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A Rare Low-Grade Glioma in the Posterior Fossa Mimicking High Grade Characteristics - A Case Study and Literature Review

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Abstract

Introduction: Gliomas of the posterior fossa are a rare and complex subset of central nervous system tumors, comprising approximately 1% of all gliomas. These tumors are typically categorized by their location into cerebellar and brainstem gliomas and are further divided into low- and high-grade forms. While high-grade gliomas (HGG) in the cerebellum are exceedingly rare, there is limited research on their clinical behavior and outcomes. Accurate diagnosis and classification are crucial for effective treatment planning, particularly given the evolving role of genetic analysis in identifying tumor subtypes.

Case description: We present a rare case of a posterior fossa lesion with hydrocephalus, initially radiologically suggestive of a high-grade cerebellar glioma. However, histopathological and immunohistochemical analysis revealed an IDH-mutant astrocytoma, WHO grade II, with a CDKN2A deletion. This unusual finding underscores the importance of advanced diagnostic techniques, including genetic testing, in accurately diagnosing posterior fossa gliomas and ensuring appropriate treatment decisions. The tumor exhibited characteristics distinct from typical high-grade gliomas, demonstrating the need for caution in radiological interpretation and the value of comprehensive molecular profiling.

Practical implications: This case highlights the critical role of genetic analysis in the diagnosis and management of posterior fossa tumors, particularly in

distinguishing low-grade astrocytomas from high-grade gliomas. It also emphasizes the importance of incorporating advanced diagnostic tools to guide treatment decisions, as radiation therapy should be reserved for recurrent or unresectable cases. The findings contribute to the growing understanding of cerebellar gliomas and reinforce the need for personalized, data-driven treatment approaches.

Keywords: Posterior fossa; Glioma; IDH-mutant astrocytoma; Genetic analysis; Case report; Cerebellar tumors

Introduction

Gliomas of the posterior fossa are a rare and intriguing subset of central nervous system tumors, representing approximately 1% of all gliomas in literature. These tumors are traditionally classified by their location into cerebellar and brainstem gliomas and are further divided radiologically and histologically into low- and high-grade forms. High-Grade Gliomas (HGG) of the cerebellum, particularly rare in both adult and pediatric populations, are underrepresented in existing research, with limited data available on their clinical behavior and outcomes. The treatment of infratentorial high-grade gliomas remains a subject of debate. Emerging evidence indicates that cerebellar high-grade gliomas share clinical characteristics with supratentorial HGG, warranting similar treatment protocols. In contrast, low-grade cerebellar gliomas, all classified as grade I,exhibit excellent outcomes with complete surgical resection, obviating the need for radiation. Genetic analysis, including IDH1/IDH2 and BRAF mutations, plays a pivotal role in diagnosis and treatment planning, with radiation reserved for recurrent or unresectable cases. Our study presents an uncommon case of a posterior fossa lesion with hydrocephalus, initially radiologically suggestive of a high-grade cerebellar glioma. However, histopathological and immunohistochemical analysis revealed an IDH-mutant astrocytoma, WHO grade II, with a CDKN2A deletion. This case underscores the importance of comprehensive diagnostic tools, including advanced genetic testing, to ensure accurate classification and appropriate management of these rare tumors.

Case History

A 30-year-old female patient attended our outpatient clinic with a significant history of recurrent headaches accompanied by episodes of vomiting and blurred vision. She reported seeing several general practitioners locally initially then referred them for further evaluation and management at our facility. Apart from a single hospitalization that was a cesarean section, her other medical history was unremarkable. She did not have any documented alcohol or substance abuse history. Her family medical history was also insignificant.

The patient was seen clinically and then received an MRI of the brain with contrast. Upon that, a mass in the posterior cranial fossa was found out with characteristics of internal necrosis and peripheral enhancement. The size of the mass was approximately 4.4 x 4.7 x 4.8 cm in anteroposterior, transverse, and craniocaudal measurements, respectively. T1-weighted imaging indicated intratumoral hemorrhage with diffusion restriction. The findings on imaging suggested considerable mass effect on the fourth ventricle, with moderate obstructive hydrocephalus and mild pons pressure. Differential diagnoses were hemangioblastoma and astrocytoma.

To treat the hydrocephalus, a right parietal ventriculoperitoneal shunt was placed on July 11, 2024, utilizing a 6 cm ventricular catheter and a standard distal catheter via a burr hole medium pressure shunt. Post placement of the intervention, the patient underwent a suboccipital craniotomy for near-total excision of the midline cerebellar tumor. Intra-operative findings were a firm, vascular tumor with internal hemorrhage inside it. Histopathological examination revealed a low-grade glial neoplasm with a pattern of sheets of tumor cells showing mild atypia with no areas of necrosis or microvascular proliferation. Immunohistochemical studies showed that the tumor highlighted for GFAP and OLIG2 and was IDH1 positive, although ATRX was retained. With this history, oligodendroglioma was favored histologically but analysis with FISH failed to detect 1p19q codeletion. Combined overall impression indicated an IDH-mutant astrocytoma classified as WHO grade II with CDKN2A deletion at 9p21. The case was thoroughly reviewed in a Multidisciplinary Team (MDT) meeting, which included discussions among histopathologists, neurosurgeons, oncologists, and radiologists, among other specialists. Given the significant enhancement observed on post-contrast MRI images, testing for CDKN2A/B deletion was considered, although it was later found to be undetected. In light of this, a treatment plan involving radiotherapy was developed. Initially, a dose of 54 Gy delivered over 30 fractions was considered, but due to dose constraints, the plan was adjusted to 50.4 Gy over 28 fractions. The patient has now completed her course of radiation and is currently doing well from a clinical perspective. This case highlights the intricacy of posterior fossa tumors with varied atypical radiological features, emphasizing the need for comprehensive imaging and histopathological assessment for appropriate management strategies.

