SVOA Neurology

ISSN: 2753-9180

ScienceVolks Open Access

Research Article

Benign Pediatric Cerebellar Pilocytic Astrocytoma: A Single-Center Perspective

Mohsen Khamoushi Kahdouee[1], Amirhosein Haghir[1], Farhang Mirakhorli MD[1], Ali Sadeghzadeh Shiraz MD[2], Faezeh Hosseini MD[2], Samin Safarian MD[2], Amirmohammad Soleimanian MD[2], Seyed Ali Shariat Razavi MD[1], Neda Kamandi MD[2]*, Mohammadreza Rostami[3]

[1]Department of Neurosurgery, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran.

[2]Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran.

[3]Faculty of Medicine, Xi'an Jiaotong University, Xi'an, China.

***Corresponding Author:** Neda Kamandi, MD, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran.

https://doi.org/10.58624/SVOANE.2025.06.014

Received: November 11, 2024

Published: May 28, 2025

Citation: Kahdouee MK, Haghir A, Mirakhorli F, Sadeghzadeh Shiraz A, Hosseini F, Safarian S, Soleimanian A, Shariat Razavi SA, Kamandi N, Rostami M. Benign Pediatric Cerebellar Pilocytic Astrocytoma: A Single-Center Perspective. *SVOA Neurology* 2025, 6:3, 70-74. doi. 10.58624/SVOANE.2025.06.014

Abstract

Introduction: Astrocytomas of the cerebellum are non-malignant central nervous system tumors. They account for 10% of all brain tumors in children within the skull and 30% of all tumors in the back part of the brain in children, particularly in the first 20 years of life.

Methods: We retrospectively assessed the medical histories of patients, documenting their age at the time of operation, pre- and post-surgery MRI scans, tumor location, presence of hydrocephalus, surgical method, and neurological condition before and after surgery, post-surgery supplementary treatment, and the results.

Results: The ratio of males to females was 33:17. The age at the time of assessment varied from 3 to 19 years, while the age at the time of surgery ranged from 1 to 18 years. The follow-up period after surgery was between 1 and 12 years. In twenty-eight patients, the tumor was located in the cerebellar vermis, whereas 22 harbored hemispheric cerebellar astrocytoma. A total of 76% of these patients underwent complete surgical removal of the tumor. Nine patients required a second surgery due to remaining tumor tissue. A ventriculoperitoneal shunt was inserted in 5 out of 50 patients due to symptomatic hydrocephalus, and three patient underwent an endoscopic third ventriculostomy before surgery.

Conclusion: Astrocytomas in the cerebellum are non-malignant tumors, and in most instances, they can be entirely removed through surgery, leading to highly favorable results.

Keywords: *Pilocytic astrocytoma, Cerebellum, Brain tumor, Children, Astrocytomas*

Introduction

In children, juvenile pilocytic astrocytoma located in the cerebellum is generally recognized as a cyst-like growth with a solid section, often resulting in a positive outcome following surgical removal [1, 2]. Magnetic resonance imaging (MRI) has been instrumental in diagnosing and improving the assessment of treatment outcomes [3, 4]. Additionally, it has facilitated the identification of various subtypes of low-grade astrocytomas that differ in radiological characteristics and anatomical positioning from the traditional variant [1, 3, 5]. The purpose of this research is to investigate if the varying positions (either in the hemisphere or vermis) or consistencies (solid versus cystic) of cerebellar pilocytic astrocytomas, despite having the same histopathological characteristics, have any significance in a clinical context.

Materials and Methods

Our study retrospectively analyzed 50 individuals who underwent surgery for cerebellar pilocytic astrocytomas at our facility between January 2015 and March 2024. Each patient was required to have MRI scans before and after the operation, along with a confirmed diagnosis of cerebellar pilocytic astrocytoma. We examined the patients' medical histories, focusing on their age during surgery, MRI scans before and after surgery, tumor location, the occurrence of hydrocephalus, the method of surgery, neurological condition before and after surgery, any additional treatments following surgery, and the final results. Patients were monitored post-surgery at intervals of 1, 3, and 6 months, annually after the first year, and then once every year thereafter.

The study reassessed the tumor's position (either in the vermis or the cerebellar hemisphere) and its consistency (whether solid or cyst-like) based on these tests. All pathological test results were examined, and instances of anaplastic astrocytoma and glioblastoma multiforme were omitted from the research. Tissue samples fixed in formalin and embedded in paraffin were used to create microscopic slides. These sections, which were 4 micrometers thick, were dyed with hematoxylin and eosin. Additionally, every case underwent immunohistochemical staining to detect synaptophysin, glial fibrillary acidic protein, neuronal nuclear antigen, and Ki-67.

