Intraventricular pilocytic astrocytoma: a case report

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ABSTRACT

Background: Pilocytic astrocytomas (PA) are low-grade, benign gliomas classified as World Health Organization's (WHO) grade I, representing 6% of all gliomas and commonly found in children. Intraventricular pilocytic astrocytomas (IVPA) are rare, accounting for only 4% of PA cases.

Case Presentation: We report a 42-year-old Saudi woman with recurrent positional headaches but no neurological deficits. Magnetic resonance imaging showed a 1.2-cm non-enhancing lesion near the foramen of Monro, initially thought to be a colloid cyst. The lesion was excised via an interhemispheric transcallosal approach. Histopathology confirmed PA with biphasic tissue, glomeruloid vessels, and strong glial fibrillary acidic protein positivity, classified as WHO grade I. No further chemotherapy or radiotherapy was required.

Postoperative Outcome: Post-surgery, the patient had transient short-term memory issues, partially resolving within 6 months, and her headaches ceased. Initial imaging showed pneumocephalus, which resolved after 1 month. **Discussion:** IVPA typically remains asymptomatic until it causes hydrocephalus by obstructing cerebrospinal fluid pathways. Radiologically, they appear as well-defined lesions with mixed cystic and solid components. Complete surgical resection is crucial to minimize recurrence.

Conclusion: IVPA, though rare in adults, should be considered in the differential diagnosis of intraventricular lesions. This case highlights the significance of accurate diagnosis and surgical management for this uncommon tumor.

Keywords: Pilocytic astrocytoma, intraventricular tumor, neurosurgery, case report, adult glioma.

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Background

Pilocytic astrocytomas (PA) are benign, low-grade tumors that are typically well circumscribed. They are categorized as grade I gliomas in the World Health Organization's (WHO) classification system for tumors of the central nervous system [1]. PA represents 6% of all gliomas and is the most prevalent glioma in children, with an insignificant male predominance [2]. In adults, PA typically develops between the ages of 18 and 72 years, with a median age of 29 years. The incidence decreases significantly with age: Patients older than 50 years are rarely diagnosed with them [3]. All levels of the neuraxis may be affected by PA; however, the majority (67%) arise in the cerebellum and optic pathway [4]. Intraventricular pilocytic astrocytomas (IVPA) are extremely uncommon, representing just 4% of all pilocytic tumors [5]. Based on histopathology, PA has low-to-moderate cellularity and highly fibrillated, compact areas rich in Rosenthal fibers [6]. In PA, glial fibrillary acidic protein (GFAP) immunohistochemistry shows diffuse staining and high immunopositivity [7]. PA is linked to the absence of the isocitrate dehydrogenase gene. Furthermore, the presence of the KIAA1549-BRAF gene fusion is seen in some cases, but not all [8].

Case Report

A 42-year-old Saudi female, not known to have any other medical illnesses, presented to the Neurosurgery Outpatient Clinic of Aseer Central Hospital with an off-and-on paroxysmal positional headache. She denied any other neurological symptoms. On physical examination, she was conscious; alert; and oriented to time, place, and person (Glasgow Coma Scale score of 15). The results of cranial nerve assessment and motor and sensory examinations were normal. The patient was referred to the Radiology Department for further investigation. Magnetic resonance imaging (MRI) revealed a 1.2-cm rounded, non-enhanced lesion near the foramen of Monro. The lesion was isointense on T1 imaging and minimally hyperintense on T2 imaging. The radiological impression was consistent with a colloid cyst (Figure 1).

