LITERATURE REVIEW AND A SERIES OF CASES

# A monocenter retrospective study of the surgical outcomes of adult pilocytic astrocytoma: a small case series and review of the literature

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## Abstract

**Introduction:** The peak age for the development of pilocytic astrocytoma (PA), a type of benign cerebellar tumor, is between 10 and 20 years. Adult PA is extremely rare, and consequently, very little is understood about its characteristics.

**Methods:** We retrospectively reviewed the records of patients older than 18 years with pathologically proven PA who had surgery to remove the tumor between January 2010 and January 2020 and were followed until January 2022.

**Results:** Although 32 cases were initially flagged as PA, we included 4 patients (2 male and 2 female) with adult PA. The average age of a male patient at diagnosis was 26.75 years old, and there was no mortality or recurrence. The mean age of female patients at diagnosis was 25 years old. One female was still living after the follow-up period ended. The cause of death in one female patient was unrelated to tumor. Women had a median follow-up of 36 months, and their mean overall survival was 42 months.

**Conclusion:** PA in adults acts differently than in children. The extent of surgical resection and the location of the tumor influenced the prognosis. When possible, total resection should be the primary treatment, as it promotes good survival rates and low recurrence risk.

**Keywords:** astrocytoma; pilocytic astrocytoma; glioma; extent of resection

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## Introduction

According to the World Health Organization (WHO), pilocytic astrocytoma (PA) is a grade I tumor with a peak incidence in the first two decades of life and no gender bias. The cerebellum is the typical site of development for PA[1]. Adults PA, however, are extremely elusive[2].

Compact regions of bipolar astrocytes linked with Rosenthal fibers, protoplasmic astrocytes, microcysts, and eosinophilic granular masses are found on histology[1]. The benign and slow-growing nature of PA means that patients have a relatively good prognosis.

Surgery is the treatment of choice since it is curative in cases with complete resection. There is a wide range of reported survival rates, from 86% to 100% five years after surgery, and gross complete resection leads to improved survival. However, poor outcomes have been linked to incomplete resection and tumors in surgically inaccessible areas[3–5].

Even worse outcomes were seen in a recent study of PA patients in adulthood[2]. However, few studies have focused on adult PA patients, rendering the effect of age on prognosis unclear[2,6,7]. During the last decade, we have recorded 4 cases of PA in adults. The goal of this study was to examine the surgical outcomes of PA in adults and to identify potential influencing factors.

## Materials and Methods

The Center of Neurosurgery and Neurology cared for 32 patients with PA between 2010 and 2020. We did not include 28 patients in our study because they were either under 18 or had missing records. Four patients met our inclusion criteria and were examined retrospectively. We performed surgery on adults diagnosed with PA (astrocytoma grade I as determined via histopathology). Up until January 2022, their clinical data were reviewed. Patient demographics, clinical history, radiographic findings, operational particulars, tumor features, and pathology information were extracted. There was a general consensus that all cases presented with neurological complaints.

Preoperative magnetic resonance imaging (MRI) with and without gadolinium contrast enhancement was performed on all patients meeting these criteria. An MRI scan was performed within 72 hours after surgery in all operated cases. Based on pre-op volume (X) and post-op volume (Y), Extent of Resection (EOR) was calculated using the formula X-Y/X*100%. After obtaining EOR%, we classified, more than 99% as gross total resection (GTR), 91-99% as near-total resection (NTR), 70-90% as sub-total resection (STR), less than 70% as partial resection and as biopsy. As part of the follow-up for each patient, a series of MRI scans were acquired.

## Results

A total of 4 patients, including 2 males and 2 females, were included. Mean age of male at diagnosis was 26.75 years. At the end of follow-up, there was no male mortality and there was no recurrence. Mean age of females at diagnosis was 25.5 years. At the end of follow up period, 1 female was alive. Mean OS in females was 42 months, and there was recurrence in 1 female patient. According to Burkhard et al., while there is a minor preference for the supratentorial space in adults (55%), the cerebellum is the most common site for tumors in children (67%)[3]. PA occurred in cerebellum in 1 case and in the brainstem in 3 cases. Only data from resectable adult patients were analyzed based on preoperative imaging. Due to tumor location, a gross total resection (GTR) was attained in 1/4 of patients, and an NTR was achieved in 1/4 of patients. However, 2/4 patients attained partial resection.

