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SNI: Neuro-Oncology

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Review Article

Cerebellopontine angle pilocytic astrocytoma in adults: A systematic review

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Received: 03 July 2024
Accepted: 10 September 2024
Published: 04 October 2024

DOI
10.25259/SNI_539_2024

Quick Response Code:

ABSTRACT

Background: In adults, the cerebellopontine angle (CPA) pilocytic astrocytoma (PA) is very rare. This tumor has radiological features similar to those of a vestibular schwannoma in the few cases reported in the literature.

Methods: In this study, we conducted a systematic review in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses protocol and scrutinized all original studies pertaining to pontocerebellar angle PA in adult patients. We conducted an analysis of the clinical, radiological, and molecular components of all eligible articles. We have also reported a case involving a 67-year-old male individual in whom the PA exhibited radiological characteristics similar to an epidermoid cyst.

Results: After the screening phase, we found four cases of PA of the pontocerebellar angle. Three cases were identified that resembled vestibular schwannoma; however, in our case, the tumor resembled an epidermoid cyst. These uncommon tumors exhibit distinctive histological patterns and molecular characteristics (adenosine triphosphate dependent helicase (ATP- dependent helicase)+, Isocitrate dehydrogenase 1−), rendering them a potential differential diagnosis for glioblastoma (GBM).

Conclusion: The CPA PA has rarely been found in adult patients and should be considered in the differential diagnosis of vestibular schwannoma and epidermoid cysts. In these rare cases, the histological characteristics of PA are significant for the differential diagnosis of GBM.

Keywords: Cerebellopontine angle, Epidermoid cyst, Molecular markers, Pilocytic astrocytoma, Tumor

INTRODUCTION

Pilocytic astrocytomas (PAs) account for 5–6% of all brain tumors and are one of the most common primary brain tumors in children.[17] The cerebellum is the most frequently encountered localization site (40%), followed by supratentorial locations (35%). Other prevalent locations for the occurrence of these tumors include the optic pathway, hypothalamus, and brain stem, which account for approximately 9% of the cases, and the spinal cord.[6] This type of tumor is

extremely rare in adults, especially when located in the cerebellopontine angle (CPA). In this region, it may present as either a primary or secondary PA. Numerous researchers propose that primary PA tumors originate from the root entry zone of the cranial nerves, specifically the VII (facial) or VIII (vestibulocochlear) nerves.[2,8,17] Secondary PA tumors in the CPA typically arise from the cerebellum or brainstem, displaying exophytic growth that extends into the CPA.[2] In addition, they may mimic other types of tumors, such as vestibular schwannoma, epidermoid, dermoid tumors, and Glioblastoma (GBM) of CPA. They are substantially more prevalent than the preceding ones. PA of the CPA presents significant diagnostic challenges, as they can mimic various other tumor types, including vestibular schwannomas, epidermoid and dermoid tumors, and GBMs of the CPA. Despite the advancements in imaging technologies, radiological diagnosis remains difficult, often leading to an incorrect diagnosis.[4,8] In this article, we conducted a systematic review of the prevailing literature regarding PA localized in the CPA in adult patients. Our objective is to delineate the distinctive characteristics that distinguish PA from other lesions in the CPA. In addition, we report a rare case of PA in an adult who was misdiagnosed preoperatively as an epidermoid cyst based on neuroimaging findings. This case underscores the diagnostic complexities and the need for increased awareness and consideration of PA in differential diagnoses for CPA lesions in adults.

MATERIALS AND METHODS

The protocol of the systematic review presented herein was prepared in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) Protocols guidelines.[18]

The most recent literature research was conducted in January of 2024, utilizing the databases of Medline/PubMed, Scopus, and Google Scholar. We searched for the following terms: "Astrocytoma," "Pilocytic Astrocytoma," "CPA," or "Cerebellopontine angle," and "Brain tumor." Two independent authors, namely, P. B. and G. P., independently assessed the abstracts for their eligibility. Any disagreements were resolved through consensus with a third senior author, namely, G.D. and B.L.P. The publication date was without any restrictions.

