of vision remained normal in both the sides. Other neurological functions were normal.

On investigation MRI showed ill - defined heterogeneously enhancing lesion involving the corpus callous and medial frontal lobes, more on the left side. Grey and white matter identification was lost with minimal mass effect on part of the body of left lateral ventricle and on its left frontal horns. Stereo tactic biopsy was deferred as there was no enhancing area and the chances of negative sampling on biopsy were high. She underwent bilateral frontal craniotomy and right frontal lobotomy and decompression of the tumor. Histopathological examination showed gliomatosis cerebri, WHO grade III (Photo 1). Postoperative neurological status was the same as in preoperative condition with recent and past memory loss but her symptoms of headache had improved.

Four weeks after surgery, she was planned and received adjuvant chemo radiotherapy. External radiation to the whole brain on Telecobalt machine with right lateral and left lateral field with Boost to a total dose of 60 Gray in 30 fractions, 5fractions per week along with Radiation sensitizer Temozolomide 100mg per day as a radio sensitizer on the days of Radiotherapy. The patient tolerated Radiotherapy well. 6 months post radiotherapy the patient is doing well with improved neurological functions and returned back to her studies.

## DISCUSSION

Gliomatosis cerebri is a relatively rare primary malignant brain tumor consisting of less than 1% of total brain tumor & and is the first case reported from this institute. Previously referred to an infiltrative glial neoplasm that causes diffuse overgrowth of the affected areas of the brain with preservation of underlying structures.[3]

WHO classification of brain tumors recognizes this entity among neuroepithelial tumors of uncertain origin.[7,8] The WHO working group considered gliomatosis is appropriate when involvement of 2 lobes of brain by small elongated cells without cellular centrally necrotic center with overall preservation of underlying brain architecture. Microscopically diffusely infiltration of astrocytic cells with varying degree of differentiation is typical.

It differs from multicentric glioma in that gliomatosis involves contiguous areas whereas in multicentric glioma tumor masses occur in different sites, multicentric more often seen with GBM.[9]

The clinical diagnosis is relatively difficult due to variable clinical feature; it has known to occur in any age group more common 2nd to 3rd decade. Headache and blurring of vision and diplopia, some times seizure are most common initial symptoms at presentation.[2,4]

Before the era of MRI diagnosis was confirmed by autopsy, the advanced neuroimaging made it possible to diagnose

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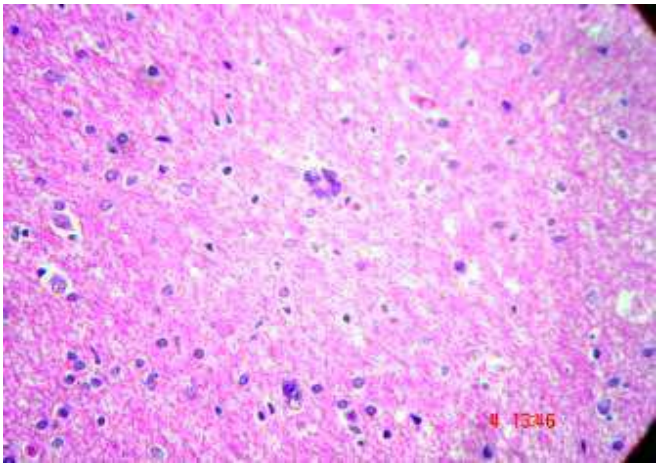

**Figure1 : Histopathological examination showed gliomatosis cerebri**

the disease ante mortem. T2-weighted MRI image considered the gold standard imaging technique, MRI images showed ill-defined heterogeneously enhancing lesion of 2 lobes with no contrast enhancement and with minimal mass effect & poor demarcation of gray &white matter. MRI FLAIR images will have better definition of lesion extent. Even MRI tends to underestimate true extent of tumor invasiveness.[10] Diagnosis of gliomatosis requires radiological- pathologic correlation, Brain biopsy & histopathological examination are mandatory.

The Brain Radiotherapy improved survival in Gliomatosis than without radiotherapy. Earlier study showed improved median survival of 11.4 months from the time of biopsy with radiotherapy compared to without radiotherapy.[11] The patient may live for years from diagnosis without RT or any cancer treatment, but the outcome for patients with radiotherapy is relatively better than the outcome of patient who receives no treatment at all.

Chemotherapy with temozolamide combined with RT could improve the treatment outcome, the earlier study showed promising results in the treatment of gliomatosis with temozolamide.[12]

## CONCLUSION

Gliomatosis cerebri is an infiltrative astrocytic tumor which has acquired the status of a distinct clinical, radiological and pathological entity where the diffuse neoplasm involves two or more lobes of cerebrum. With advances in brain imaging and molecular techniques, more data regarding the natural history of this rare disease will be accumulated, enabling us to design better therapeutic strategies to improve treatment outcome. The near whole brain Radiotherapy appears to have survival and performance benefit for a patient with low grade gliomatosis cerebri use of radiation sensitizer like temozolomide could improve treatment outcome.

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