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Case Report

Pediatric giant cell glioblastoma: a rare entity

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ABSTRACT

Giant cell glioblastoma (GCG) is a rare subtype of glioblastoma multiforme (GBM) which represents 0.8% of all brain tumors and 5% of glioblastomas. This entity is all the more rarer in the pediatric age group. And, due to the limited number of case reports and case series in literature, its epidemiology, natural history and follow up are not well documented. We present this case for its rarity. A 14-year-old girl presented with history of recent onset headache and vomiting from last 10 days. Magnetic resonance imaging (MRI) brain revealed a mass lesion in the right temporo-parietal region, suggestive of a high-grade glioma. Histopathological examination revealed features typical of giant cell glioblastoma. Giant cell glioblastoma, is an extremely rare variant of gliobastoma multiforme, merits special mention because it has been hypothesized to be associated with a longer survival compared with GBM in both adults and children, possibly because of the younger age at presentation, certain histopathological and molecular characteristics of this entity.

Keywords: Giant cell glioblastoma, Long term survival, Pediatric

INTRODUCTION

Giant cell glioblastoma (GCG) is a rare subtype of glioblastoma multiforme (GBM) which represents 0.8% of all brain tumors and 5% of glioblastomas.[1]

It has been described as a histological variant of glioblastoma with predominance of bizarre multinucleated giant cells and a high frequency of TP53 mutations, corresponding to grade IV tumors in the WHO classification of CNS tumors.[2]

Due to the rarity of this entity, the epidemiology, natural history and follow up are not well documented.

It has been hypothesized that GCG has a better outcome and higher survival rates than GBM, due to several reasons, such as the age at presentation, certain histopathological and genetic differences from usual glioblastomas.[3]

CASE REPORT

A 14-year-old girl presented with history of recent onset headache and vomiting for the last 10 days. On examination, the patient did not have any focal neurological deficit. Magnetic resonance imaging (MRI) brain revealed a mass lesion in the right temporo-parietal region, hypointense on T1 and hyperintense on T2 with ring enhancement on contrast administration. It was associated with perilesional edema and mass effect. The radiological features were suggestive of high grade glioma. The patient underwent right temporo-parietal craniotomy and gross total excision of the tumor. Per operatively, the tumor was very vascular and was seen invading the dura. She had an uneventful peri-operative course and was discharged on 7th post-operative day.

Histopathological examination revealed a high-grade neoplasm predominantly composed of multinucleated giant cells with bizarre hyperchromatic nuclei (Figure 1).

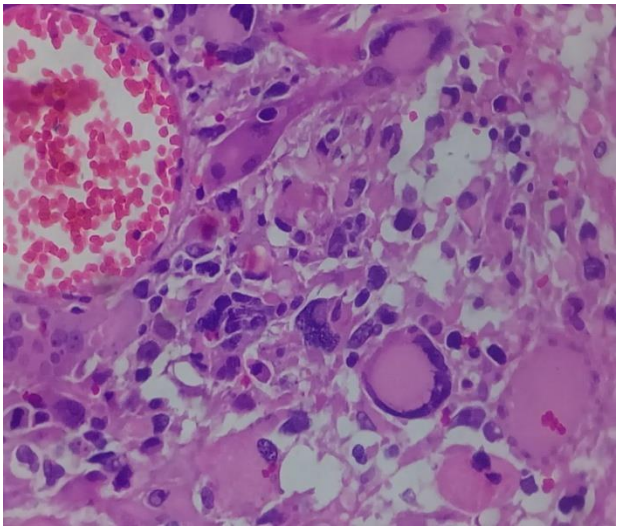

Figure 1: Neoplasm showing numerous multinucleated giant cells with bizarre nuclei (H and E, 40x).