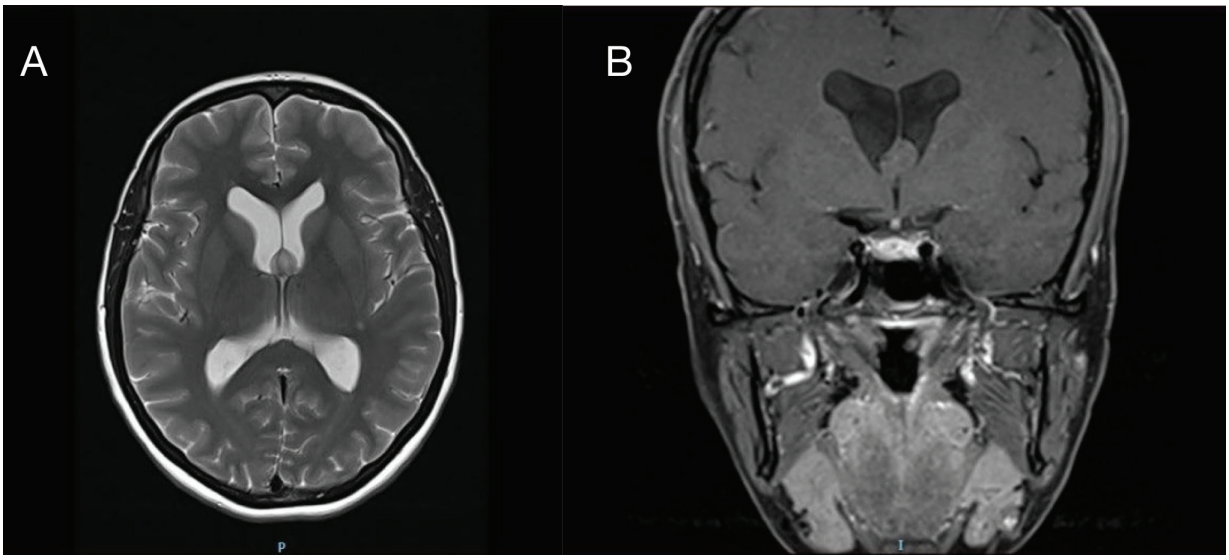

Figure 1. *(A) Axial MRI view shows an intraventricular lesion. (B) Coronal MRI view with contrast shows an intraventricular lesion.*