Discussion

The World Health Organization (WHO) classification of Central Nervous System (CNS) tumors has undergone significant advancements, with the 2021 version (WHO CNS5) introducing crucial updates in tumor taxonomy and grading systems to reflect the evolving understanding of molecular and genetic features [1]. Gliomas, the most common primary brain tumors, are now classified based on integrated histopathological and molecular profiles, enhancing diagnostic accuracy and treatment stratification [1]. Astrocytomas, oligodendrogliomas, and glioblastomas represent the primary subtypes of gliomas. Glioblastoma, the most aggressive form, is characterized by diffuse infiltration, rapid progression, and resistance to standard therapies [1]. In contrast, oligodendrogliomas, marked by IDH mutations and 1p/19q codeletions, exhibit better prognoses and therapeutic responses [2]. Molecular markers, such as IDH mutations and ATRX loss, play pivotal roles in differentiating these subtypes, with IDH-mutant astrocytoma associated with more favorable outcomes compared to their IDH-wild type counterparts [3,4].

Posterior fossa gliomas are rare, comprising1% to 4.6% of all gliomas [5]. These tumors are typically categorized into cerebellar and brainstem gliomas, with further subdivision into low- and high-grade types based on radiological and histopathological criteria [6,7]. High-Grade Gliomas (HGGs) in the cerebellum are particularly uncommon and often excluded from clinical trials due to their presumed aggressive behavior and poorer outcomes compared to supratentorial gliomas [8,9]. However, recent

studies suggest comparable survival outcomes for cerebellar and supratentorial HGGs when managed with surgical resection and adjuvant therapies, challenging this exclusionary approach [5,10]. Cerebellar Glioblastoma Multiforme (GBM) often presents with rapid local progression and brainstem invasion, factors associated with poor prognoses [5,9]. Median survival for cerebellar HGG ranges from 11 to 32 months depending on tumor grade and extent of resection (8). Surgical resection followed by adjuvant radiation and chemotherapy, including Temozolomide (TMZ), improves survival [2]. Current evidence suggests that cerebellar gliomas should be managed according to standard protocols for HGGs, advocating their inclusion in clinical trials [5,11]. In contrast, Low-Grade Gliomas (LGGs) in the cerebellum, often classified as WHO grade I, typically achieve excellent outcomes with gross total resection alone. Recurrence may necessitate additional interventions such as radiation therapy [10,12]. Malignant transformation, while rare, underscores the importance of long-term follow-up [10].

Radiological evaluation, particularly MRI, remains critical in glioma diagnosis and management. Advanced techniques, including Diffusion-Weighted Imaging (DWI), Apparent Diffusion Coefficient (ADC) mapping, and MR spectroscopy, aid in tumor grading and predicting molecular subtypes [13-15]. For instance, the T2-FLAIR mismatch sign is a highly specific imaging biomarker for IDH-mutant astrocytomas without 1p/19q codeletions [14]. Elevated D-2-hydroxyglutarate levels detected via MR spectroscopy further confirm the presence of IDH mutations [15].

Pathology remains the diagnostic gold standard, though multidisciplinary collaboration between neurosurgeons, neuroradiologists, and pathologists is essential to minimize diagnostic errors and refine treatment strategies [16-18]. Immunohistochemistry (IHC) is indispensable for confirming molecular profiles, with IDH1 R132H mutation and ATRX loss serving as key diagnostic markers [2,4]. Homozygous deletion of CDKN2A/B, even in histologically low-grade tumors, warrants a WHO grade 4 classifications [1]. We present a rare case of an IDH-mutant astrocytoma in the posterior fossa with clinical and radiological features mimicking a high-grade lesion. The patient presented with hydrocephalus and cerebellar symptoms. MRI revealed a T2 hyperintense, enhancing lesion without a T2-FLAIR mismatch, initially suggesting a high-grade glioma. Histopathological examination revealed mild atypia, absence of necrosis, and low microvascular proliferation. IHC confirmed IDH1 positivity, ATRX retention, and GFAP/OLIG2 expression, while fluorescence in situ hybridization (FISH) for 1p/19q codeletion was negative. The tumor was ultimately classified as an IDH-mutant astrocytoma, WHO grade II, with CDKN2A deletion at 9p21, highlighting the diagnostic complexities of posterior fossa gliomas [2,6]. This case underscores the need for comprehensive, multidisciplinary evaluations incorporating radiological, histopathological, and molecular analyses to achieve accurate diagnoses and guide optimal management strategies. While conventional MRI and pathology remain invaluable, advanced imaging and molecular diagnostics are integral to the modern classification and treatment of gliomas. Further research and inclusion of rare posterior fossa gliomas in clinical trials are imperative to refine therapeutic approaches and improve patient outcomes [5,11].

Conclusion

In conclusion, the 2021 WHO CNS classification refines the approach to glioma diagnosis and management, integrating molecular markers like IDH, ATRX, and CDKN2A/B, along with advanced imaging techniques to enhance prognosis prediction and treatment planning. This evolving framework is key to improving outcomes for patients with brain tumors.

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