Results

This investigation included 50 children who fulfilled the study's eligibility requirements: they were diagnosed with cerebellar astrocytoma and had undergone contrast-enhanced MRI before surgery, as well as regular MRI monitoring for a minimum of 12 months post-surgery. The gender distribution was 33 males to 17 females. The ages at assessment varied from 3 to 19 years, with an average of 13.46 years (standard deviation ±4.26 years), while the ages at surgery ranged from 1 to 18 years, with an average of 7.89 years (standard deviation ±4.82 years). The follow-up period spanned from 1 to 12 years, averaging 4.8 years (standard deviation ±2.77 years). Headaches were the most frequently reported initial symptom, affecting 78% of patients. Additional symptoms at the onset included vomiting (72%), difficulty walking (48%), neck stiffness (22%), double vision (8%), visual problems (6%), and developmental delays (6%). Initial neurological assessments revealed papilledema in 82% of patients, ataxia in 58%, nystagmus in 30%, dysmetria in 22%, speech difficulties in 4%, and impaired rapid alternating movements in 4%. Symptoms had been present for a duration ranging from 10 days to eight months. A summary of the initial symptoms and neurological observations can be found in Table 1.

Table 1. Patient's presenting signs and symptoms

Presentation	Patients (%)
Symptoms	
Headache	39 (78%)
Vomiting	36 (72%)
Gait disturbance	24 (48)
Neck stiffness	11 (22%)
Diplopia	4 (8%)
Visual impairment	3 (6%)
Developmental delay	3 (6%)
Signs	
Papilledema	41 (82%)
Ataxia	29 (58%)
Nystagmus	15 (30%)
Dysmetria	11 (22%)
Dysarthria	2 (4%)
Dysdiadochokinesia	2 (4%)

In the MRI scans, solid tumors were found in 13 patients, which is 26% of the total, while cystic tumors with a mural nodule were identified in 23 patients, making up 74% of the total. Pilocytic astrocytomas were present in the vermis of 28 patients (56% of the total), with 22 of these patients having cystic tumors and the remaining six having solid tumors in the vermis. Brain MRIs revealed hemispheric lesions in 24 patients (48% of the total), with 17 of these lesions (70.8%) on the left and 7 (29.2%) on the right. Among these 24 patients, 18 (75%) had cystic cerebellar tumors with a mural nodule, and 6 (25%) had solid cerebellar tumors.

The World Health Organization (WHO) has a system for categorizing tumors of the central nervous system (CNS), which includes cerebellar tumors. The WHO criteria state that tumors displaying a two-phase structure, consisting of densely packed bipolar cells and more dispersed multipolar cells, along with Rosenthal fibers and eosinophilic granular bodies, are classified as pilocytic astrocytoma, which is a WHO grade 1 tumor. The presence of Glial fibrillary acidic protein (GFAP) is particularly notable in the bipolar cells. Typically, the Ki-67 labeling index, which indicates cell proliferation, is less than 1%.

Before surgery, 80% of the patients, amounting to 40 individuals, were diagnosed with hydrocephalus. Nine patients experienced lethargy as a result of obstructive hydrocephalus and underwent surgical intervention. Of these, five had a ventriculoperitoneal shunt placed, while three patients were treated with endoscopic third ventriculostomy. Following the operations, no patients needed surgical intervention for hydrocephalus.

During the procedures, patients were positioned upright. For tumors located in the vermis, a suboccipital craniotomy was conducted without removing the C1 arch. For tumors in the hemispheres, a suboccipital paramedian cut was made on the affected side. The goal was to completely remove the tumor in all cases, except for four where the cerebellar peduncle was involved. Complete removal was achieved in 38 patients (76%), while a partial resection was performed in 12 patients (24%). The presence of any remaining tumor was detected in the early postoperative MRI scans.

In the period following surgery, there was no ongoing occurrence of hydrocephalus. Out of 12 patients, nine required a second surgery because the remaining tumor had grown, and in these cases, the tumor was completely removed. Three other patients, who had not had the entire tumor removed in the initial surgery, received radiotherapy afterwards. Early complications after the surgery included ataxia in 18% of patients, difficulty swallowing in 12%, mutism also in 8%, and cerebrospinal fluid (CSF) leakage in 8%. There were no fatalities following the surgery.

