**IMCC Journal of Science**
2023, 3, 31-40
https://myjournal.imcc.edu.ph/

# Intraventricular Pilocytic Astrocytoma: A Case Series

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**ABSTRACT**
Pilocytic astrocytoma (PA) is a low-grade glial tumor and the most common pediatric intracranial tumor, typically located in the infratentorial region. The cerebellum is the most frequent site for PA, and it usually affects children and young adults. While PA can arise anywhere in the central nervous system, its occurrence in the ventricles, particularly the lateral ventricles, is exceptionally rare, accounting for approximately 4% of all pilocytic tumors. We present a series of three cases with the uncommon intraventricular location of PA and an unusual presentation in old age in one of the cases. Due to its unusual location and unexpected age of presentation, intraventricular PA (IVPA) can be misinterpreted in adults. Some IVPAs are highly vascular, and patients may experience profuse intraoperative blood loss if the tumor is not identified on preoperative imaging. Therefore, we aim to highlight the occurrence of such unusual presentations of IVPA, as very few cases have been reported in the literature. It is essential to consider IVPA in adult patients with calcified intraventricular lesions.

**KEYWORDS:** adult; intraventricular; lateral ventricle; pilocytic astrocytoma

**ARTICLE INFO:** Received: 18 September 2023; Accepted: 25 November 2023; Volume: 03; Issue: 02; Type: Case Series

## 1. Introduction

Pilocytic astrocytoma (PA) has been reclassified in the recent 5th edition of the World Health Organization (WHO) classification of central nervous system (CNS) tumors as a circumscribed glioma [1]. PA is an astrocytic neoplasm characterized by varying proportions of bipolar hair-like (pilocytic) cells, compact and loose or myxoid regions, Rosenthal fibers, and eosinophilic granular bodies. It accounts for 17.6% of all childhood primary brain tumors. It is the most common glial tumor in the pediatric age group, comprising around 6% of all gliomas and predominantly affecting males [2]. Although PA can occur anywhere in the CNS, the most common locations are the cerebellum and optic pathway [3]. Patients typically present with symptoms due to mass effect and ventricular obstruction, leading to raised intracranial pressure. Slow growth can lead to diagnostic delays due to subtle symptoms. Intraventricular location is very rare, accounting for around 10% of cases [4,5]. Most of these tumors are located

in the third and fourth ventricles, with very few in the lateral ventricles. Intraventricular pilocytic astrocytoma (IVPA) may exhibit calcification as a degenerative change and can also be highly vascular. Here, we report a series of three rare cases of IVPA, one of which occurred in an elderly patient. Early preoperative diagnosis of IVPA is crucial, as some highly vascular patients may experience complications such as profuse intraoperative blood loss. Informed consent was obtained from all patients for participation in the study after approval from the ethical committee. The objective of the study was to highlight the occurrence of PA at unusual locations and in unusual age groups, which will aid in the timely diagnosis of IVPA cases and facilitate proper intraoperative and postoperative management.

## 2. Case Presentation

### *2.1 Case 1*

**Patient demographics and clinical presentation:** A 64-year-old female patient presented with difficulty walking, persistent left-sided headache, memory loss, and bowel and bladder incontinence that had been developing gradually over the past two months. She had no co-morbidities such as hypertension or diabetes and no history of trauma or surgery. The headache was persistent but not associated with loss of consciousness, nausea, or projectile vomiting. The general physical examination was unremarkable. She was well-oriented to time, place, and person, and the cranial nerve examination was normal. The Glasgow coma scale was E4M6V5, with all four limbs moving normally and bilateral pupils normal with spontaneous eye opening.

**Imaging findings:** The patient was sent to radiology for further investigation. Magnetic resonance imaging (MRI) revealed a well-defined, heterogeneously enhancing T1 hypointense and T2 isointense lesion in the region of the left foramen of Monro, with grossly dilated bilateral lateral ventricles, suggestive of central neurocytoma (Figure 1).

**Pathological findings:** Blood investigations were within normal limits. The patient underwent diagnostic and therapeutic surgical resection. A fronto-parietal craniotomy was performed, and the mass was excised. Intraoperative squash cytology revealed multiple tissue pieces measuring 0.5 cm x 0.4 cm x 0.3 cm, and smears were easily spread. The smears showed a moderately cellular lesion with ill-defined cell borders and long hair-like processes in a fibrillary background, with occasional eosinophilic granular bodies and Rosenthal fibers (Figure 2A). The squash cytology findings were suggestive of a low-grade glial tumor with piloid features. A biopsy was subsequently obtained, which revealed a moderately cellular tumor mass comprising bipolar piloid cells arranged in a diffuse pattern in a fibrillary background, along with loose hypocellular edematous areas showing eosinophilic granular bodies, occasional Rosenthal fibers, and calcification in the form of a fair number of psammoma bodies. The histological findings were suggestive of PA CNS WHO Grade 1 (Figure 2B). Immunohistochemistry (IHC) revealed IDH negativity, SOX10 positivity, p16 positivity, and an MIB-1 labeling index of 1-2%. The diagnosis was confirmed as pilocytic astrocytoma on IHC.

