**Introduction**

Pediatric Brain Tumors (BT) are the most common solid tumors in children and the second among all childhood malignancies after leukemia [1].

Glioblastoma Multiforme (GBM) or grade IV gliomas-tumors is the most frequent primary BT [2]. However, the incidence in children is very low, 0.8 per 100,000 children [3]. It is a clinically, histologically and genetically quite heterogeneous, highly malignant tumor [4].

Giant Cell Glioblastoma (GCG) as a sub type of GBM [5], is a rare variant with an incidence of 0.8% of BT and 5% of GBM. Supposedly, it has a better prognosis.

We report a case of GCG in a 12-year-old boy along with a clinicopathological and therapeutic description and literature review.

**Case presentation**

A 12-year-old male child presented with a two-month history of progressively worsening headache and vomiting. On neurological examination, the child was conscious, had left kinetic cerebellar ataxia and multiple caféau-lait spots in the trunk. A contrast-enhanced CT scan showed an intra axial mass in the left cerebellar region with significant surrounding edema causing mass effect and onset of amygdalas engagement.

To further evaluate the lesion, a Magnetic Resonance Imag-

The differential diagnosis based on imaging and clinical presentation include low-grade tumors such as ganglioglioma, oligodendroglioma and Pleomorphic Xanthoastrocytoma (PXA) [15]. According to Pant et al, the common features shared by GCG and PXA include numerous giant cells, prominent reticulin stroma, lymphocytic infiltrates and evident circumscription. Supposedly, GCG and PXA include numerous giant cells, prominent reticulin stroma, lymphocytic infiltrates and evident circumscription. 

Figure 1a: Dosimetric image showing the Planning Target Volume (PTV) coverage by radiation fields during the first series of irradiation.

Figure 1b: Dose-volume histogram showing the RT dose received by the Planning Target Volume (PTV) and the different organs at risk during the first series of irradiation.
sis, with immunopositivity for p53 and GFAP and immunonegativity for neuronal markers [14].

Treatment approaches include maximum safe resection along with adjuvant RT [6]. Use of CH has been described as well, although protocols are quite variable [14]. The standard regimen for RT and CH includes fractionated focal irradiation in daily fractions of 2 Gy given 5 days per week for 6 weeks, for a total of 60 Gy and continuous daily TMZ (75 mg per m² of body surface area per day, 7 days per week from the first to the last day of RT), followed by 6 cycles of adjuvant TMZ (150 to 200 mg per m² for 5 days during each 28-day cycle) [15]. The outcome of the GCG patients treated with the more intensive CH regimen was not analyzed [16]. To be noted that surgery by itself, may offer 32 weeks of mean survival time. RT has proven beneficial, adding 25 weeks to the total mean survival time [14].

The GCG is associated with longer survival times compared to GBM, ranging from 15 months up to 17 years in adults and from 14 months up to 12 years in children [16]. It was linked to a higher degree of complete tumor resection than in GBM patients [7] due to its predominance in locations within the cerebral hemispheres and its less infiltrative behavior such as our case [7]. Therefore, the more superficial and localized the tumor, the better the prognosis [6]. Palma et al. involved the beneficial prognostic influence of lumphocytic infiltration in malignant BT and considering that giant monstrous cells are implicated in the host’s enhanced immune response by manifying the antigenic stimulus, that would explain the slower evolution of this tumor [15]. Besides, a study of 18 pediatric patients by Karremann et al. showed no significant difference in median age, male preference, median clinical history, and prognosis between GCG and GBM [7]. Spinal metastasis from primary GBM is extremely rare and occurs relatively late [18,19]. Treatment modalities include surgery for decompression, RT in total dose of 25-40 Gy and intrathecal CH. Because of the diffuse nature of the disease, surgery is generally unsuitable. Therefore, RT is used most commonly. There is no obvious survival advantage of one therapy over the other and the prognostic is poor [18,19].

Conclusion

This case report is focused on the rarest situation of GBM subtype (GGC) which is even few reported, especially in the pediatric population.

Differentiating it from GBM on a clinical and radiological features is difficult because of similar characteristics and is entirely based on histopathological examination with immunohistochemistry study.

The case presented shows a GCG with non-specific symptoms progressed rapidly over a short time treated by complete removal followed by RT and CH. Unfortunately, the evolution in our patient with spinal leptomeningeal metastases is not habitual and the prognosis is similar to that of common GBM. Future studies with larger cohorts and molecular pathological analyses are still needed to corroborate the findings of the present case report and to establish aconsensual management of this entity.

References