There were 64 items identified, 24 of which were duplicates. Eighteen articles have been excluded as they pertain to pediatric patients (age ≤18), and one item was not found. We excluded 19 reports for the following reasons: Secondary to neurolymphomatosis (n = 4); secondary to primary central nervous system (CNS) lymphoma (n = 3); secondary to malignant lymphoma in the pelvis (n = 1); secondary to bone lymphoma (n = 1); associated with cutaneous telangiectasia (n = 2); primary spinal epidural non-Hodgkin lymphoma (n = 3); secondary to bone lymphoma (n = 1); related intravascular lymphomatous (n = 1); multifocal ependymoma infiltrated by lymphoma (n = 1); postmortem examination (n = 1); papers in languages other than English (n = 1); and review of literature (n = 2). Nevertheless, we excluded studies published in languages other than English, pediatric cases (Age ≤18 years), review studies, and meta-analyses. A systematic abstract screening of the references (forward search) was performed to identify additional records. Results obtained were summarized using the PRISMA 2020 flow diagram for new systematic reviews, which included searches of databases and registers only [Figure 1]. The PRISMA 2020 flow diagram templates are distributed under the terms of the Creative Commons Attribution (CC BY 4.0) license, which allows others to distribute, remix, adapt, and build on this work for commercial use, provided that the original work is properly cited. To see a copy of this license, please visit https://creativecommons.org/licenses/by/4.0/.

Our findings reported in Table 1 suggest that 75% of PA in CPA adults involved male patients (3/4) and that no prevalence of side effects was reported (50% left). Two studies reported that the internal acoustic canal (IAC) was implicated, and the clinical onset lacked any specific symptoms (100%). Two authors have described an expansion of the IAC, which has been interpreted as a potential radiological indication of schwannoma.[4,8]

The surgery procedure was performed with an intraoperative microscope, neuronavigation, and electrophysiological neuromonitoring (Somatosensory evoked potentials (SEP), motor evoked potential (MEP), electroencephalogram (EEG) free run, facial, and acoustic monitoring). The histological and molecular examinations were performed, and sections of hematoxylin-eosin stained sections and immunohistochemistry sections were obtained from the paraffin-embedded and formalin-fixed specimen. It was conducted according to the Declaration of Helsinki and approved by the Institutional Review Board of the Department of Neurosurgery at Fabrizio Spaziani Hospital, Frosinone, Italy. For scientific purposes, written informed consent was obtained from the patient.

RESULTS

The literature search resulted in a total of 64 results. After our systematic review reported in Figure 1, we included three studies in our literature reviews. In addition, we present another case of a 67-year-old male with a PA demonstrating radiological characteristics similar to an epidermoid cyst.

A Caucasian man, aged 67, presented with gait disorders, including ataxic gait, episodes of disorientation, positive Romberg sign, and short-term memory deficits, for approximately 12 months. On magnetic resonance imaging

