International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Paediatric Giant Cell Glioblastoma: A Rare Case Study and Review of Treatment Strategies

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Abstract: ***This paper presents a rare case of Giant Cell Glioblastoma GCG, a distinct variant of classical Glioblastoma Multiforme GBM, in a 15-year-old adolescent. GCG, characterized by a predominance of bizarre, multi-nucleated giant cells, represents 5 of all glioblastomas, with an even lower incidence in pediatric patients. Despite a generally poor prognosis, this variant is associated with longer survival in both adults and children. The patient, presenting with holocranial headaches, was diagnosed with a High Grade Glioma in the left fronto parietal region. Post-operative histopathological examination confirmed a high-grade GCG, WHO Grade IV. The patient underwent adjuvant radiotherapy and chemotherapy with Temozolomide, showing no signs of recurrence or toxicities after 12 months. This case study emphasizes the importance of histopathological confirmation and the role of radiotherapy followed by chemotherapy for progression and recurrence-free survival in GCG.***

Keywords: Giant Cell Glioblastoma (GCG), Glioblastoma multiforme (GBM), Paediatric, Prognosis

1. Introduction

WHO recognises Giant Cell Glioblastoma (GCG) as a distinct variant of classical GBM. It is characterized by a predominance of bizarre, multi-nucleated giant cells, an occasionally abundant reticulin network, and a high frequency of TP53 mutations. It represents 5% of all glioblastomas and incidence in adults is 0.8% of all brain tumors and even less in pediatric patients. Clinically, despite a poor prognosis for GCG in most reports, this variant is associated with a longer survival compared with GBM in both adults and children.

2. Differential Diagnosis

On imaging the differential is primarily:

- Conventional Glioblastoma: usually indistinguishable
- Cerebral metastasis
- Meningioma: if abutting the dura

Histologically,Pleomorphic Xanthoastrocytoma(PXA) is one important differential diagnosis as both have common the presence of giant tumor cells, infiltration of lymphocytes, deposition of reticulin and circumscription.Neuronal markers Positive in PXAs are negative in giant cell glioblastomas.

3. Case Report

A 15yr old adolescent presented with a history of holocranial headache,not relieved with analgesics. MRI Brain showed Grade II/III Glioma in the left fronto parietal region measuring 3 x 2 cm. Left Parietal Craniotomy and Excision of Space of occupying lesion was done uneventfully.

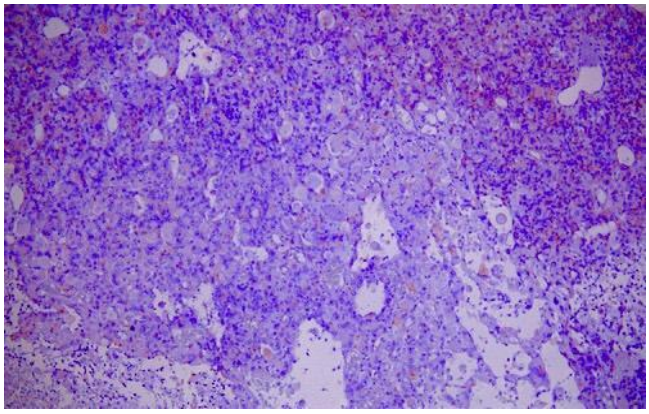
Post operative HPE: HIGH GRADE GLIOMA -GIANT CELL GLIOBLASTOMA, WHO GRADE IV.

Post operative MRI: Craniotomy defect noted in left parietal region measuring 26x37x22mm cystic areas noted in Left parietal lobe with peripheral T2 hypo intense areas showing blooming on GRE with peripheral rim enhancement & T2 white matter edema.

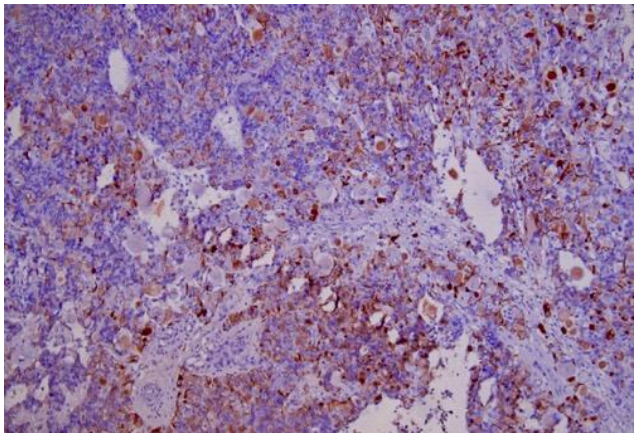
Patient was referred to MNJIO&RCC for further treatment, where we planned for ADJUVANT RADIOTHERAPY a total dose of 60GY IN 30 FRACTIONS AT 2.0GY PER FRACTION WITH CONCOMITANT TEMOZOLOMIDE 75mg per square meter(100mg/day on days of RT), followed by ADJUVANT TEMOZOLOMIDE 150mg per square meter (200 mg/day for 5 days every 28 day cycle). Patient is on Adjuvant Temozolomide since 12 months without any signs of recurrence or toxicities of Temozolomide.

