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Case Report

Giant Fourth Ventricular Ependymoma Presenting with Acute Hydrocephalus in a Young Adult

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Abstract

Background: Fourth ventricular ependymomas predominantly occur in children, and adult cases presenting with massive tumor size and acute hydrocephalus are uncommon. Surgical management is particularly challenging when tumors are adherent to the floor of the fourth ventricle, where aggressive resection may result in significant neurological morbidity. Recent molecular classification has further highlighted the heterogeneity of posterior fossa ependymomas.

Case Description: A 27-year-old man presented with a 1-month history of progressive headache and nausea, followed by impaired consciousness due to acute obstructive hydrocephalus. Imaging revealed a giant tumor, approximately 5 cm in diameter, occupying the entire fourth ventricle. Emergency external ventricular drainage resulted in rapid neurological improvement. Definitive tumor resection was subsequently performed via a midline suboccipital craniotomy using the telovelar approach. Although most of the tumor was safely removed, a small portion densely adherent to the floor of the fourth ventricle was intentionally preserved to avoid neurological injury. Postoperative contrast-enhanced MRI demonstrated no residual enhancing lesion. Histopathological examination revealed ependymoma with focal anaplastic features (WHO grade III). Molecular analysis showed predominantly posterior fossa group A (PFA) characteristics with focal posterior fossa group B components. Adjuvant radiotherapy (54 Gy) was administered. The postoperative course was uneventful, and the patient returned to his previous occupation without neurological deficits.

Conclusion: This case demonstrates that giant fourth ventricular ependymomas in adults, with predominantly PFA molecular features, can be effectively managed using a telovelar approach that prioritizes neurological safety, combined with appropriate adjuvant radiotherapy, resulting in favorable functional and oncological outcomes.

Keywords: Fourth Ventricular Ependymoma; Telovelar Approach; Hydrocephalus; Posterior Fossa Tumor

Abbreviations

PFA: Posterior Fossa Group A; PFB: Posterior Fossa Group B; MRI: Magnetic Resonance Imaging

Introduction

Ependymomas are glial tumors arising from ependymal cells lining the ventricular system and account for 2–3% of all intracranial tumors [1]. They occur more frequently in children, with posterior fossa ependymomas commonly originating from the fourth ventricle. In adults, intracranial ependymomas are relatively rare and exhibit heterogeneous clinical behavior [2].

Gross-total resection is consistently reported as the most important prognostic factor in the management of intracranial ependymomas [3,4]. However, achieving complete resection can be technically challenging, particularly in large fourth ventricular tumors adherent to the ventricular floor. Excessively aggressive resection in this region may result in severe neurological deficits, including cranial nerve dysfunction and long-term brainstem injury [5].

Recent advances in molecular profiling have identified two major subgroups of posterior fossa ependymomas: posterior fossa group A (PFA) and posterior fossa group B (PFB) [6,7]. PFA tumors are more common in children and are generally associated with a poorer prognosis, whereas PFB tumors are more frequent in adolescents and adults and are associated with more favorable outcomes [7,8]. However, the clinical significance of intratumoral molecular heterogeneity remains incompletely understood.

Here, we report a rare case of a giant fourth ventricular ependymoma in a young adult presenting with acute hydrocephalus. We emphasize the role of the telovelar approach, surgical decision-making that prioritizes neurological safety, and the implications of mixed PFA and PFB molecular features.

Case Presentation

A 27-year-old man with no significant medical history developed progressive headache and nausea approximately 1 month prior to admission. He was initially evaluated at a local internal medicine clinic and treated conservatively; however, his symptoms failed to improve. Subsequent computed tomography revealed a large posterior fossa mass occupying the fourth ventricle with associated obstructive hydrocephalus, and the patient was referred to our institution.

On admission, the patient's level of consciousness progressively deteriorated. Emergency external ventricular drainage was therefore performed, resulting in immediate improvement in consciousness. A head Computed Tomography scan (CT) revealed a tumor with mixed low- and high-density areas filling the fourth ventricle, as well as hydrocephalus (Figure 1A, B). Magnetic resonance imaging (MRI) demonstrated a well-circumscribed, heterogeneously enhancing tumor measuring approximately 5 cm in maximum diameter, occupying the entire fourth ventricle and compressing the brainstem (Figure 2A–E).

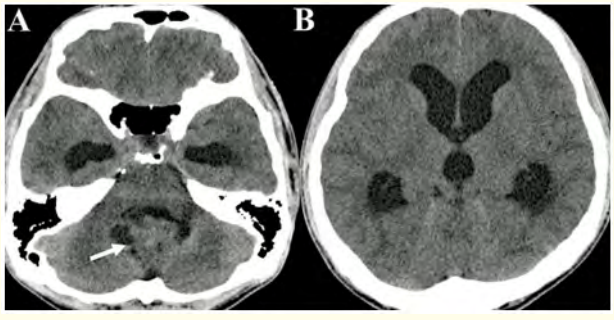

Figure 1

A: A tumor filling the fourth ventricle was identified, with findings showing a mixture of low- and high-density areas. Arrow: tumor.

