Cerebellar High-Grade Glioma In A 6-Year-Old Female Child : A Literature Review And Case Analysis

Hayet OUADI, Amira BENHAFRI, Lynda ATROUNE
Department of Neurosurgery, SHE Ali Ait Idir, Algiers, Algeria

Correspondant author: *Hayet OUADI.Department of Neurosurgery, SHE Ali Ait Idir, Algiers, Algeria*

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ABSTRACT

Background : Paediatric cerebellar glioblastoma is an exceptionally rare clinical entity, with very few cases described in the literature. These tumors have been reported with an incidence of less than 3% and are associated with poor prognosis despite aggressive treatment.
Case Presentation : We report the case of a 6-year-old female child who presented with balance and gait disturbances for one month. Clinical examination revealed cerebellar ataxia, dysmetria, dysdiadochokinesia, and left hemiparesis. Imaging showed a paramedian solid infratentorial mass lesion in the right cerebellar hemisphere, infiltrating the superior and middle cerebellar peduncles and the brainstem. The patient underwent tumor resection following ventriculoperitoneal shunt placement. Postoperative complications included intracavitary hematoma, cerebellar syndrome aggravation, and right seventh cranial nerve palsy.
Histopathological examination revealed a high-grade (4) diffuse astrocytoma (glioblastoma). Due to complications, radiotherapy was not administered, and the patient succumbed two months after surgery.
Conclusion : Pediatric cerebellar glioblastomas remain rare but highly aggressive tumors with limited treatment options and poor survival rates. Multimodal therapeutic approaches, including maximal safe resection and adjuvant therapy, are critical to improving patient outcomes. Ongoing research is needed to enhance early detection and therapeutic strategies.

Key words:*Pediatric glioblastoma; Cerebellar tumor; Posterior fossa; High-grade astrocytoma; Poor prognosis*

INTRODUCTION

Paediatric cerebellar glioblastoma is an exceptionally rare clinical entity, with very few cases described in the literature (1). They have been reported with an incidence of less than 3%. In the majority of reported cases, prognosis is extremely poor, despite surgical and oncological management (1). Here, we present a case of cerebellar high-grade glioma diagnosed in a 6-yearold female child.

CASE REPORT

Clinical Presentation : A 6-year-old female child presented with chief complaints of balance and gait disturbances since one month. On examination, cerebellar signs were positive, including cerebellar ataxia, dysmetria, and dysdiadochokinesia, associated with left hemiparesis, without cranial nerve involvement.

Imaging Findings : Preoperative magnetic resonance imaging (MRI) showed a lesion in the right cerebellar hemisphere. Contrast-enhanced MRI was suggestive of a paramedian solid infratentorial mass lesion of 4.2 x 3 x 3 cm in the right cerebellar hemisphere, infiltrating the superior and middle cerebellar peduncles and the brainstem, with surrounding edema causing a mass effect, notably on the fourth ventricle, which remained patent without signs of hydrocephalus upstream.

