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(CASE REPORT)

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Adult deep midline pilocytic astrocytoma: A case report highlighting diagnostic and management challenges with brief literature review

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Abstract

Adult deep midline pilocytic astrocytoma (PA) is a rare, slow-growing World Health Organization (WHO) grade 1 central nervous system tumor arising in deep structures such as the brainstem, hypothalamus, or thalamus. Recognition of this entity is important because its deep location often precludes complete surgical resection. Adults may experience higher recurrence rates than children, and misdiagnosis can lead to inappropriate or overly aggressive treatment.

We report the case of a 34-year-old man with progressive headaches, nausea, and diplopia. Neuroimaging revealed a cystic hypothalamic mass with an avidly enhancing mural nodule extending toward the optic chiasm. Histopathologic examination demonstrated the classic biphasic pattern and Rosenthal fibers characteristic of PA. The tumor showed a low Ki-67 proliferation index. Molecular testing was negative for IDH1 mutations and the common BRAF-KIAA1549 fusion. Because of the tumor location, maximal subtotal resection was performed, resulting in improvement of hydrocephalus and visual symptoms.

At the 18-month follow-up, the patient remained clinically stable, and MRI showed a residual lesion with no significant change. However, at 26 months, imaging demonstrated slight enlargement of the residual nodule, consistent with tumor progression. To preserve neurological function, targeted therapy with a BRAF/MEK inhibitor was initiated. The patient was advised to undergo long-term surveillance with serial MRI, given the possibility of delayed recurrence following initial treatment.

This case underscores the diagnostic and therapeutic challenges of deep midline PA in adults. It highlights the value of multidisciplinary management and molecular profiling in guiding treatment when gross total resection is not feasible.

Keywords: Pilocytic Astrocytoma; Hydrocephalus; BRAF/MEK Inhibitor; Rosenthal Fibers; Grade 1 Gliomas; Supratentorial

1. Introduction

Pilocytic astrocytoma (PA) is a circumscribed astrocytic glioma, classified as a CNS WHO grade 1 tumor, that predominantly affects children and adolescents. [1,2] Its occurrence in adults is uncommon and may present a diagnostic challenge, particularly when it arises in deep midline diencephalic structures rather than in the more familiar cerebellar location. Hypothalamic and optic-chiasmatic PAs are especially challenging because their clinical

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manifestations, including headache, visual disturbance, endocrine dysfunction, and obstructive hydrocephalus, overlap with those of other sellar, suprasellar, and third-ventricular lesions. [3]

Although often histologically low grade, adult PAs do not invariably follow the indolent course expected from pediatric experience; adult series have documented clinically meaningful rates of progression and recurrence, particularly when complete resection is precluded. [4,5] In the hypothalamic region, the surgical objective must be balanced against the risk of hypothalamic, visual, and neuroendocrine morbidity. Moreover, increasing recognition of MAPK pathway driver alterations has made integrated histomolecular assessment increasingly relevant to diagnostic confirmation and treatment planning. [6]

We report this case to highlight the diagnostic uncertainty of a cystic, enhancing deep midline tumor in an adult, the value of tissue diagnosis and multidisciplinary decision-making, and the need for long-term surveillance after intentional subtotal resection.

2. Case Presentation

2.1. Clinical Presentation

A 34-year-old male presented with a several-month history of progressive headaches, intermittent nausea, and episodes of diplopia. He initially attributed these symptoms to stress and eyestrain but sought medical attention after experiencing a sudden, severe headache accompanied by vomiting and a brief episode of confusion. He had no significant past medical history and denied any personal or family history of neurofibromatosis type 1 (NF1) or other neurological conditions. His presentation was consistent with a progressive mass effect and early obstructive hydrocephalus in the deep midline structures.

2.2. Physical Examination and Diagnostic Workup

A neurological examination revealed mild bilateral papilledema and subtle ataxia, but no motor or sensory deficits. Laboratory tests were unremarkable. An urgent non-contrast computed tomography (CT) scan of the head showed a well-circumscribed, partially cystic lesion in the deep midline, causing effacement of the third ventricle and early hydrocephalus.

A subsequent contrast-enhanced magnetic resonance imaging (MRI) demonstrated a predominantly cystic mass with a vividly enhancing mural nodule within the hypothalamus, extending toward the optic chiasm. The lesion showed no evidence of diffusion restriction. The radiological differential diagnosis included a pilocytic astrocytoma, a craniopharyngioma, a low-grade glioma, or a more aggressive lesion such as a high-grade astrocytoma.