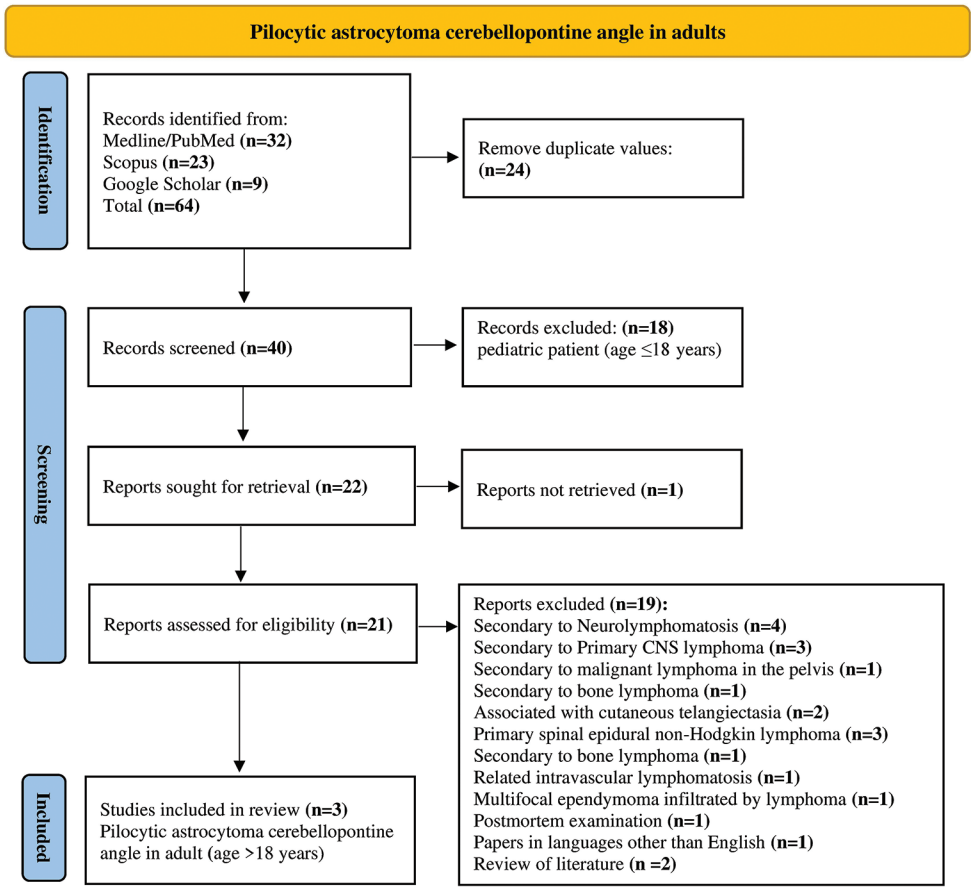

Figure 1: Flow diagram of systematic reviews of pilocytic astrocytoma in adults of the cerebellopontine angle.

Table 1: Studies that reported that PA was located in the cerebellopontine angle in adults.

Authors	Age (y)*	Sex (M/F)*	Side	Radiological aspect	IAC* involvement	Symptoms
Beutler *et al.*[4]	58	M	Left	Schwannoma	Yes, with the enlargement of IAC*	Light-headedness and ataxia
Dutta *et al.*[8]	55	M	Left	Schwannoma	Yes	Diminution of hearing in his left ear with mild headache
Bhradwaj *et al.*[5]	23	F	Right	Cystic schwannoma or arachnoid cyst	NR*	Right hearing loss
Present case	67	M	Right	Epidermoid	No	Gait disorders with ataxic

*y: Years, M: Male, F: Female, IAC: Internal acoustic canal, NR: Not reported, PA: Pilocytic astrocytoma

(MRI) of the right cerebellar pons, we observed a tumor fusiform morphology with a maximum dimension of 4.5 × 1.7 cm. The lesion was characterized by hyperintensity both on T2-weighted and fluid-attenuated inversion recovery, accompanied by edema of the adjacent cerebellar tissue. The gadolinium-enhanced T1-weighted sequence revealed an irregular distribution and a predominant concentration at the periphery. Based on the radiological features depicted in Figure 2, it is assumed that this formation is an epidermoid cyst.

In the present case, MRI revealed that there was a mass in the CPA with characteristics that suggest an epidermoid cyst. The lesion was hypointense on T1-weighted images and hyperintense on T2-weighted images, with no significant enhancement after gadolinium administration. Diffusion-