Discussion

Multiple factors influence patient outcomes following treatment, including the thoroughness of the tumor removal, whether the brain stem was affected, instances of tumor return, the effectiveness of radiotherapy, and the tumor's specific location [1, 6, 7]. Advances in diagnostic imaging and surgical methods have led to better rates of morbidity and mortality [3, 6]. It is widely accepted among researchers that fully removing the tumor is a critical determinant of the likelihood of recurrence and the survival rate [1, 2, 7, 8]. Gjerris et al. [9] reported a 94% survival rate, while Zakrzewski et al. [10] documented an 88% survival rate over a span of 25 years. Despite this, there have been occurrences of cerebellar astrocytomas reappearing even after being fully excised [2, 11, 12]. In our study, complete tumor removal was achieved in 76% of cases during the initial surgery, and these patients remained free of tumors for an average mean follow-up of 4.8 years.

Typically, identifying cerebellar pilocytic astrocytomas is a prolonged process due to the tumor's gradual growth. The initial symptoms mainly stem from hydrocephalus, which occurs when the tumor blocks the flow of cerebrospinal fluid (CSF), and from issues associated with the tumor invading cerebellar tissue. An analysis of the initial symptoms and neurological assessments showed no notable distinction between symptoms caused by hydrocephalus and those caused by cerebellar invasion, regardless of the tumor's location or how it appears on radiological scans [7, 8].

Austin et al. [11] reported a 23% recurrence rate of tumors in their study, while Due-Tonnessen et al. [13] found that 27% of their patients experienced growth in the remaining tumor. In our own research, we did not observe any tumor regrowth, although 12 out of 50 patients had some tumor left after the initial surgery. Of these, nine underwent a second surgery to remove the remaining tumor, which was completely excised as confirmed by subsequent imaging. There have also been cases reported where the leftover tumor shrank on its own after the first surgery. The strategy for managing these patients is based on several factors: the size of the remaining tumor as seen on postoperative MRI, the patient's overall health, and the potential risks associated with another surgery [13-15].

We opt for a second surgery when the tumor can be safely removed, but in cases where there is a significant risk of complications, such as with patients whose brain stem is affected, we prefer to monitor them with regular MRI scans and clinical assessments.

The literature presents mixed views on the benefits of radiation therapy. Most studies suggest that the long-term outcomes for patients who undergo only surgery are not significantly different from those who receive both surgery and radiation therapy [6, 16, 17]. There are concerns about the adverse effects of radiotherapy on the developing brain, including reports of low-grade cerebellar astrocytomas becoming malignant after radiation treatment [17, 18]. In our study, four patients received postoperative radiotherapy because their tumors were not fully removed during the initial surgery due to involvement of the brain stem. We did not see any enhancement in prognosis following radiotherapy, as reported by Sgouros et al. [19], although our sample size was too limited to draw statistical conclusions. Our preference is to opt for a second surgical procedure when feasible, to avoid the potential hazards associated with radiotherapy.

In our study, MRI scans revealed that 13 patients presented with solid tumors, while 37 had cystic tumors. Focusing on the tumor's location, 28 were found in the cerebellar vermis. Within this group, 22 patients exhibited a cystic tumor in the midline with a mural nodule, and six had a solid tumor in the vermis. Additionally, brain stem involvement was noted in four of the six patients with a solid vermian tumor. Post-operative radiotherapy was administered to four patients due to remaining tumor tissue. The other patients had tumors in the cerebellar hemispheres; of these, 16 had cystic and six had solid tumors. A second surgery was performed on five patients to address residual tumors. We achieved a surgical mortality rate of 0% and were able to completely remove the tumor in 90% of the cases through a combination of initial and follow-up surgeries.

Overall, patients experienced a higher rate of complete tumor removal when the tumor had not spread to the brain stem or the base of the fourth ventricle. Our findings suggest that cystic tumors tend to have better outcomes than solid ones, particularly as the involvement of the brain stem can increase complications associated with solid tumors.

The debate over whether to fully remove cystic lesions is ongoing. Sgouros et al. [19] found little difference in outcomes between complete and partial removals of cystic tumors. Conversely, Pencalet et al. [20] advocate for the removal of any visibly abnormal cyst walls to prevent recurrence. In our cases, we achieved complete resection of all cystic tumors, especially removing the cyst wall when preoperative MRI showed contrast enhancement.

While there have been instances of cerebellar astrocytomas leading to leptomeningeal spread and malignant changes as noted in the literature, such complications did not occur in any of the cases in our study [14, 21].

While the long-term survival rates for our patient group remain unknown, it appears that those with cystic tumors generally have better outcomes than those with solid tumors, regardless of where the tumors are located. Tumors in the vermis more frequently resulted in permanent neurological damage. The most significant predictor of a less favorable prognosis was involvement of the brain stem. Preoperative shunting for hydrocephalus was not deemed necessary in every case.