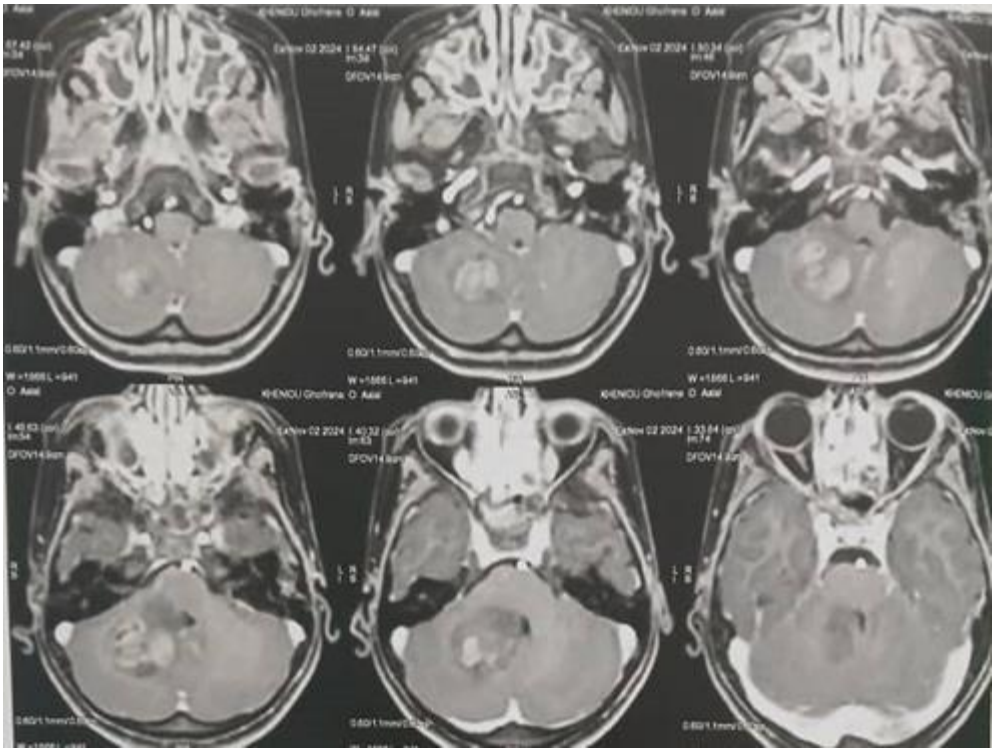

Figure 1: Pre operative magnetic resonance imaging showing a lesion in the right cerebellar hemisphere

Surgical and Postoperative Course : The patient was admitted to the hospital and initiated on corticosteroid therapy while awaiting surgical intervention. On the following day, she developed signs of elevated intracranial pressure (ICP), and a brain CT scan demonstrated dilation of the ventricular system. Consequently, a ventriculoperitoneal shunt was placed. The patient subsequently underwent tumor resection. Postoperative complications included an intracavitary hematoma requiring surgical revision, along with an aggravation of cerebellar syndrome and a seventh right cranial nerve palsy.

Histopathology: Histopathological examination revealed a high-grade (4) diffuse astrocytoma (glioblastoma) according to the 2021 WHO classification. Immunohistochemical stains were positive for GFAP, Olig2, and P53.

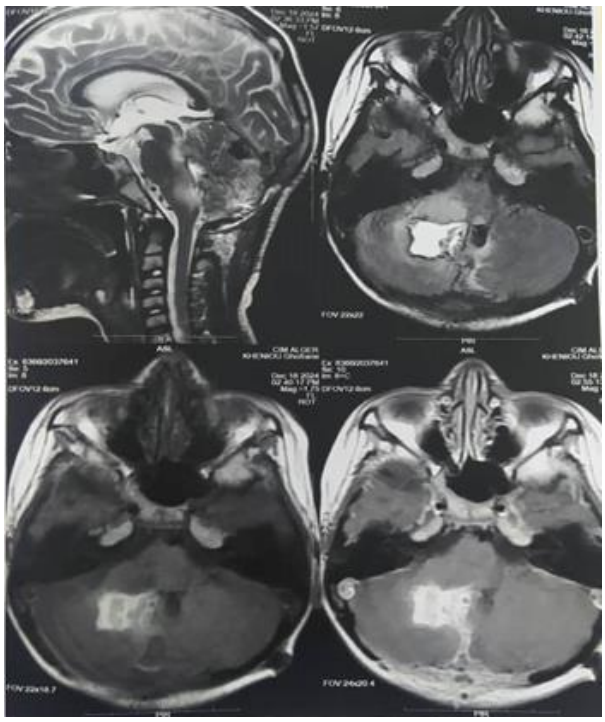

Figure 2 : Post operative magnetic resonnance imaging showing an intracavity hematoma with residual tumor infiltratinf the brainstem.