2.3. Multidisciplinary Tumor Board Discussion

The case was presented at the multidisciplinary tumor board. The discussion centered on the diagnostic challenge of an adult deep midline pilocytic astrocytoma, its rarity, and the need to differentiate it from other enhancing lesions. Given the tumor location in the hypothalamus and proximity to the optic chiasm, the team concluded that a gross total resection would pose a significant risk of morbidity. The consensus favored a strategy of maximal safe surgical resection, with the primary goal of obtaining a definitive tissue diagnosis and relieving the hydrocephalus. Postoperative management included long-term surveillance with serial MRI to monitor tumor progression.

2.4. Special Studies and Pathology

Microscopic examination of the surgical specimen revealed the classic biphasic pattern of a pilocytic astrocytoma, consisting of densely fibrillated areas with Rosenthal fibers and loosely textured microcystic areas with eosinophilic granular bodies, consistent with a WHO grade 1 tumor. (**Figure 1 A, B, C, D**) Immunohistochemical (IHC) staining showed the tumor cells were positive for glial fibrillary acidic protein (GFAP), while the Ki-67 proliferation index was low (< 2%). ATRX expression was retained, and IDH1 R132H staining was negative. Molecular studies confirmed the absence of an IDH mutation and revealed no BRAF-KIAA1549 fusion, which is more common in pediatric cases, but did not detect any other actionable alterations.

2.5. Treatment, Outcome, and Follow-up

The patient underwent subtotal resection, leaving a small residual focus in situ to preserve critical hypothalamic and optic tract integrity. Surgery relieved the hydrocephalus, and his postoperative course was uneventful, with near-complete resolution of headaches and diplopia. He was monitored with serial surveillance MRIs. At 18 months, he

remained clinically well, and MRI showed stable residual disease. At 26 months, however, imaging showed slight enlargement of the residual nodule, consistent with recurrence. Given the slow progression and excellent functional status, the tumor board recommended targeted therapy with a BRAF/MEK inhibitor, based on molecular evidence of MAPK pathway involvement, on the concept of targeting broader, alternative alterations within the MAPK/ERK signaling pathway that still drive tumor growth, rather than immediate re-irradiation or repeat surgery. This approach aimed to control progression while preserving quality of life.