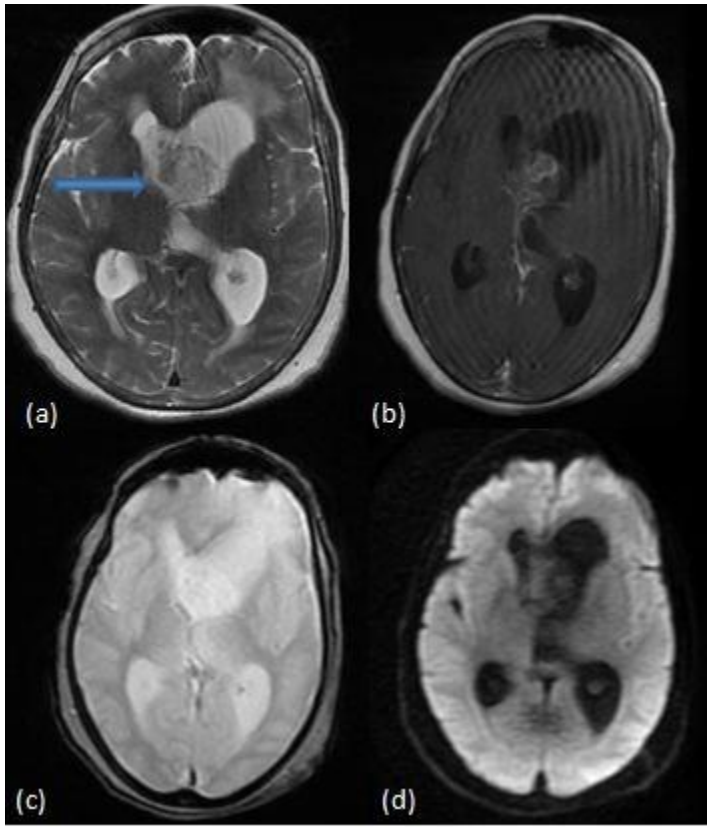


**Figure 1.** T2W axial MR image shows T2 hyperintense lesion in left frontal horn causing compression of bilateral foramen of Monroe with dilatation (arrow) of bilateral lateral ventricles (a); post-contrast T1W axial MR image shows the lesion showing heterogenous enhancement (b); GRE axial MR image shows no blooming in the lesion (d); diffusion-weighted MR image shows no diffusion restriction (d).

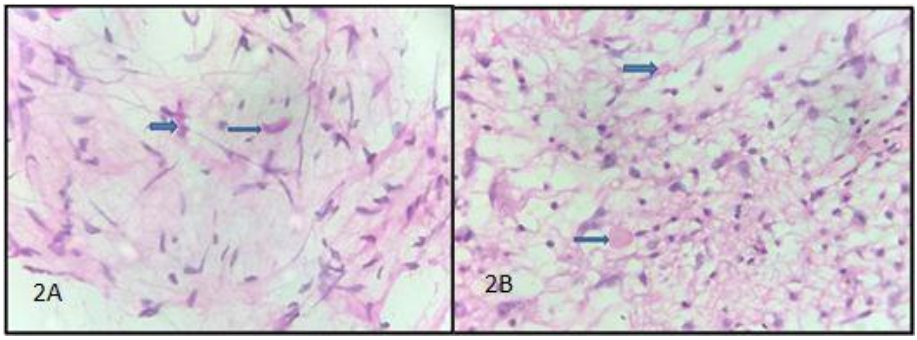


**Figure 2.** Squash cytology smear shows bipolar cells with long hair like piloid processes dispersed in fibrillary background with occasional Rosenthal fiber (arrows) (2A, HE stain, x40); histology tissue section shows bipolar piloid cells in fibrillary background surrounded by loose areas with occasional eosinophilic granular bodies (arrows) (2B, HE stain, x40).

**Treatment outcome:** The patient showed significant improvement in her clinical condition on subsequent follow-up for six months.