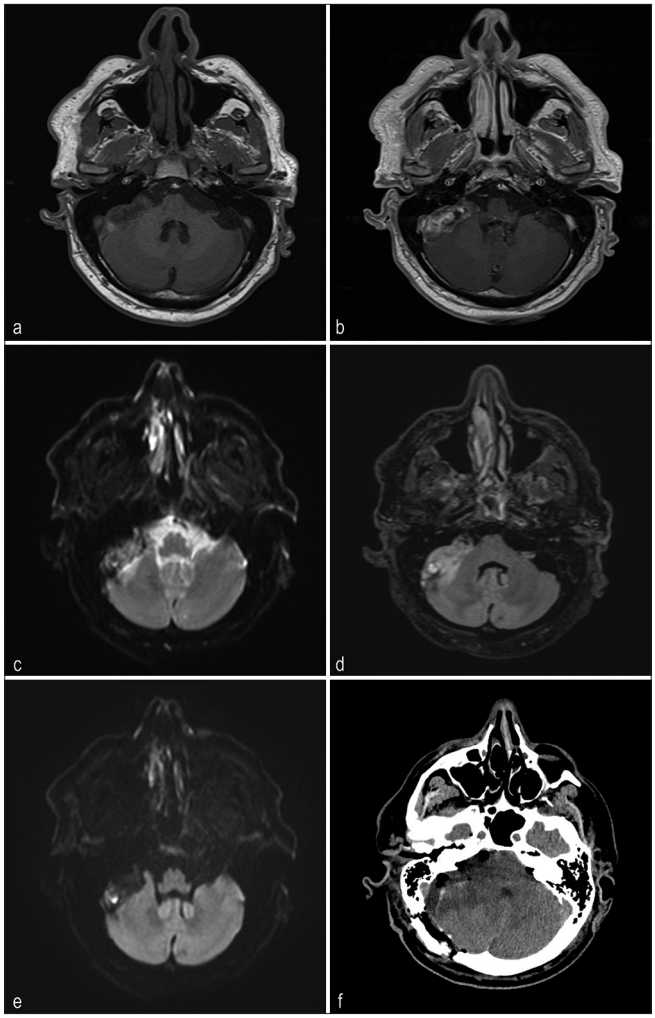

Figure 2: Magnetic resonance imaging (MRI) of pilocytic astrocytoma of cerebellopontine angle right in a 67-year-old man. (a) The T1-weighted axial MRI demonstrates a lesion of 4.5 × 1.7 cm maximum diameter, exhibiting a hypointense signal. (b) Gadolinium-enhanced T1-weighted axial MRI shows an extra-axial lesion with outfit and not homogeneous signal enhancement. This is mainly noticeable at the periphery of the lesion. (c) In the T2-weighted axial MRI, the lesion exhibits signal hyperintensities and appears to have an interface with nerve tissue. (d) In the T2 – fluid-attenuated inversion recovery-weighted axial MRI, the lesion shows signal hypertension and edema in the cerebellar parenchyma. (e) Diffusion-weighted imaging indicates a restricted diffusion of the lesion. (f) A postoperatively computed tomography scan revealed the presence of air bubbles at the lesion site and moderate edema of the cerebellar parenchyma.

weighted imaging showed a weak restricted diffusion pattern of an epidermoid cyst, as reported in Table 2.

The patient underwent a retrosigmoid craniotomy approach to remove the tumor. An intraoperative neurophysiological evaluation was performed. On examination of the microscope images, it was observed that the tumor originated from the lower cranial nerve (IX–XI) and that a distinct cleavage plane existed between the cerebellum and brain stem. The tumor, which had an irregular margin and was brown-red, occupied the right lateral cerebellomedullary cistern and developed into the cerebellopontine cistern. It had an irregular margin and was brown-red. The condition caused a dislocation of the cochleovestibular facial bundle and involved the cranial nerve VII. Histology was critical in determining whether this tumor was really a primary CPA glial tumor. Histological examination revealed features of PA such as a biphasic growth pattern, an alternation between compact areas with microcystic areas, low proliferation activity, and p53 negative. As shown in Figure 3, the neoplasm has infiltration features and numerous blood vessels with hyaline walls.

Table 2: Radiological features for the differential diagnosis of PA and epidermoid cyst.

MRI sequence	Epidermoid cyst	PA
T1WI	Hyperintense (75%)	Hyperintense
T2W2	Isointense (65%)	Isointense
PD/Intermediate	Hyperintense (95%)	Hyperintense
FLAIR	Hyperintense	Hyperintense
DWI	Restricted diffusion	Restricted diffusion
T1 C+	Usually margin of the cyst may show minimal enhancement (35%)	Nodular enhancement in the center and thin rim at the margins
T1WI	Hyperintense (75%)	Hyperintense
T2W2	Isointense (65%)	Isointense
PD/Intermediate	Hyperintense (95%)	Hyperintense
FLAIR	Hyperintense	Hyperintense
Diffusion	Restricted	Restricted

PA: Pilocytic astrocytoma, MRI: Magnetic resonance imaging, T1WI: T1-weighted image, FLAIR: Fluid-attenuated inversion recovery, DWI: Diffusion-weighted imaging, PD: Proton density weighted, T1 C+: T1-weighted contrast-enhanced

Notwithstanding, the GBM and PA exhibit significant distinctions at the molecular level, which are the root cause of their distinct biological behaviors and clinical outcomes. In our case, the differential diagnosis with GBM proved to be challenging due to the overlapping molecular characteristics, as illustrated in Table 3.