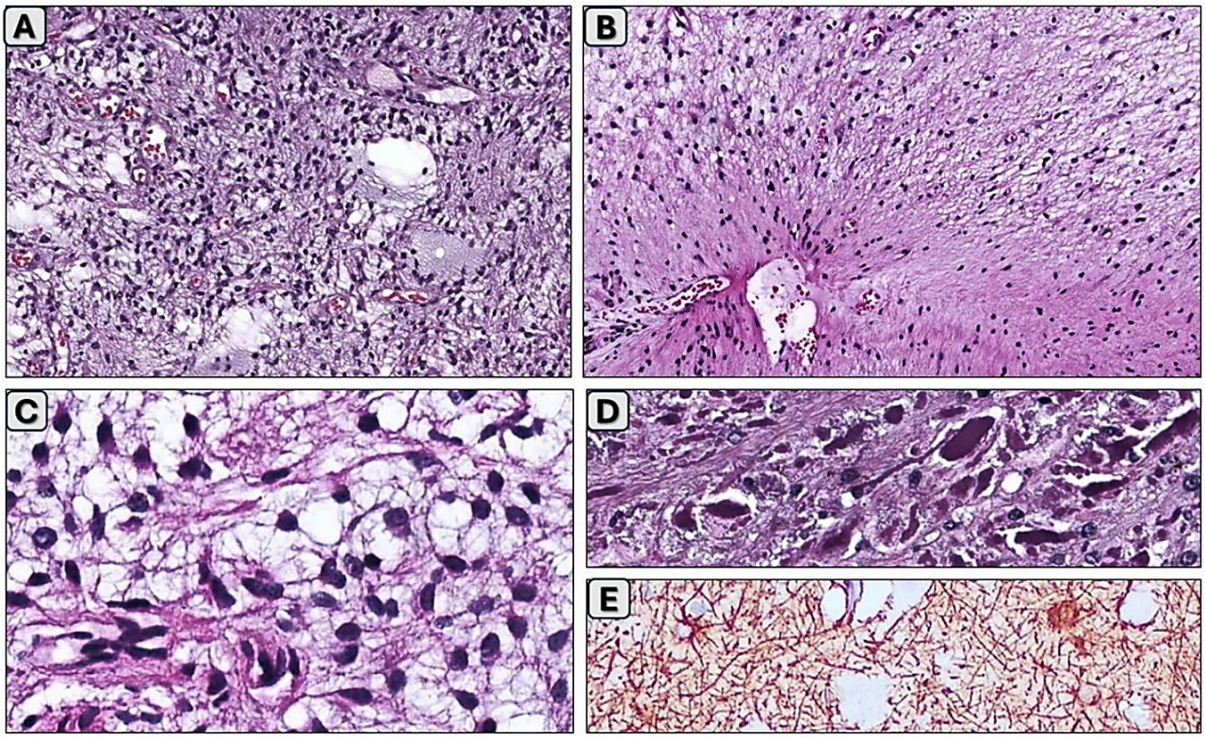

Figure 1 Microscopic features of pilocytic astrocytoma

1A**:** Low-power view showing classic biphasic appearance with cellular areas alternating with hypocellular, microcystic areas (H&E X20); 1B**:** Intermediate-power view showing piloid astrocytic cells with long delicate hair-like processes (H&E X40); 1C**:** High-power view showing oval and spindled tumor nuclei with bland chromatin, large, vacuolated cytoplasm, and degenerative-type atypia (H&E X60); 1D: High-power view showing brightly eosinophilic, corkscrew-shaped Rosenthal fibers; 1E: Tumor cells positive for GFAP

3. Discussion

3.1. Background (History, epidemiology, risk factors, and WHO classification)

Pilocytic Astrocytomas (PAs), first described by Henry Cushing in 1931, are benign, well-circumscribed tumors classified as WHO grade 1 glioma. [7,8] They were historically referred to as cystic cerebellar astrocytoma or juvenile pilocytic astrocytoma. PAs are the most common primary brain tumor within the pediatric population, often found in adolescents, with approximately 75% of patients diagnosed prior to 20 years of age. [7,9] In contrast, PA is rare in adults, resulting in limited literature within this population. [2,10,11]

Progression-free survival (PFS) and overall survival (OS) of patients diagnosed with PA depend on factors such as mutation, histopathology, location, extent of surgical resection, and age. [12] PA frequently demonstrates BRAF translocations in pediatric populations, associated with greater PFS, whereas adults commonly harbor BRAF V600E and non-BRAF mutations. [13]

Histopathologically, pediatric tumors exhibited a higher proportion of vasculopathy, pilomyxoid changes, dispersed growth patterns, and elevated miotic activity. In contrast, a greater prevalence of calcification, necrosis, inflammatory infiltration, hemosiderin deposition, and compact growth patterns was observed in adults. [6,14] Factors such as brisk mitotic activity, pilomyxoid changes, absence of calcification, hemosiderin deposition, and exuberant vasculopathy were associated with shorter PFS and OS. [14]

Most pediatric PAs are located infratentorially within the posterior fossa. In contrast, adult tumors have a higher rate of supratentorial localization; both locations have a better prognosis than midline or spinal cord tumors. [2,7,13] Furthermore, complete resection yields a more favorable outcome, with a 95% 10-year survival rate. [7] Overall, age-related distinctions in PA demonstrated a worsening prognosis with age, as well as an increased likelihood of tumor progression and recurrence among adults. [10]

3.2. Pathogenesis, Pathophysiology

PA is fundamentally a MAPK-pathway-driven neoplasm. Most sporadic tumors harbor a single activating alteration, classically a tandem duplication of chromosome 7q34 producing a KIAA1549-BRAF fusion. The resulting constitutively active BRAF kinase stimulates downstream MEK/ERK signaling and supports tumor-cell proliferation. [4,6,13] Less frequent mechanisms include activating alterations in BRAF, FGFR1, KRAS, or NF1; loss of neurofibromin in neurofibromatosis type 1 similarly removes the negative regulation of RAS/MAPK signaling. [6] Adult PAs have a more heterogeneous molecular profile than pediatric tumors, and KIAA1549-BRAF fusion is less common; its absence therefore does not exclude the diagnosis. [3,4]

The biphasic histology in this case, with compact fibrillated areas and Rosenthal fibers alternating with loose microcystic regions containing eosinophilic granular bodies, is characteristic. [12] Retained ATRX expression, negative IDH1 R132H staining, and a low Ki-67 index further support a circumscribed rather than diffuse astrocytoma, although no single marker is diagnostic. [15]