3.1 Histopathology & IHC

High Grade Glioma-Giant Cell Glioblastoma, WHO Grade IV.



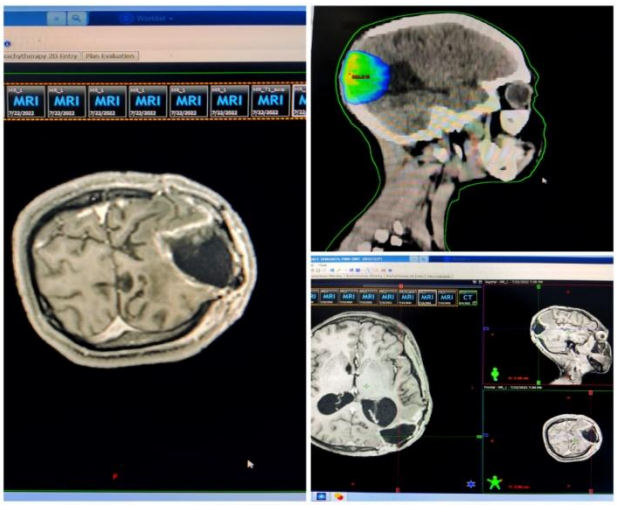
Histopathology of the excised mass showing multinucleated tumor giant cells with areas of necrosis



Immunohistochemistry of tumor: Positive for GFAP and Vimentin, negative for EMA.

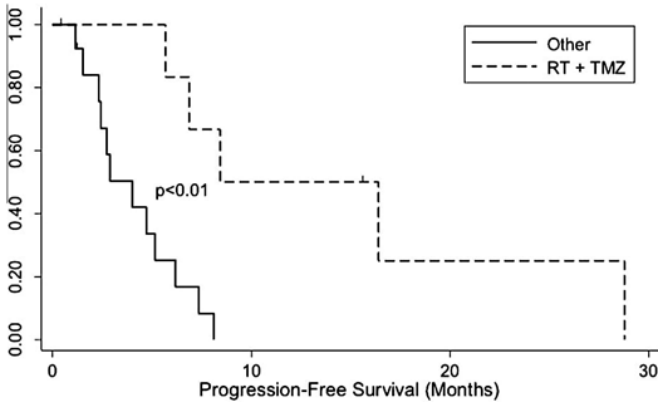
IHC:
GFAP Positive
EMA Negative
VIMENTIN Positive.

3.2 Imaging and Radiotherapy Planning

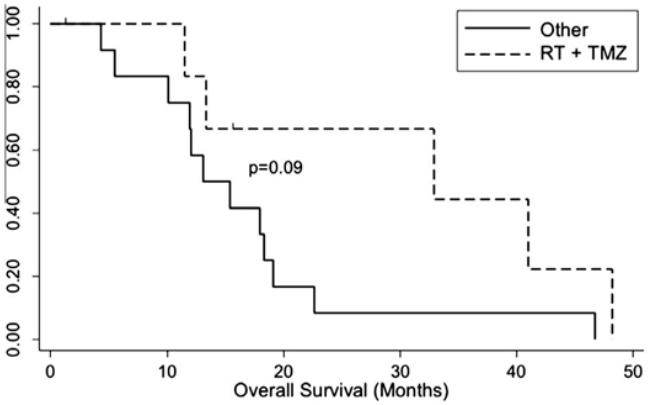


3.3 Statistical Analysis

Impact of adjuvant radiation (RT) and temozolomide (TMZ) therapy on progression-free survival (months) in giant cell glioblastoma patients. Censored patients are represented by vertical dash marks.



Impact of adjuvant radiation (RT) and temozolomide (TMZ) therapy on overall survival (months) in giant cell glioblastoma patients. Censored patients are represented by vertical dash marks.



3.4 Conclusion

GCG is very rare in the pediatric age group, but when diagnosis is confirmed, it offers better hope for such patients because they usually have a longer survival period than classic GBM patients.

Reporting of such case highlights the importance of HPE confirmation and the role of RADIOTHERAPY followed by CHEMOTHERAPY for PROGRESSION AND RECURRENCE FREE SURVIVAL

3.5 Prognostic Factors

1) Young Age
2) Tumor Size
3) Extent Of Resection
4) Radiation Therapy

Acknowledgments
I would like to acknowledge the department of Radiation Oncology, MNJIO & RCC, Osmania Medical College, Hyderabad.

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Author Profile

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