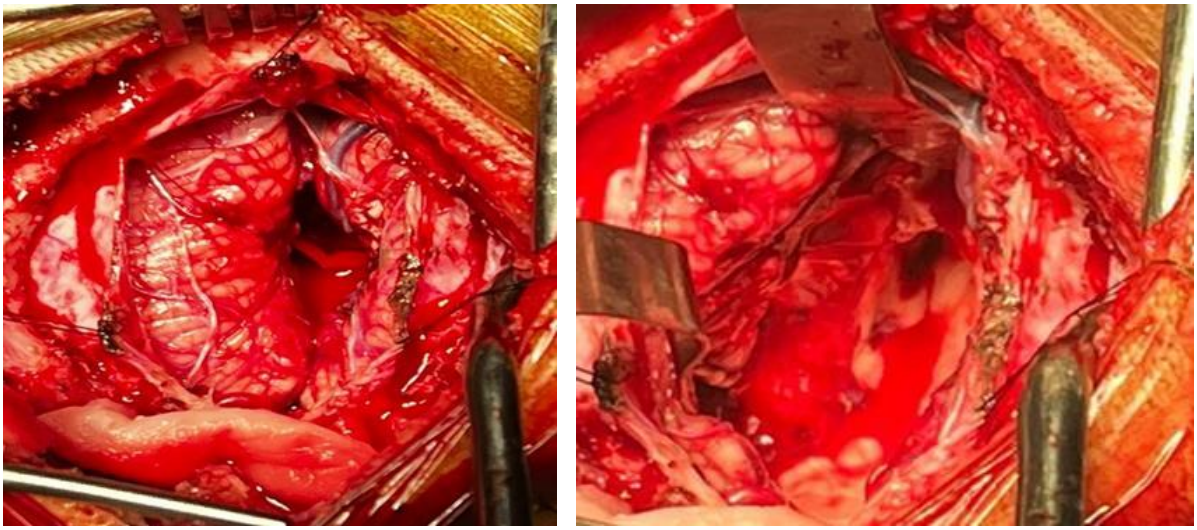

Figure 3: Intra operative views of the cerebellar glioblastoma resection

DISCUSSION:

Glioblastoma multiforme (GBM) is the most common primary malignant brain tumor in adults, but its occurrence in the pediatric population is rare, accounting for approximately 3% of all pediatric brain tumors [1]. Pediatric GBM differs significantly from its adult counterpart

in terms of molecular characteristics, clinical behavior, and treatment response. The mean age of onset for pediatric GBM.typically ranges between 8.8 to 12.7 years [1], whereas in the present case, the patient was a sixyear-old female, indicating an atypically early onset [2].

In terms of anatomical distribution, pediatric glioblastomas most frequently arise in the supratentorial regions, particularly the frontal lobe, which is involved in 25-35% of cases [3]. However, infratentorial gliomas especially those localized in the cerebellum are more commonly found in younger children under the age of 11 [1]. This anatomical predilection is consistent with our case, which involved a cerebellar glioblastoma in a six-year-old child. Cerebellar GBMs represent a rare subset of pediatric high-grade gliomas (pHGGs), and their infratentorial location presents unique diagnostic and therapeutic challenges [4].
The clinical presentation of pediatric high-grade gliomas varies based on the tumor's location and the child's age. Supratentorial tumors typically cause seizures, visual disturbances, or motor deficits, whereas cerebellar gliomas more commonly present with cerebellar ataxia, headaches, nausea, and signs of increased intracranial pressure [1.5]. In our patient, the primary symptom was cerebellar ataxia, which correlated well with the tumor's infratentorial location.
Magnetic resonance imaging (MRI) with contrast remains the gold standard for initial diagnosis. GBMs typically appear as solid masses with heterogeneous or rim enhancement, central necrosis, and varying degrees of peritumoral edema [6]. In cerebellar GBMs, peritumoral edema tends to be less prominent, and tumor margins are often better defined than their supratentorial counterparts [7]. These imaging features can be helpful in distinguishing cerebellar GBMs from other posterior fossa tumors in children, such as medulloblastomas or ependymomas.
Advanced imaging modalities, such as diffusion-weighted imaging (DWI) and the apparent diffusion coefficient (ADC), further aid in differentiating high-grade gliomas from other tumors [6]. He et al. demonstrated that cerebellar HGGs show higher ADC values compared to primary CNS lymphomas (PCNSLs), consistent with lower tumor cellularity in GBMs. The mean ADCmin for HGGs was 0.83×10mm/sec, compared to 0.53 10 mm^2/sec in PCNSLs ($p < 0.001$) [7]. Although not definitive, these quantitative imaging biomarkers may improve diagnostic accuracy when integrated with conventional MRI findings.
The cornerstone of treatment for pediatric GBM is maximal safe surgical resection followed by adjuvant radiotherapy and chemotherapy [8]. Gross total resection (GTR) has been associated with improved survival in multiple pediatric trials [9]. Nevertheless, GTR is often not feasible in cerebellar tumors due to their proximity to critical structures like the brainstem and deep cerebellar nuclei [1,10]. In our case, only partial resection was achieved, as the brainstem-infiltrating component was deemed inoperable due to high surgical risk.
Postoperative complications significantly impacted the patient's clinical course. The inability to initiate timely radiotherapy due to a malfunctioning ventriculoperitoneal shunt necessitating a ventriculocisternostomy, followed by a severe pulmonary infection, further contributed to the adverse outcome. Timely initiation of radiotherapy is crucial, as delays are associated with significantly reduced overall survival [11]

The optimal radiation dose for pediatric high-grade gliomas is approximately 59.4-60 Gy. administered in 1.8-2 Gy fractions over 30-33 sessions [7]. Missed or postponed radiotherapy is known to negatively affect prognosis. Chemotherapeutic regimens, including those used in the HITGBM trials, typically comprise vincristine, cisplatin, and temozolomide [5.12]. However. the effectiveness of these regimens remains limited, especially in cerebellar GBMs, which often have a more aggressive clinical course than their cortical counterparts [1].