Conclusion

To sum up, after an average follow-up of 4.8 years, there was no clear distinction in outcomes between patients with solid versus cystic pilocytic tumors, or between those with tumors in the cerebellar hemispheres versus the vermis. Although complete tumor removal was less likely in isolated cases, its impact on long-term prognosis remains uncertain.

Ethical Consideration

Written informed consent was obtained from the parents or legal guardians before the procedure. Because the data were collected retrospectively, there was no need for patients' consent. Moreover, the institutional review board (IRB) of our medical school approved the study protocol.

Funding

None.

Conflict of Interest

The authors declare no conflict of interest.

References

1. Desai KI, Nadkarni TD, Muzumdar DP, Goel A. Prognostic factors for cerebellar astrocytomas in children: a study of 102 cases. Pediatr Neurosurg. 2001;35(6):311-7.
2. Bonfield CM, Steinbok P. Pediatric cerebellar astrocytoma: a review. Child's Nervous System. 2015;31:1677-85.
3. Gol A, McKissock W. The cerebellar astrocytomas: a report on 98 verified cases. J Neurosurg. 1959;16(3):287-96.
4. Ilgren E, Stiller C. Cerebellar astrocytomas: therapeutic management. Acta neurochirurgica. 1986;81:11-26.
5. Viano JC, Herrera EJ, Suárez JC. Cerebellar astrocytomas: a 24-year experience. Child's nervous system. 2001;17:607-10.
6. Garcia DM, Marks JE, Latifi HR, Kliefoth AB. Childhood cerebellar astrocytomas: is there a role for postoperative irradiation? International Journal of Radiation Oncology* Biology* Physics. 1990;18(4):815-8.
7. Lapras C, Patet J, Lapras C, Mottolese C. Cerebellar astrocytomas in childhood. Child's Nervous System. 1986;2:55-9.
8. Schneider Jr JH, Raffel C, McComb JG. Benign cerebellar astrocytomas of childhood. Neurosurgery. 1992;30(1):58-63.
9. Gjerris F, Klinken L. Long-term prognosis in children with benign cerebellar astrocytoma. J Neurosurg. 1978;49(2):179-84.
10. Zakrzewski K, Fiks T, Polis L, Liberski PP. Posterior fossa tumours in children and adolescents. A clinicopathological study of 216 cases. Folia Neuropathol. 2003;41(4):251-2.
11. Austin EJ, Alvord EC. Recurrences of cerebellar astrocytomas: a violation of Collins' law. J Neurosurg. 1988;68(1):41-7.
12. Pagni CA, Giordana MT, Canavero S. Benign recurrence of a pilocytic cerebellar astrocytoma 36 years after radical removal: case report. Neurosurgery. 1991;28(4):606-9.
13. Due-Tønnessen BJ, Helseth E, Scheie D, Skullerud K, Aamodt G, Lundar T. Long-term outcome after resection of benign cerebellar astrocytomas in children and young adults (0–19 years): report of 110 consecutive cases. Pediatr Neurosurg. 2002;37(2):71-80.
14. Dirven C, Mooij J, Molenaar W. Cerebellar pilocytic astrocytoma: a treatment protocol based upon analysis of 73 cases and a review of the literature. Child's nervous system. 1997;13:17-23.
15. Smoots DW, Geyer JR, Lieberman DM, Berger M. Predicting disease progression in childhood cerebellar astrocytoma. Child's Nervous System. 1998;14:636-48.
16. Ferbert A, Gullotta F. Remarks on the follow-up of cerebellar astrocytomas. J Neurol. 1985;232:134-6.
17. Kleinman GM, Schoene WC, Walshe TM, Richardson EP. Malignant transformation in benign cerebellar astrocytoma: Case report. J Neurosurg. 1978;49(1):111-8.
18. Scott RM, Ballantine HT. Cerebellar astrocytoma: malignant recurrence after prolonged postoperative survival: Case report. J Neurosurg. 1973;39(6):777-9.
19. Sgouros S, Fineron PW, Hockley AD. Cerebellar astrocytoma of childhood: long-term follow-up. Child's Nervous System. 1995;11:89-96.
20. Pencalet P, Maixner W, Sainte-Rose C, Lellouch-Tubiana A, Cinalli G, Zerah M, et al. Benign cerebellar astrocytomas in children. J Neurosurg. 1999;90(2):265-73.
21. Morikawa M, Tamaki N, Kokunai T, Nagashima T, Kurata H, Yamamoto K, et al. Cerebellar pilocytic astrocytoma with leptomeningeal dissemination: case report. Surg Neurol. 1997;48(1):49-51.