*2.2 Case 2*

**Patient demographics and clinical presentation:** A 16-year-old male patient presented to the neurosurgery outpatient department with a complaint of gradual blurring of vision and headache that had been present for two months. The headache was persistent but did not cause loss of consciousness. The patient had also experienced left upper and lower limb paresis for one month. Cranial nerve examination and bilateral pupil examination were normal. The patient's Glasgow coma scale was E4M5V4, indicating disorientation to time, place, and person. The patient had spontaneous eye opening and moved to localize pain.

**Imaging findings:** MRI revealed an intraventricular space-occupying lesion involving the body and trigone of the right lateral ventricle, suggestive of ependymoma. A right fronto-parietal craniotomy was performed, and the mass was excised.

**Pathological findings:** Blood investigations were normal. A fronto-parietal craniotomy was performed with excision of the mass. Intraoperative squash cytology revealed bipolar piloid cells in a fibrillary background along with occasional eosinophilic granular bodies, suggesting a low-grade glial tumor. Histopathological examination revealed alternating hypercellular areas comprising bipolar cells with hair-like processes arranged in a fibrillary background along with hypocellular areas showing eosinophilic granular bodies and numerous psammoma bodies (Figure 3). The histological features were consistent with circumscribed glioma, pilocytic astrocytoma, CNS WHO Grade 1.

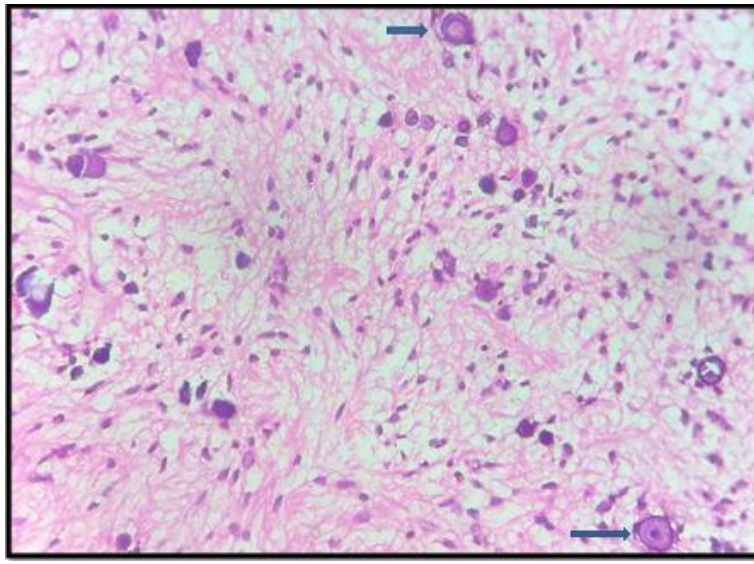

**Figure 3.** Histology tissue section shows bipolar piloid cells in fibrillary background with fair number of psammoma bodies (arrows) (HE stain, x40).

**Treatment outcome:** The patient showed improvement in vision and gradually regained power in both limbs postoperatively. There was no recurrence over the six-month follow-up period.

*2.3 Case 3*

**Patient demographics and clinical presentation:** A 12-year-old male patient was admitted to the neurosurgery ward with complaints of headache and vomiting for the past three months, along with gradual vision loss. The vomiting was projectile and not associated with nausea. The headache was persistent without loss of consciousness. There was no history of trauma. Cranial nerve examination was normal. The Glasgow coma scale was E4M6V5; the patient was well oriented and moving all four limbs with spontaneous eye opening.

**Imaging findings:** MRI revealed a large lobulated lesion in the suprasellar region. The lesion was hypointense on T2/FLAIR and T1-weighted sequences and exhibited heterogeneous contrast enhancement. The lesion measured 3.7 cm x 2.3 cm x 2 cm and completely obliterated the third ventricle. These findings suggested a suprasellar mass lesion, possibly a craniopharyngioma (Figure 4).

**Pathological findings:** Blood investigations were normal. The patient was scheduled for diagnostic and therapeutic resection. A fronto-parietal craniotomy was performed, and the mass was excised. Intraoperative squash cytology revealed multiple