B: Significant ventricular enlargement was observed.

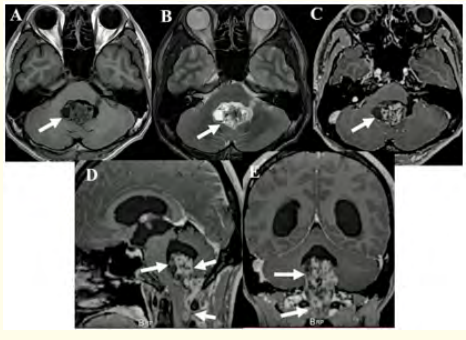

Figure 2: Preoperative MRI.

Arrow: tumor

A: T1 WI axial MRI revealed a tumor containing low to iso signal intensity components posterior to the fourth ventricle.

B: T2 WI axial MRI showed a tumor involving areas ranging from low to high signal intensity.

C: The tumor showed heterogeneous gadolinium enhancement.

D: The T1 gadolinium-enhanced sagittal section showed a tumor measuring 5 cm in length extending to the upper cervical spinal cord level.

E: A giant tumor measuring 5 cm in the longitudinal axis and 3.5 cm in the transverse axis was identified on the T1 gadolinium-enhanced coronal section.

Definitive surgical resection was performed via a midline suboccipital craniotomy using the telovelar approach [9]. The cerebellomedullary fissure was carefully opened without splitting the cerebellar vermis, allowing wide exposure of the fourth ventricle [10]. The tumor was internally decompressed and circumferentially dissected under direct visualization (Figure 3A). Although most of the tumor was safely separated from the surrounding structures, a portion was found to be densely adherent to the floor of the fourth ventricle. To minimize the risk of brainstem injury, this adherent component was intentionally preserved (Figure 3B).

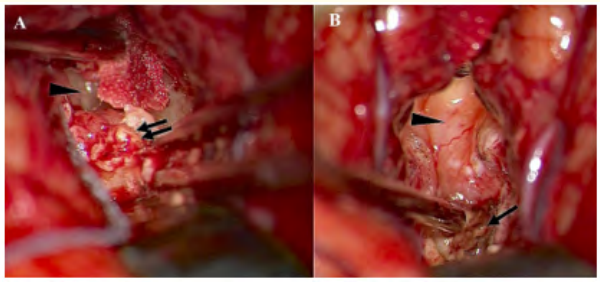

Figure 3: Surgical view.

A: Using a Telovelar approach, we were able to capture the overall morphology of the tumor. The tumor was soft and prone to bleeding. Double arrows: tumor, arrowhead: floor of the fourth.

B: Most of the tumor was removed, but there was a portion that was highly adherent to the floor of the fourth ventricle, which was left behind (arrow).

Postoperative contrast-enhanced MRI demonstrated no apparent residual enhancing lesion, consistent with near-total to gross-total resection (Figure 4A, B). The postoperative course was uneventful, and no new neurological deficits were observed.

Histopathological examination confirmed the diagnosis of ependymoma with focal anaplastic features, corresponding to WHO grade III [11] (Figure 5A, B, C, D). Molecular analysis revealed that the tumor was predominantly PFA, with focal areas exhibiting PFB characteristics [6,7]. Given the presence of anaplastic components and the predominantly PFA molecular profile, adjuvant focal radiotherapy was administered to a total dose of 54 Gy [12].

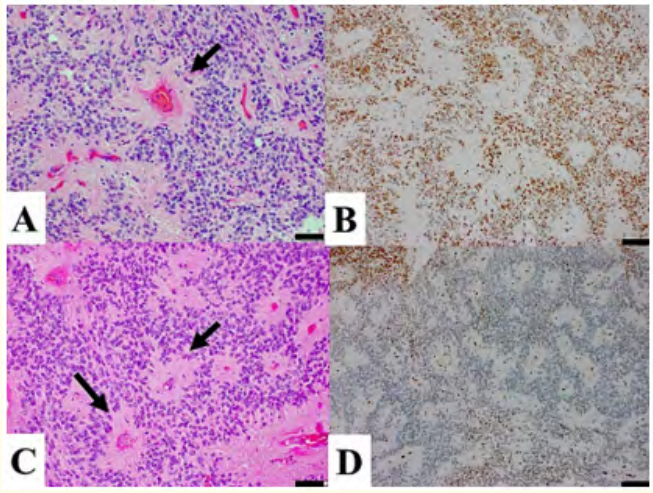

Figure 4: Histopathological and molecular features supporting the diagnosis and subclassification of ependymoma (PFA vs PFB).