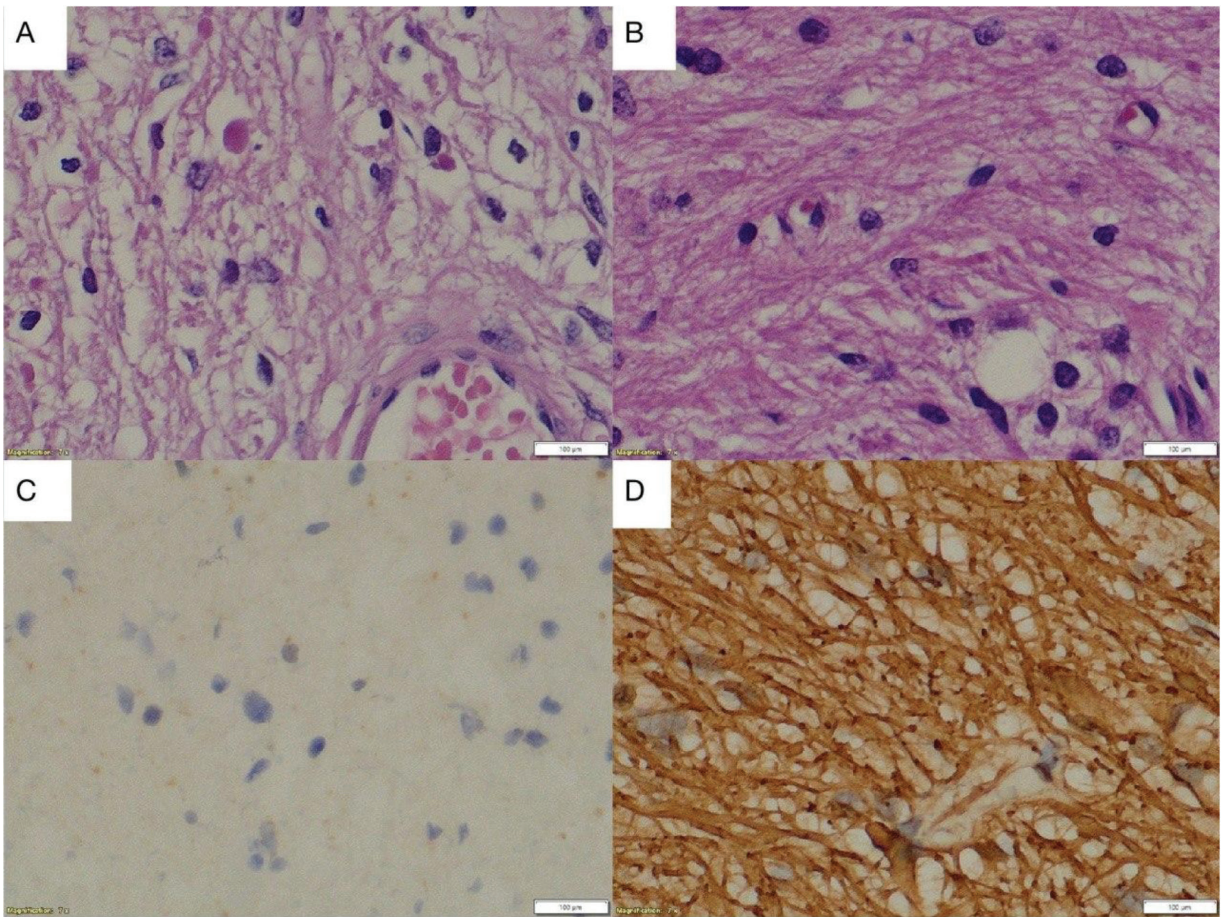

Figure 2. *(A and B) Hematoxylin and eosin stained sections of the tumor. (C) Ki67 immunohistochemistry. The Ki67 labeling index was <1%. (D) GFAP immunohistochemistry.*

Subsequently, the patient underwent surgery for tumor excision by the interhemispheric transcallosal approach. The tumor was resected and sent to the histopathology and cytology lab. Microscopically, the tissue showed proliferation with a biphasic appearance, compact fibrillar portions, elongated nuclei, and microcystic portions. In addition, there were glomeruloid vessels. The histomorphological and immunohistochemical findings were consistent with PA (Figure 2). The tumor was classified as WHO grade 1 for central nervous system tumors;