During the immediate postoperative period, the patient did not experience new-onset neurological deficits, except for a Grade II deficit of the VII ipsilateral cranial nerve second House Brackmann Facial Nerve Grading System. However, he exhibited psychomotor agitation and disorientation in both space and time. This has been improving gradually. During the 3rd day after the surgery, the patient experienced cardiac atrial flutter, accompanied by a high ventricular response, resulting in an irreversible coma. The patient underwent treatment for hydrocephalus through the use of an external ventricular shunt. Heart failure and cardiac

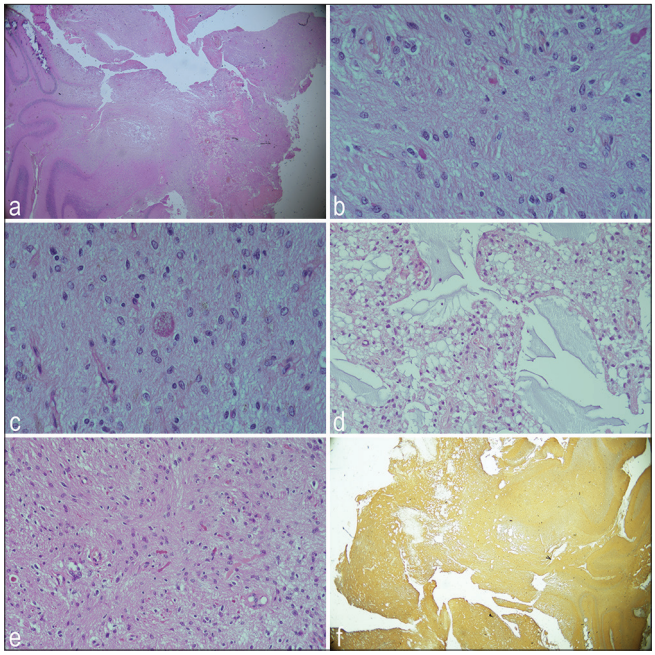

Figure 3: Histological examination of pilocytic astrocytoma of cerebellopontine angle right in a 67-year-old man. (a) Haematoxilin-Eosin (H&E) ×10 biphasic growth pattern characterized by compact areas alternating with microcystic areas, (b) H&E ×20 globuli ialini, (c) H&E ×40 granular body, (d) H&E ×20 microcystic areas, (e) H&E Rosenthal fibers, and (f) positive expression of the glial fibrillary acidic protein.

Table 3: Molecular features for differential diagnosis between PA and GBM in adults.

Molecular markers	PA	GBM
ATRX	+	+
GFAP	+	+
OLIG-2	+	+
BRAF (KIAA1549-BRAF fusion)	+	-
p53	-	+
IDH1 (R132H/H09)	-	-
H3K27m	+	+
Mitosis	Low activity	Abundant
Microvascular changes	Absent	Focal
Necrosis	Absent	Focal
Cellular malignant neoplasm	Low	Highly
	+	-

PA: Pilocytic astrocytoma, GBM: Glioblastoma, IDH1: Isocitrate dehydrogenase 1, GFAP: Glial fibrillary acidic protein, H3K27m: Lys-27-Met mutations in histone 3 genes, OLIG-2: Oligodendrocyte transcription factor 2, KIAA1549-BRAF: K-B fusion Gene, ATRX: ATP-dependent helicase

arrhythmia can cause an embolic ischemic stroke of the brain stem. The patient passes away within a few weeks. The patient had no previous history of cardiac arrhythmias or other known cardiac disorders, except for arterial hypertension, which was adequately compensated with ramipril therapy.

DISCUSSION

PA is a rare entity in adults, but it is one of the most common tumors in children. Moreover, the supratentorial site is the most common localization, and the pontocerebellar angle is a rare location.[4,6] Due to this, the radiological features of CPA tumors provide crucial information regarding the differential diagnosis. However, a radiological diagnosis can be challenging and sometimes wrong.[4,8] After a systematic literature review, we selected 3 papers that focused on primary PA in CPA in adult patients. The authors of this study evaluated the demographic, radiological, and intraoperative characteristics. Bhradwaj *et al.* reported a case in which the radiological manifestation of the cyst lesion resembled an arachnoid cyst or a cystic schwannoma.[5] Moreover, the mild hearing loss reported in two cases may have been misleading in order to reach a correct diagnosis.