Progressive residual disease may reflect persistent oncogenic signaling despite low-grade histology. Comprehensive molecular profiling at progression is therefore important to establish whether an actionable MAPK alteration is present. [4,6]

3.3. Comparative Analysis of Our Case with Existing Literature.

This patient's headache, nausea, diplopia, papilledema, and early hydrocephalus reflected the mass effect and cerebrospinal fluid obstruction reported in adult hypothalamic PAs. [3] Its cystic lesion with a vividly enhancing mural nodule and no diffusion restriction was compatible with PA. However, age and imaging alone could not establish the diagnosis in this deep midline site. [16] In the 127-patient series by Theeler et al., 17% of adult PAs involved the optic pathway or hypothalamus, and 22% underwent pathology revision after specialist review, underscoring the importance of integrated neuroradiological and neuropathological assessment. [4]

Subtotal rather than gross-total resection was appropriate given the absence of a safe plane at the hypothalamus and optic apparatus. Bond et al. reported that 57 of 79 recurrences (73%) occurred after subtotal resection. [5] Stable residual disease at 18 months and mild enlargement at 26 months therefore represented progression of an at-risk residual lesion, not failure of a function-preserving strategy.

Unlike many pediatric tumors, this PA lacked a KIAA1549-BRAF fusion, and no actionable alteration was reported, illustrating adult PA diversity. MAPK-directed therapy at progression may offer a tissue-sparing option, but agent selection should follow confirmation of a targetable alteration. [4,6]

Abbreviations

- Pilocytic astrocytoma (PA);
- Immunohistochemical (IHC);
- Progression-free survival (PFS)
- Overall survival (OS)

4. Conclusion and What Have We Learned from This Case?

This case adds an adult-onset, deep midline hypothalamic PA with classic histology, absent KIAA1549-BRAF fusion, intentional residual disease, and subsequent slow radiographic progression. It reinforces that the label “low grade” should not be equated with negligible clinical risk when a tumor occupies the hypothalamus or optic chiasm.

In adults with progressive headache, nausea or vomiting, diplopia, papilledema, gait disturbance, or altered mentation, the combination of increased intracranial pressure and a deep midline lesion should prompt urgent assessment for obstructive hydrocephalus and timely multidisciplinary review. A cystic mass with an enhancing mural nodule and no

diffusion restriction may suggest PA, but these features are not pathognomonic; definitive diagnosis requires correlation of imaging with representative histology, immunophenotyping, and molecular findings.

The principal management lesson is that maximal safe resection, rather than anatomical radicality, may be the most appropriate initial strategy when visual, hypothalamic, or neuroendocrine function is at risk. However, intentional residual tumors should be coupled with structured, long-term MRI surveillance, since clinical well-being and early radiographic stability do not preclude later progression. At progression, reassessment by a neuro-oncology multidisciplinary team, including review of pathology and broad molecular testing, is essential to distinguish residual PA from an alternative diagnosis or biologically higher-risk process and to determine whether a molecularly matched therapy is justified.

By documenting these diagnostics, surgical, and surveillance challenges, this report encourages clinicians to recognize adult deep midline PA as a rare but important differential diagnosis. It supports individualized, function-preserving, molecularly informed care.

Compliance with ethical standards

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Statement of ethical approval

This work did not involve any studies on human or animal subjects performed by the authors. It was a retrospective review of archival pathology material collected during routine clinical care, and all data were fully de-identified before analysis.

Data access statement

All relevant data are included in the paper.

Author contributions

All authors contributed equally to producing this manuscript.

Statement of informed consent

The patient was lost to follow-up, and all attempts to reach the patient were unsuccessful. Therefore, the paper has been sufficiently anonymized to maintain patient confidentiality.

References

[1] Torp SH, Solheim O, Skjulsvik AJ. The WHO 2021 Classification of Central Nervous System tumors: a practical update on what neurosurgeons need to know—a minireview. Acta neurochirurgica. 2022 Sep;164(9):2453-64.

[2] Pisharody V, Grigorian J, Chandler KE, Chisango ZM, Chow J, Mao S, Ma T, Jahangiri A, Chern JJ, Hoang KB. 161 Pediatric Versus Adult Pilocytic Astrocytoma (PA): Largest Comparison Cohort of Clinical Course & Genomic

Profile Demonstrates BRAF-KIAA1549 Fusion is Associated With Worse Prognosis in Children Compared to Adults. Neurosurgery. 2025 Apr 1;71(Supplement_1):37.