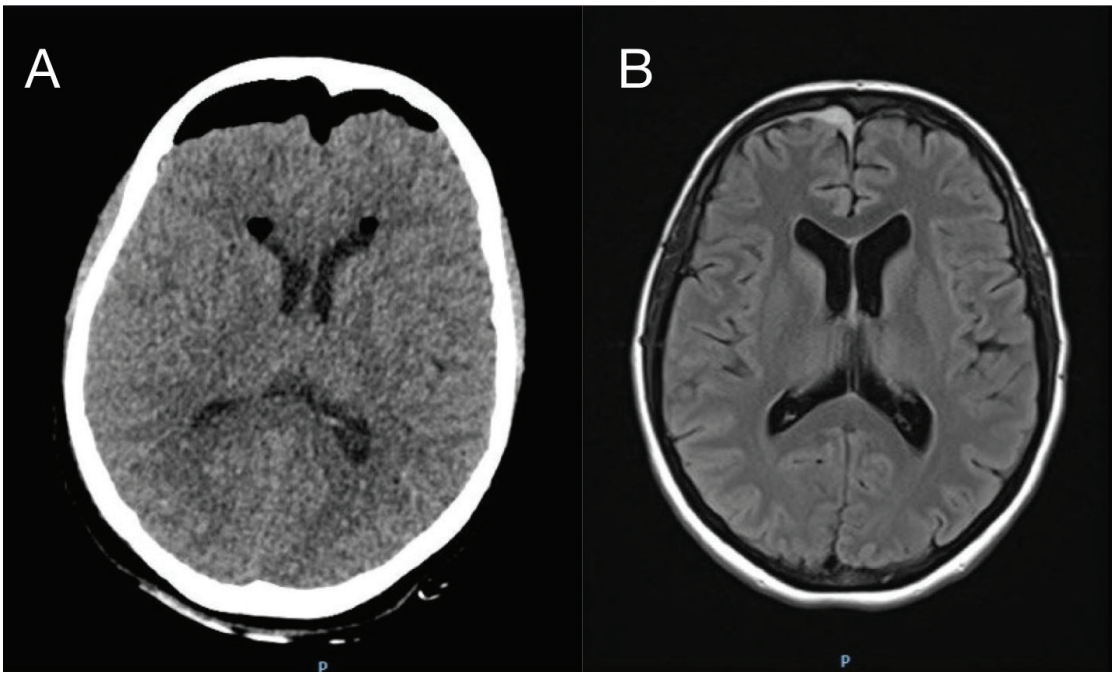

Figure 3. *(A-Left) Postoperative computed tomography showing pneumocephalus. (B-Right) One month after the operation, MRI revealed that pneumocephalus had resolved.*

therefore, neither chemotherapy nor radiotherapy was not administered to the patient.

Postoperatively, the patient had short-term memory impairment, which resolved partially after 6 months; however, her headache had completely subsided. Postoperative computed tomography showed pneumocephalus (Figure 3A), which had resolved 1 month after the operation based on MRI (Figure 3B).

Discussion

Intraventricular tumors are relatively asymptomatic until they grow and block the cerebral spinal fluid (CSF) pathway, resulting in hydrocephalus or signs of increased intracranial pressure such as headache, nausea and vomiting, visual disturbances, and cranial nerve palsies [3,9]. The radiological findings for IVPA typically include a rounded shape with well-circumscribed margins; however, they can occasionally be irregular in shape. The majority of IVPA are mixed cystic and solid, with very few being purely cystic. The cystic areas are highly T2 hyperintense/T1 hypointense, similar to CSF, whereas the solid parts are moderately T2 hyperintense/T1 hypointense with heterogeneous enhancement [10]. Bond et al. [11] conducted a systematic literature review and discovered that 13% of 46 adults with PA had tumor recurrence. All recurrent tumors were first treated with subtotal resection or radiotherapy alone. Therefore, gross total resection should always be the goal in adults with PA [11].

Conclusion

IVPA in adults is an exceedingly rare entity. This case underscores the importance of considering IVPA in the differential diagnosis of intraventricular lesions, even in adult patients. Accurate diagnosis through imaging and histopathological examination is crucial for appropriate management. Surgical resection remains the primary treatment modality, with gross total resection being the goal to minimize the risk of recurrence. Our patient's favorable postoperative outcome, characterized by the resolution of headaches and partial recovery from short-term memory impairment, highlights the efficacy of surgical intervention. Ongoing monitoring and follow-up are essential to ensure long-term tumor control and to address any potential postoperative complications. This case contributes to the limited but growing body of literature on adult IVPA, emphasizing the need for heightened awareness and expertise in managing such a rare tumor.

What is new?

Intraventricular pilocytic astrocytomas in adults are rare but should be considered in differential diagnoses. Surgical resection is key for effective management and reducing recurrence risk.

List of Abbreviations

CSF	Cerebrospinal fluid
CT	Computed tomography
GCS	Glasgow Coma Scale
GFAP	Glial fibrillary acidic protein
IDH	Isocitrate dehydrogenase
IVPA	Intraventricular pilocytic astrocytoma
KIAA1549-BRAF	KIAA1549-BRAF gene fusion
MRI	Magnetic resonance imaging
PA	Pilocytic astrocytoma
WHO	World Health Organization

Conflict of interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

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Consent for publication

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

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

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Summary of the case

1	**Patient (gender, age)**	42 years, female
2	**Final diagnosis**	Intraventricular pilocytic astrocytoma tumer
3	**Symptoms**	Off-and-on paroxysmal positional headach
4	**Medications**	Symptomatic treatment given
5	**Clinical procedure**	Surgery for tumor excision
6	**Specialty**	Neurosurgery