Patients were followed for a median of 36 months after hospital release, with a range of 2 months to 144 months. From a total of 4 adults diagnosed with PA, 3 were clinically stable or improved after 42 months on average and 1 had passed away in that time. One of four patients experienced a recurrence. We anticipate that the survival rate reflects the overall positive prognosis of this tumor type, as demonstrated by previous studies.

## Discussion

Most commonly found in children and young adults, PA are slow-growing, benign, low-grade glioma tumors with a clear boundary[1,8]. PA is the most frequent glioma in children, comprising as many as 25% of all brain tumors removed through neurosurgical practice. However, PA is uncommon in adults, accounting for only 2.3% of all brain tumors[2], leading to a lack of knowledge regarding their features. This ten-year retrospective study discusses the demographics and treatment outcomes of a cohort of adult patients with PA treated at the Clinical Center of Neurosurgery and Neurology.

Recently, cases of PA in patients older than 30 years have been noted. A case was detected by Rossi et al. in the hypophysis of a man aged 40, and Kehler reported four cases in people aged 40 and up, albeit the specific details of these cases were not specified[9,10]. Of the 37 cases discussed by Lee et al., only one was older than 20 years[11]. Burkhard et al. reported a recent series of 55 participants, of whom 11 were 30 or older[3]. Only 4 of our 32 patients with PA were 18 or older.

While PA can appear anywhere along the neuraxis, it is most often found in midline structures such the cerebellum, optic chiasm, and brainstem. The cerebellum is the most common origin of PA, though in our series 1 case was located in the cerebellum and 3 were located in the brainstem. Patients presenting with PA have varying clinical manifestations depending on the size and site of the lesion. Our study found that the majority of people with posterior fossa PA also had difficulties walking due to symptoms of increased intracranial pressure, such as headaches, vomiting, and impaired vision.

Multiple studies have demonstrated that the location and extent to which a tumor was excised after surgery significantly impact prognosis. A univariable Cox proportional hazards regression analysis of 865 adult patients with PA from the USA National Cancer Institute Surveillance, Epidemiology, and End Results Program database indicated that gross total resection reduced the risk of death by a factor of 0.2 compared to subtotal resection or biopsy[12]. Similarly, Stüer et al. found that individuals whose tumors were only partially removed had a fourfold higher recurrence rate than those whose tumors were removed completely[2]. Total tumor removal is not only related to improved outcomes, but also curative. The patient in our study who underwent a total tumor removal had no tumor recurrence or death. Optic chiasm and brainstem PAs are associated with poorer outcomes due to the increased difficulty of access and complete excision. In our series, one patient with brainstem PA had a recurrence of their tumor, while another patient with brainstem PA passed away during follow-up.

Depending on the study, the prognosis for PA in adults can range from excellent to poor[2,3,8,12]. This is due to conflicting data showing either greater survival rates or worse prognoses and a higher likelihood of tumor recurrence following surgical excision. In the National Cancer Institute database study, survival rates decreased dramatically with age, from 96.5% at 5–19 to 52.9% at over 60 years of age[12].

Among 44 adult patients in a retrospective study, 5-year survival was 87% and progression-free survival (PFS) was 72%[2]. Brown et al. found PFS and OS rates of 95% after 5 years in a prospective study comprising 20 patients[12].

Based on the data we gathered, PA in adults may not be completely benign. One of four patients in our study experienced tumor recurrence and one died during the study period, with an overall survival rate of 42 months. The discrepancy between this study's findings and those of other investigations may be attributable, in part, to the small sample size of adult patients studied in this study.

## Conclusions

Adult PA is an uncommon tumor with a similar clinical presentation to that of childhood/adolescence. The standard of care appears to be surgical resection. The prognosis is favorable if entire resection can be performed. To confirm these preliminary findings, further studies are needed. Extent of resection and tumor site were shown to influence prognosis. We recommend careful monitoring and follow-up for adult PA patients.

## Disclosures

**Conflict of Interest:** All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

**Ethical approval:** All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This study was approved by the Research Ethics Committee, Faculty of Medicine, Neurosurgery Department, Uzhhorod National University.

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