(A, B) Representative images of posterior fossa group A (PFA) ependymoma.

(A) Hematoxylin and eosin (H&E) staining demonstrating a moderately cellular tumor with uniform nuclei and characteristic ependymal features, including perivascular pseudorosette formation (arrow) (scale bar = 100 μm).

(B) Immunohistochemistry for H3K27me3 showing loss of nuclear expression, supporting classification as PFA ependymoma (scale bar = 50 μm).

(C, D) Representative images of posterior fossa group B (PFB) ependymoma.

(C) H&E staining demonstrating similar ependymal morphology with perivascular pseudorosettes (arrow) (scale bar = 50 μm).

(D) H3K27me3 immunostaining demonstrating retained nuclear expression, supporting classification as PFB ependymoma (scale bar = 50 μm).

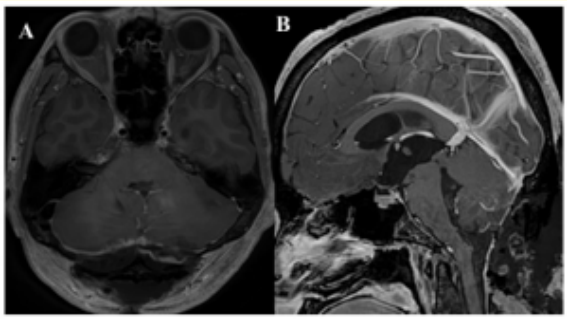

Figure 5: Postopeative MRI.

A: Axial section, B: Sagittal section.

The postoperative contrast-enhanced MRI did not reveal any clearly enhanced areas.

At follow-up, the patient remained neurologically intact, showed no radiological evidence of tumor recurrence, and had resumed his previous occupation.

Discussion

Fourth ventricular ependymomas in adults are rare and often present with nonspecific symptoms such as headache and nausea, which may lead to diagnostic delay [2]. In this case, initial conservative treatment contributed to delayed diagnosis and subsequent acute neurological deterioration due to obstructive hydrocephalus. Emergency cerebrospinal fluid diversion was crucial in stabilizing the patient and enabling safe definitive surgical management [13].

The extent of resection remains the most important prognostic factor in ependymoma treatment [3,4]. However, large tumors occupying the entire fourth ventricle frequently exhibit dense adhesion to the ventricular floor, making complete resection hazardous [5]. In such situations, preservation of neurological function should take precedence over anatomical radicality. Intentional subtotal resection followed by adjuvant therapy represents a reasonable and widely accepted management strategy [12].

The telovelar approach has become the preferred surgical corridor for fourth ventricular tumors, as it provides wide exposure of the ventricular cavity through the cerebellomedullary fissure without splitting the cerebellar vermis [9,10]. Compared with the traditional transvermian approach, the telovelar approach is associated with a lower risk of cerebellar mutism and long-term cerebellar dysfunction [14]. In this case, this approach provided excellent visualization of the tumor–brainstem interface, facilitating maximal safe resection and enabling clear identification of regions where further dissection would have posed unacceptable neurological risk.

Molecular classification has added an important dimension to the prognostic assessment of posterior fossa ependymomas [6,7]. PFA tumors are typically associated with younger age at presentation and poorer outcomes, whereas PFB tumors generally occur in older patients and are associated with a more favorable prognosis [7,8]. The present case is notable for demonstrating predominantly PFA molecular features in an adult patient, with focal PFB components, suggesting intratumoral molecular heterogeneity. Although the clinical implications of such mixed molecular profiles remain unclear, this heterogeneity may partly explain the favorable clinical course observed despite the presence of high-risk features.

The identification of focal anaplastic components (WHO grade III) further supported the use of adjuvant radiotherapy [11,12]. Postoperative radiotherapy at a total dose of 54 Gy is commonly recommended for high-grade or incompletely resected ependymomas and likely contributed to durable local tumor control in this patient.

Further accumulation of similar adult cases with molecular analysis will be necessary to clarify the clinical significance of mixed PFA and PFB characteristics and to optimize surgical and adjuvant treatment strategies.

Conclusion

In conclusion, this case demonstrates that adult patients with giant fourth ventricular ependymomas, including those with predominantly PFA molecular characteristics, can achieve excellent functional and oncological outcomes through a surgical strategy that emphasizes neurological safety using the telovelar approach, combined with appropriate adjuvant radiotherapy

Ethical Approval and Patient Consent

Written informed consent for publication of this case and accompanying images was obtained from the patient.

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Conflict of Interest

The authors declare that they have no conflict of interest.

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