Prognosis for pediatric glioblastoma remains dismal, with median overall survival rates rarely exceeding one year [13-15]. In a large cohort analysis, Karremann et al. found that patients with cerebellar HGGs had significantly worse outcomes than those with cortical tumors. The median OS for cerebellar HGG was 0.92 years compared to 2.03 years for cortical HGG ($p=0.0064$), and five-year OS was 16% vs. 32%, respectively [1]. These findings highlight cerebellar location as an independent predictor of poor prognosis.

The presence of residual tumor following surgery, especially when involving critical structures such as the brainstem, further compromises survival outcomes. In our case, the patient succumbed just two months postoperatively a particularly short interval, though consistent with previous reports showing a median survival of only 1.2 months following glioblastoma progression [9.16] Recurrence rates remain high, reaching up to 90% in some series, reflecting the highly aggressive nature of pediatric GBM [5].

Molecular profiling has become increasingly important for diagnosis, prognosis, and therapeutic stratification in pediatric gliomas. One pivotal molecular marker is epidermal growth factor receptor (EGFR). EGFR overexpression is frequently observed in supratentorial GBMs and is associated with resistance to radiotherapy and poor prognosis [1.7]. Interestingly, cerebellar GBMs often lack EGFR amplification, potentially contributing to a relatively more favorable treatment response [7,12,13].

In our patient, EGFR was not expressed, consistent with the expected cerebellar GBM molecular phenotype. However, the potential benefit was not realized due to severe postoperative complications.

One of the most significant recent discoveries in pediatric neuro-oncology is the H3K27M mutation, which is strongly associated with diffuse midline gliomas (DMGs), a WHO grade IV tumor [8].

H3K27M-mutant tumors have an extremely poor prognosis and are frequently resistant to standard therapies. These tumors also show a higher propensity for ventricular and spinal dissemination [8].

Chang et al. reported that pediatric patients harboring the H3K27M mutation are more than three times as likely to die from the disease than those without it [13]. In our case, H3K27M mutation testing was not performed due to technical and resource constraints a limitation commonly encountered in low- and middle-income countries.

Molecular testing for additional markers, such as IDH1/2 mutations, MGMT promoter methylation, and TP53 mutations, is increasingly recommended to guide treatment [1]. Karremann et al. found that none of the six cerebellar HGGs they tested harbored the H3.3 mutation, suggesting a distinct pathogenesis compared to supratentorial GBMs [1]. These findings underline the need to consider cerebellar GBMs as biologically and clinically distinct entities, which may require different therapeutic approaches and prognostic tools.

To improve prognostication, Chang et al. recently developed a nomogram-based survival prediction model for cerebellar high-grade gliomas in adults [13]. This model incorporates prognostic factors such as WHO grade, surgical resection status, and adjuvant therapy use. While the model was designed for adults, its principles may be extended to pediatric populations, highlighting the continued importance of maximal resection and prompt radiochemotherapy initiation.

Analysis of SEER database data confirmed that gross total resection, radiation, and chemotherapy are all independently associated with improved survival in cerebellar HGGs. In multivariate Cox regression analysis, hazard ratios for survival were 0.553 for GTR, 0.515 for radiation, and 1.662 for chemotherapy ($p<0.001$ for all) [13]. These findings underscore the value of a comprehensive, multimodal approach to treatment, even when faced with limited resources.

CONCLUSION:

Cerebellar glioblastomas in pediatric patients, though rare, present significant challenges due to their aggressive behavior and poor prognosis. Early and accurate diagnosis, along with a multimodal therapeutic approach combining surgical resection, radiation, and chemotherapy, is crucial for improving outcomes. Despite the advancements in treatment, survival rates remain low,
emphasizing the need for ongoing research into novel therapies and better strategies for early detection. Long-term neurological follow-up remains essential to manage the complex sequelae of these tumors and to ensure the best quality of life for affected children.

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