As this condition is extremely rare, there is limited data on the follow-up and outcomes of these patients. Bhradwaj *et al.* reported a 3-month follow-up showing an improvement in preoperative symptomatic.[5] Beutler *et al.* reported a worsening of VII cranial nerve paresis without any follow-up information.[4] Finally, Dutta *et al.* did not provide any information on outcomes.[8]

All the papers included in this systematic review were considered suspicious for radiological evidence of schwannoma lesions in CPA. We have reported the first case of PA of CPA with radiological mimicking of an epidermoid cyst.

These authors have further reported that tumors originate from the VIII cranial nerve.[4,8] Regardless, the radiological and intraoperative characteristics of our case have prompted us to suspect the epidermoid cyst of the lower cranial nerve.

Additionally, during the intraoperative examination, it was observed that a CPA tumor had a cleavage plane between the brain stem and cerebellum. These features were essential in determining that this tumor was a primary CPA glial tumor, and they emphasize the extremely rare conditions we analyze in this paper.[2,5,10,22] Numerous authors offer various theories regarding the origin of primary PA of CPA. However, in recent times, it has become more widely accepted that they originate from the medial velum of the lateral recess of the fourth ventricle or that they may arise as primary leptomeningeal gliomas, possibly in leptomeningeal neuroglial heterotopias.[4,17,19]

Posterior cranial fossa GBM is rare in adults, accounting for <1% of all GBM cases.[12,21] The reasons for its rarity are unknown. Numerous reported studies have only included a limited number of patients from a single institution, and the clinical data regarding primary posterior cranial fossa GBM is inadequate.[1,11,12] The cerebellum is the most common site

in the posterior cranial fossa, whereas GBM is extremely rare in the CPA. The cerebellar GBM has a poor prognosis compared to supratentorial GBM, and the median overall survival is approximately 9 months, which is worse than that of supratentorial GBM patients.[11,12]

In our literature review, we found that four papers reported the GBM of CPA. A gliosarcoma was identified within one of the tumors. To the best of our knowledge, there have been only four reported cases that have been localized within the CPA. This case presents a challenge due to the involvement of the IAC and the radiological aspect of other tumors of the CPA, which strongly suggests vestibular schwannoma.[22,23,25,26] Despite the lack of definitive mechanistic data, as per the current literature, we hypothesize that there exist two potential theories regarding the origin of GBM's tumor cells. In all CNS components, heterotopic glioneuronal cell nests have been described and are considered to be the source of primary gliomatosis. Moreover, glioneuronal heterotopias have also been reported as uncommon lesions in the CPA.[21,26] It is possible that microscopic or macroscopic glioneuronal heterotopias are the source of a GBM that is otherwise disconnected from all CNS tissue. Histological analyses have shown that the most proximal CN segments adjacent to the brainstem root entry zones are covered with glial cells, including astrocytes and oligodendrocytes. These cells myelinate the nerve segment just proximal to the first myelinating periphery.[21,22]

GBM is characterized by a complex array of genetic alterations that contribute to its aggressive nature.[15,16] These include frequent epidermal growth factor receptor (EGFR) amplifications and mutations, particularly the EGFR variant III variant, which drives uncontrolled cell proliferation and survival through pathways such as phosphatidylinositol 3-kinase (PI3K)/protein kinase B (AKT) and RAS/RAF/MEK/ERK.[3,13,16,20] In addition, PTEN mutations lead to the activation of the PI3K/AKT pathway, promoting cell survival and proliferation. Mutations in the TP53 gene, common in secondary GBM, result in impaired DNA damage response and increased cell survival, while telomerase reverse transcriptase promoter mutations enable limitless replicative potential by upregulating telomerase.[7] Chromosomal aberrations, including gains of chromosome 7 and losses of chromosome 10, further contribute to the genomic instability characteristic of GBM.[15,24]

In contrast, PA typically exhibits a simpler genetic landscape, primarily involving alterations in the BRAF gene. The most common alteration is the KIAA1549-BRAF fusion, which results from a duplication event on chromosome 7 and leads to constitutive activation of the BRAF kinase.[9] This activation drives the MAPK/ERK signaling pathway, promoting cell proliferation and survival. Less commonly, the BRAF V600E mutation also activates this pathway. In cases associated with neurofibromatosis type 1 (NF1), mutations in the NF1 gene result in increased RAS signaling, further activating the MAPK/ERK pathway.[13]

In contrast to GBM, PA typically does not involve modifications in the p53 or RB pathways, indicating its more benign nature. PA has a less aggressive molecular profile and a slower growth rate than GBM, which is highly malignant and invasive. These molecular distinctions are critical in determining and shaping the clinical management and prognostic outcomes for patients with these tumors.