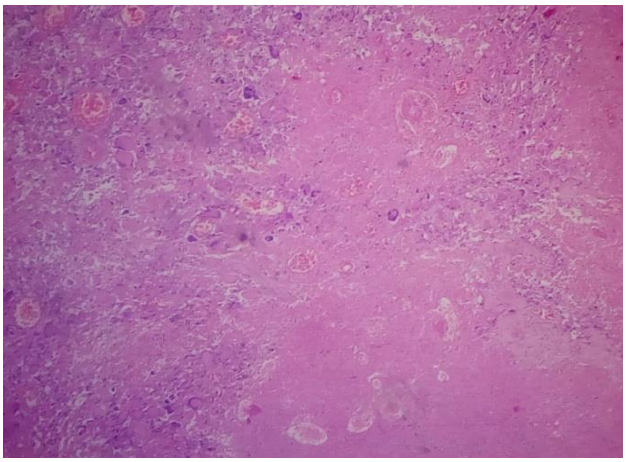

Figure 2: Areas of geographic necrosis with adjacent giant cell component. (H and E, 4X).

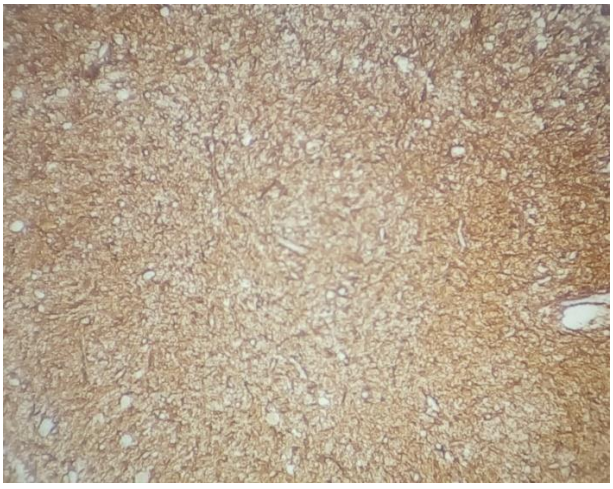

Figure 3: Immunohistochemistry with GFAP showing strong diffuse positivity (4X).

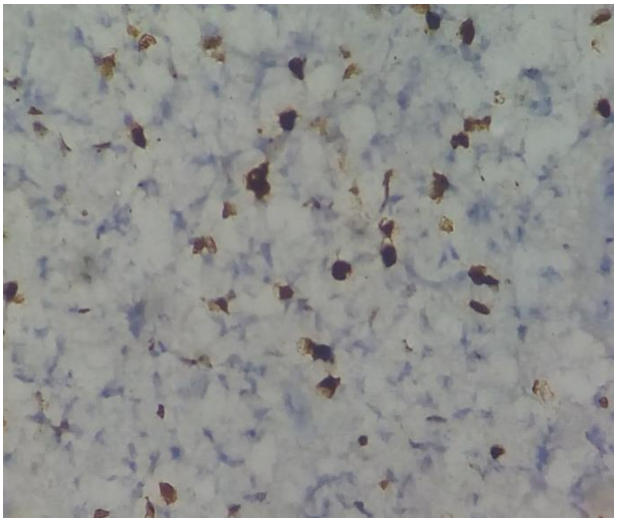

Figure 4: Ki67 showing high profileration index.

Large areas of geographic necrosis (Figure 2) and atypical mitotic figures were noted. Immunohistochemical studies showed strong diffuse positivity of GFAP (Figure 3) as well as a high proliferation index (Ki 67 - 30% - 40%) (Figure 4). A diagnosis of giant cell glioblastoma was made.

DISCUSSION

Giant cell glioblastoma, WHO grade IV, is an extremely rare variant of glioblastoma multiforme (GBM) constituting about 5% of all glioblastomas. It was previously called 'monstrocellular sarcoma' because of the prominent stromal reticulin network, but strong expression of GFAP has firmly established its astrocytic nature.[1]

The mean age at presentation was 41 years, which was 2 decades younger than the mean age of presentation of usual glioblastoma. However, a wide variation in age range has been described, including even children.[1] Some studies have documented male predominance, while others have documented equal gender predilection.[1,2] This subtype is even more rare in the pediatric age group with only 53 cases having been reported in literature.[4] Other studies have reported that GCG comprised of 3% of all cases of glioblastomas in the pediatric age group.[5]