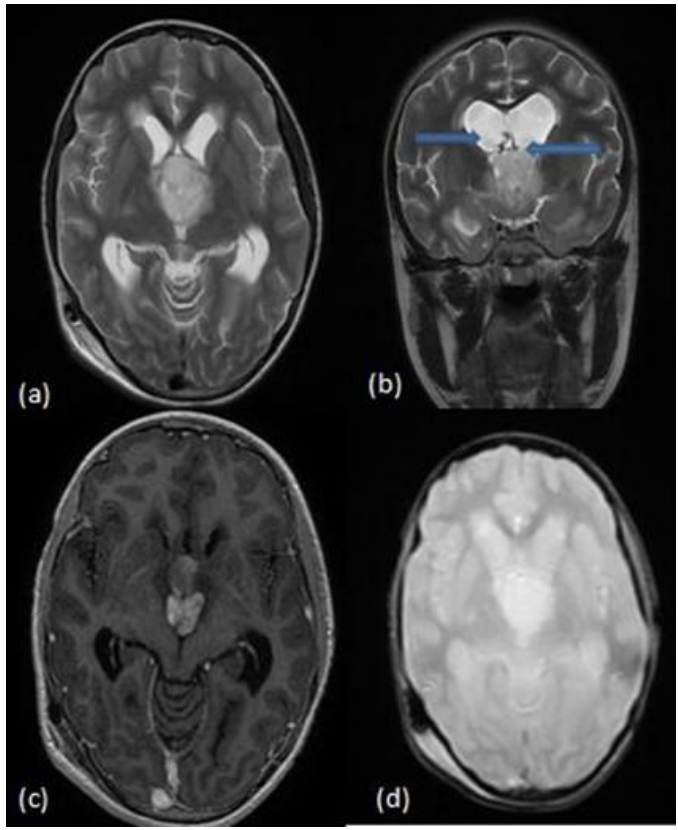


**Figure 4.** T2W axial and T2W coronal MR images show T2 hyperintense lesion in third ventricle (arrow) causing compression of bilateral foramen of Monro with dilatation of bilateral lateral ventricles (a-b); post-contrast T1W axial MR image shows the lesion showing heterogenous enhancement (c); GRE axial MR image shows no blooming in the lesion (d).

HE- and PAP-stained smears showing bipolar cells with long hair-like processes in a fibrillary background, along with a few Rosenthal fibers and foci of calcification. These findings were suggestive of a low-grade glial tumor with piloid features. Subsequent histopathological examination of the biopsy tissue showed compact areas of bipolar piloid cells arranged in a fibrillary background alternating with loose areas containing plenty of eosinophilic granular bodies and Rosenthal fibers (Figure 5). Histological findings were consistent with pilocytic astrocytoma CNS WHO Grade 1.

**Treatment outcome:** The patient recovered well in the postoperative period, and no recurrence or complication was noted over a six-month follow-up period.

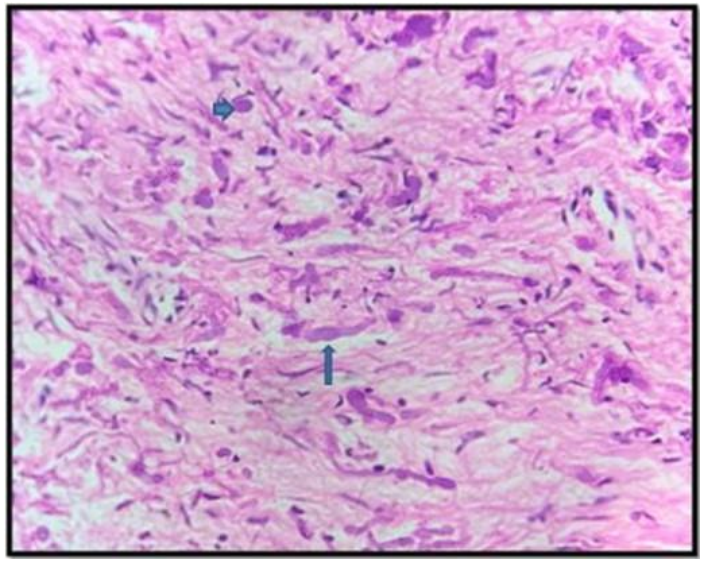

**Figure 5.** Histology tissue section shows bipolar piloid cells arranged in diffuse pattern in fibrillary background along with plenty of Rosenthal fibers (arrow) and eosinophilic granular bodies (arrowhead) (HE stain, x40).