In our case, the morphological and immunohistochemical features of the lesion are characterized by a positive ATP-dependent helicase (ATRX), a negative Isocitrate dehydrogenase (IDH)1/IDH2, and restricted mitotic and proliferative activity. From a histological point of view, the differential diagnosis was made with GBM[15] because both could show IDH wild-type.[2,14,19,22] The PA of the CPA has the mutation of ATRX, mutation of Lys-27-Met mutations in histone 3 genes, and non-mutated IDH 1, which are also present in the current classification of the GBM.[15] Notwithstanding the patient's age, the histological diagnosis of PAs was substantiated by the absence of microvascular changes, necrosis, and mutations in the tumor suppressor gene p53. In addition, the low index of mitosis and the presence of Rosenthal fiber were well confirmed.

The role of radiological imaging in the diagnosis and management of CPA tumors is crucial. MRI is the preferred imaging technique, providing comprehensive details on the tumor's location, size, and association with adjacent structures.

To summarize, the diagnosis of PA of the CPA is exceedingly uncommon among adults. To accurately diagnose this condition, a comprehensive approach is required, including a thorough radiological assessment, intraoperative findings, and a detailed histopathological examination. The unique molecular profile of PA, which is different from GBM, is an essential guide for differential diagnosis and clinical management. This case emphasizes the importance of considering PA in the differential diagnosis of CPA tumors, even in older adults, and emphasizes the necessity of meticulous evaluation to avoid misdiagnosis and ensure suitable treatment.

Limitations

In this study, we present a systematic review of the English literature regarding three cases of PA of CPA and a new case we found in our clinical practice. Few cases possess statistical significance and can only provide indicative and not definitive information. Furthermore, the reported cases are not presented uniformly. Given the limited number of cases, only observational and retrospective studies are feasible.

CONCLUSION

This systematic review demonstrates that PA of the CPA is an uncommon occurrence among adult patients. It is important to consider this factor when considering the differential diagnosis of vestibular schwannoma, epidermoidal cyst, and GBM. We have identified the initial instance of a PA at the CPA, which resembles an epidermoid cyst, in a 67-year-old male. It is possible to distinguish between PA and GBM in the CPA by examining the tumor's morphological characteristics, including the presence of cellular malignancy, mitotic activity, microvascular changes, and necrosis. Our findings highlight the importance of a thorough radiological and histopathological evaluation in preventing misdiagnosis and ensuring appropriate treatment for these uncommon tumors.

Author's contributions

P.B., G.P.: Conceptualization. G.D.A., B.L.P.: methodology; P.B., G.P.: Software; G.D.A., B.L.P.: Validation; P.B., P.L.: Formal analysis; C.Q., P.A.: Investigation; P.L., P.F., V.M.: Resources; P.B., G.P.: Data curation; P.B., G.P., P.L.: Writing – original draft preparation; G.D.A., B.L.P., and P.F.: Writing review and editing; V.M.: Visualization; G.D.A., P.F., and P.B.: Supervision; G.D.A., B.L.P. and P.F.: Project administration. All authors have read and agreed to the published version of the manuscript.

Acknowledgments

Prof. MD Giovanni Condemi.

Ethical approval

The study was conducted in accordance with the Declaration of Helsinki and approved by the Institutional Review Board of Ospedale F. Spaziani (I2022-012816).

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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How to cite this article: Bruzzaniti P, Pennisi G, Lapolla P, Familiari P, Maiola V, Quintiliani C, *et al.* Cerebellopontine angle pilocytic astrocytoma in adults: A systematic review. Surg Neurol Int. 2024;15:363. doi: 10.25259/SNI_539_2024