Classically, patients presented with symptoms of raised intracranial pressure and hemiparesis diplopia, aphasia, vertigo, and seizures were also seen in a minority of cases.[2] The most common location are the frontal and temporal lobe, and rarely the multifocal.[3]

Histologically, giant cell glioblastomas are characterized by a predominance of multinucleated giant cells with abundant cytoplasm, with abundant reticulin fibers, and it was used to be called monstrocellular brain tumor.[1] Immunohistochemically, both giant cells and non-giant cells are positive for glial fibrillary acidic protein

(GFAP), S100 protein and vimentin; in giant cells nuclei are positive to proliferation markers like Ki-676. At cytogenetic and molecular analyses, microsatellite instability is more frequent in GC than GBM (30% vs 7.8%), TP53 mutation is observed in 83.3%, but epidermal growth factor receptor is uncommon (8.3%). Chromosome 10 deletion is found in all non-giant cells of the GC; at GBM it is found in 45%. GCG has more p53 mutations than GBM, but p16 deletion, MDM2 and CD4K amplifications are rare.[3]

Long term survivors (LTS) are those who remain disease free for 3 years or more after initial diagnosis. According to Shinojima et al, young age at presentation and female gender are both predictors for long-term survival in cases with GBM. The authors also found that giant cell elements were exclusively seen in long term survival cases of GBM.[11] Currently, efforts are focused on detecting molecular predictors for survival. They found that p53 mutation is associated with younger age and prolonged survival, while MDM2 amplification correlated with worse prognosis.[11] This is probably the basis for the hypotheses that GCG is associated with a long-term survival when compared with the usual GBM. However, surprisingly, in a unique series of 18 cases of pediatric GCG by Karremann M et al, the widely-acclaimed hypothesis that GCG may imply a better prognosis than GBM could not be substantiated.[2]

Thus, our case can be considered 'typical' of GCG because of the typical location, clinical presentation, radiological features and histo-morphology described in literature, except for the atypical age group, which makes our case a unique one.

CONCLUSION

Giant cell glioblastoma has been described as a histological variant of glioblastoma, WHO grade IV, with predominance of bizarre multinucleated giant cells and a high frequency of TP53 mutations. This variant merit special mention because it has been hypothesized to be associated with a longer survival compared with GBM in both adults and children, possibly because of the younger age at presentation, certain histopathological and molecular characteristics of this entity. Due to the rarity of these cases, epidemiology, natural history and follow up are not well documented, especially in pediatric cases. We present this case for its rarity.

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REFERENCES

1. Kleihues P, Burger PC, Aldape KD. Glioblastoma. In: Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, editors. WHO Classification of Tumors of the Central Nervous System. Lyon, France: IARC Press; 2007:33-49.
2. Karremann M, Butenhoff S, Rausche U, Pietsch T, Wolff JEA, Kramm CM. Pediatric giant cell glioblastoma: New insights into a rare tumor entity. Neuro Oncol. 2009;11(3):323-9.
3. Belsuzarri TAB, Araujo JFM, Catanoce AP. Giant cells glioblastoma: case report and pathological analysis from this uncommon subtype of glioma. Rare Tumors. 2015;7(1):5634.
4. Jain SK, Sundar IV, Sinha VD, Sharma V, Bhasme V, Goel RS. Giant cell glioblastoma in a child: a rare case report. Asian J Neurosurg. 2012;7(3):144-6.
5. Borkar SA, Lakshmiprasad G, Subbarao KC, Sharma MC, Mahapatra AK. Giant cell glioblastoma in the pediatric age group: Report of two cases. J Pediatr Neurosci. 2013;8(1):38-40.
6. Katoh M, Aida T, Sugimoto S. Immunohistochemical analysis of giant cell glioblastoma. Pathol Int. 1995;45:275-82.
7. Shinojima N, Kochi M, Hamada JI. The influence of sex and the presence of giant cells on postoperative long-term survival in adult patients with supratentorial glioblastoma multiforme. J Neurosurg. 2004;101:219-26.

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