## 3. Discussion

Intraventricular tumors can originate from the ependymal lining, subependymal plate of the ventricular wall, choroid plexus, or glial structures lining the septum pellucidum [6]. The differential diagnoses for intraventricular tumors include choroid plexus papilloma, central neurocytoma, ependymoma, subependymal giant cell astrocytoma, and low-grade glial tumors, including pilocytic astrocytoma [7]. Other less common intraventricular tumors include meningioma, lymphoma, and metastasis. Choroid plexus papilloma consists of true papillae with fibrovascular cores lined by cuboidal to columnar epithelium. Central neurocytoma comprises a monomorphic population of oligodendrocyte-like cells. Ependymoma exhibits prominent perivascular pseudorosettes. Subependymal giant cell astrocytoma displays giant pyramidal-like cells with a ganglionic appearance and is associated with tuberous sclerosis. Meningioma shows syncytial sheets, whorls, and lobules of meningothelial cells with intranuclear pseudoinclusions. Lymphoma histology reveals a monomorphic population of atypical lymphoid cells dispersed singly. However, pilocytic astrocytoma exhibits bipolar piloid cells arranged in a diffuse fibrillary background along with Rosenthal fibers and eosinophilic granular bodies.

Pilocytic astrocytoma is commonly seen in the pediatric age group with a mean age of presentation of 12 years, and its incidence declines with age [8]. It is very rare in adults, with only a few cases reported in the literature [8]. Therefore, these tumors often remain misdiagnosed and overlooked. One of the cases in our series involved a 64-year-

**Table 1.** Summary of clinical cases.

| Characteristics | Case 1 | Case 2 | Case 3 |
|---|---|---|---|
| Age in years/sex | 64/female | 16/male | 12/male |
| Symptoms | Headache, loss of memory, loss of bowel and bladder control | Blurring of vision, headache, left upper and lower limb paresis | Headache, vomiting and gradual vision loss |
| Radiological findings | Central neurocytoma | Ependymoma | Craniopharyngioma |
| Squash cytology findings | Low grade glial tumor with piloid features | Low grade glial tumor | Low grade glial tumor with piloid faetures |
| Histopathological findings | Pilocytic astrocytoma (CNS WHO Grade 1) | Pilocytic astrocytoma (CNS WHO Grade 1) | Pilocytic astrocytoma (CNS WHO Grade 1) |
| Immunohistochemistry findings | IDH negative, SOX10 positive, p16 positive, MIB labeling index 1-2% | Not available | Not available |
| Treatment outcome | Good | Good | Good |
| Six months follow-up | No recurrence and complications | No recurrence and complications | No recurrence and complications |

old patient with intraventricular pilocytic astrocytoma. Intraventricular pilocytic astrocytoma is extremely rare, accounting for approximately 4-15.6% of all pilocytic astrocytomas [9]. PA typically presents as a cystic tumor with a mural nodule and may be vascular. It sometimes presents with extensive calcification as a degenerative phenomenon, as seen in long-standing low-grade tumors. Calcification may be very extensive and may sometimes obscure the diagnosis of pilocytic astrocytoma. Therefore, care should be taken when reporting intraventricular tumors with extensive calcification.

By causing mechanical obstruction, ventricular tumors produce signs and symptoms of raised intracranial pressure, such as headache, blurring of vision, vomiting, and focal neurological deficits, with seizures as long-term complications. The usual age of presentation of PA is childhood, and it is very uncommon to find PA after 50 years of age [10]. One of the cases in our series presented at 64 years of age, which is a very rare occurrence (Table 1). Additionally, none of our cases were diagnosed as pilocytic astrocytoma on radiology preoperatively. This is important for proper intraoperative management of patients with highly vascular intraventricular PA, who may experience profuse blood loss complications [5]. However, the limitation of our study was that immunohistochemistry (IHC) was available in only one of the cases. As IHC is more confirmatory, its availability in all cases would have enhanced the diagnostic reliability of the cases. Increasing the sample size and incorporating more extensive IHC markers would have been beneficial in addressing these limitations.

## 4. Conclusion

Limited literature is available on the occurrence of PA at an unusual age or in an unusual intraventricular location. It is important to consider IVPA in adult patients with calcified intraventricular lesions. As these lesions can often be missed on radiology and can lead to misdiagnosis in pathology, patients may experience intraoperative complications of massive blood loss in extremely vascular IVPA cases if preoperative arrangements are not made to control bleeding.

## Acknowledgment

Acknowledgment is extended to all faculty members and residents of the neurosurgery department, NSCB Medical College, Jabalpur, India for their contribution of samples for the study. The invaluable comments and recommendations from the anonymous reviewers are gratefully appreciated.

## Conflict of Interest Statement

The authors declare no conflict of interest.

**Author Contributions:** Design conception, data collection, data analysis, and preparation of draft, Diya Bajaj (D.B.); Design conception, data collection, data analysis, and preparation of draft, A.G.; Data collection and data analysis, N.Y.; Design conception, data analysis, and preparation of draft, J.B.; Design conception and data analysis, S.R. All authors have read and agreed to the published version of the manuscript.

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