





Original Article

# Intraventricular Pilocytic Astrocytoma in Adults: A 25-year Single-Center Case Series and Systematic Review of the Literature

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## Background

Pilocytic astrocytomas (PA) are the most common gliomas in children/adolescents but are less common and poorly studied in adults. Here, we describe the clinical presentation, surgical management, and outcomes of surgically treated adult patients with intraventricular (IV) PA and review the literature.

## Methods

Consecutive adult patients treated for IV brain tumors at a tertiary academic center over 25 years (1997–2023) were identified. Clinical data were reviewed retrospectively for adult IV PA patients. A systematic literature review was conducted following the Preferred Reporting Items for Systematic Reviews and Meta-analyses guidelines.

## Results

Eight patients with IV PA were included. Median age was 25 years (range, 18–69 years), and 4 (50%) were female. The most common tumor location was the lateral ventricle (5, 63%), followed by the fourth ventricle (3, 37%). Subtotal and near total resection were the most common surgical outcomes (6 patients, 75%), followed by gross total resection in 2 (25%). Progression or recurrence occurred in 3 patients (37%), requiring repeat resection in 2 patients. The 5-year overall survival and progression-free survival were 67% and 40%, respectively. In addition, 42 cases were identified in the literature.

## Conclusions

PAs in adults are rare and an IV location is even more uncommon. The findings demonstrate the challenges in caring for these patients, with overall- and progression-free survival outcomes being poorer than the general adult PA population. Findings support the employment of surgical techniques and approaches that favor gross total resection when possible. Further studies are needed to better characterize this unique presentation.

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## Introduction

Pilocytic astrocytomas (PA) are the most common gliomas in children and adolescents, but they can also arise in adults. The World Health Organization classifies PAs as grade 1 tumors with characteristically benign, slow, and well-circumscribed growth patterns.<sup>1</sup> Infrequently, they invade surrounding structures or show malignant behavior such as anaplasia, dissemination, or local recurrence. PAs account for approximately 5%–6% of all gliomas, and amongst all brain tumors, their proportion declines from 11.4% (in pediatric patients) to 0.8% in patients older than 19 years.<sup>2</sup> In the United States in 2019, 1069 patients were diagnosed with PAs, comprising 1.2% of all brain tumors, most cases were in patients less than 15 years old (62%), with fewer occurring between the ages of 15 and 39 (27%) and rarely occurring after the age of 40 (11%).<sup>3</sup> Previous reports suggest that PAs in adults tend to be more aggressive compared to pediatric cases, with a higher recurrence rate and an increased incidence of malignant transformation.<sup>2,4, 5, 6</sup> However, the literature focused on adult patients with PAs is less well-defined due to the rare nature of this diagnosis, and significant gaps remain in our understanding of the clinical presentation, management, and outcomes.

Most PAs arise near the midline, with the cerebellum being the most common location and, less frequently, the optic pathway, hypothalamic area, diencephalon, or brain stem.<sup>7</sup> Intraventricular (IV) PAs are rarely encountered, and the literature describing outcomes after this presentation is limited. Of note, IV tumors present significant surgical challenges due to their proximity to critical brain structures and difficult-to-access anatomic locations.<sup>8</sup> Although technological advances have added tools for the resection of supratentorial IV tumors achieving gross total resection (GTR) is still challenging.<sup>9,10</sup> Furthermore, fourth ventricular tumors are challenging to treat surgically due to the limited availability of approaches and/or techniques.<sup>11</sup>

The preferred treatment for PAs is GTR, which is commonly curative. However, achieving GTR in patients with IV PAs is frequently challenging, and the morbidity associated with incomplete resection of PAs is considerable. Hence, we aim to describe the clinical presentation, surgical management, and clinical outcomes of surgically treated adult patients with IV PAs in a consecutive, single-center case series. Additionally, we performed a systematic review of the literature to summarize the current evidence on the management of adult patients with IV PAs.

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## Section snippets

### Study Design and Inclusion Criteria

We performed a retrospective review of consecutive patients treated for intracranial IV tumors at a tertiary academic center in the United States from January 1997 to March 2023. The institutional review board approved this study (# 202210136), and the need for informed consent was waived due to its retrospective nature. An initial list of medical records was obtained using diagnostic codes specific to IV brain tumors. Inclusion criteria were: (a)  $\geq 18$  years, (b) radiologic confirmation of an...

## Patient and Tumor Characteristics

Over the last 25 years, 8 cases who were surgically managed for IV tumors at our center were found to have PA by histopathological diagnosis and were included in this study (Figure 1). The clinical and tumor characteristics of all cases are described in Table 1. Median age was 25 years (range, 18–69 years). The male-to-female ratio was 1:1. The most common clinical presentation was headache (4 cases, 50%), followed by seizures in 2 cases (25%). Of note, 3 cases (38%) presented with imaging...

## Discussion

In this 25-year single-center experience, 8 adult patients with IV PAs underwent surgical resection. Craniotomy was more common than endoscopy. Patients with supraventricular tumors were more likely to get a transcortical approach than an interhemispheric approach. For fourth ventricular tumors, 3 different surgical approaches were employed (paramedian, telovelar and transvermian). GTR was achieved only on 2 patients (25%), with 6 patients receiving STR/NTR (75%). Progression/recurrence...

## Conclusions

Overall, these results and literature review demonstrate the challenges that exist in caring for adult IV PA cases. The literature reflects that PAs in adults are uncommon, and IV locations are even rarer. Radiological features can be misleading, so consideration for this diagnosis is suggested in the differential for IV neoplasms. Our findings support a goal of GTR for IV PA tumors. Surgeons should select the surgical technique and approach that offer the greatest opportunity for GTR when...

## CRediT authorship contribution statement

**Timothy Woodiwiss:** Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Supervision, Validation, Writing – original draft, Writing – review & editing. **Juan Vivanco-Suarez:** Data curation, Investigation, Methodology, Validation, Visualization, Writing – original draft, Writing – review & editing. **Tyson Matern:** Data curation, Investigation, Supervision, Writing – original draft, Writing – review & editing. **Kathryn L. Eschbacher:** Data curation, Supervision, Writing –...

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Timothy Woodiwiss and Juan Vivanco-Suarez were contributed equally

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