[3] Costanzo R, Rosetti V, Tomassini A, Fuschillo D, Lofrese G, Iacopino DG, Tosatto L, D'Andrea M. Hypothalamic hemangioma-like pilocytic astrocytoma in an adult patient: A systematic review with a focus on differential diagnosis and neurological presentation. Journal of Clinical Medicine. 2024 Jun 17;13(12):3536.

[4] Theeler BJ, Ellezam B, Sadighi ZS, Mehta V, Tran MD, Adesina AM, Bruner JM, Puduvalli VK. Adult pilocytic astrocytomas: clinical features and molecular analysis. Neuro-oncology. 2014 Jun 1;16(6):841-7.

[5] Bond KM, Hughes JD, Porter AL, Orina J, Fang S, Parney IF. Adult pilocytic astrocytoma: an institutional series and systematic literature review for extent of resection and recurrence. World Neurosurgery. 2018 Feb 1;110:276-83.

[6] Collins VP, Jones DT, Giannini C. Pilocytic astrocytoma: pathology, molecular mechanisms and markers. Acta Neuropathologica. 2015 Jun;129(6):775-88.

[7] Knight J, De Jesus O. Pilocytic Astrocytoma [Internet]. PubMed. Treasure Island (FL): StatPearls Publishing; 2021. Available from: https://www.ncbi.nlm.nih.gov/books/NBK560614/

[8] [8] Louis DN, Perry A, Wesseling P, Brat DJ, Cree IA, Figarella-Branger D, Hawkins C, Ng HK, Pfister SM, Reifenberger G, Soffietti R. The 2021 WHO classification of tumors of the central nervous system: a summary. Neuro-oncology. 2021 Aug 1;23(8):1231-51.

[9] Johnson DR, Brown PD, Galanis E, Hammack JE. Pilocytic astrocytoma survival in adults: analysis of the Surveillance, Epidemiology, and End Results Program of the National Cancer Institute. Journal of neuro-oncology. 2012 May;108(1):187-93.

[10] Mair MJ, Wöhrer A, Furtner J, Simonovska A, Kiesel B, Oberndorfer S, Ungersböck K, Marosi C, Sahm F, Hainfellner JA, Rössler K. Clinical characteristics and prognostic factors of adult patients with pilocytic astrocytoma. Journal of Neuro-oncology. 2020 May;148(1):187-98..

[11] Brossier NM, Strahle JM, Cler SJ, Wallendorf M, Gutmann DH. Children with supratentorial midline pilocytic astrocytomas exhibit multiple progressions and acquisition of neurologic deficits over time. Neuro-Oncology Advances. 2022 Jan 1;4(1):vdab187

[12] Mitschang D, Kleineidam H, Hinz F, Sahm F, Unterberg A, Krieg S, Dao Trong P, Lenga P. Adult pilocytic astrocytomas: challenging the benign paradigm with surgical risks, recurrence dynamics, and molecular insights. Neurosurgical Review. 2025 Sep 19;48(1):658.

[13] Yu T, Hou J, Zheng L, Zhong J, Pan X, Zhang J, Gong J, Chen X, Zhou Q, Wei Y, Chen N. Pilocytic astrocytoma in pediatric and adult patients: a single-center analysis of 650 cases. Virchows Archiv. 2026 Feb 15:1-0.

[14] Sincu A, Orasanu CI, Voda RI, Bosoteanu M, Enciu M, Vamesu S, Ghitoi SA, Deacu M. Unpredictable Evolution of Pilocytic Astrocytoma in Adults: A Case Series and Diagnostic Challenges. The American Journal of Case Reports. 2025 Dec 12;26:e947774.

[15] Hu WM, Wang F, Xi SY, Zhang X, Lai JP, Wu HY, Liu LL, Sai K, Zeng J. Practice of the new integrated molecular diagnostics in gliomas: experiences and new findings in a single Chinese center. Journal of Cancer. 2020 Jan 1;11(6):1371.

[16] Park S, Seo JH, Song SW, Cho YH, Hong CK, Kim JH, Kim HS, Park JE, Nam SJ, Kim YH. Survival outcomes and prognostic factors in surgically treated adult pilocytic astrocytomas. Frontiers in Oncology. 2025 Apr 1;15:1525427.