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Case Report

The Extra-axial Cerebellopontine Angle Medulloblastoma in an Adult Patient: A Case Report and Review of Literature

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ABSTRACT

Background and Importance: Medulloblastoma (MB) is regarded as a scarce primary brain neoplasm in adulthood, originating from the fourth ventricle or the cerebellum, and cerebellopontine angle (CPA)-MB has been described less often in the literature. Few cases of CPA MB have been reported in the English-written literature, most of which are intra-axial, mainly in children; adults' extra-axial CPAMB is even scarcer. To the best of the authors' knowledge, 12 cases of extra-axial CPA MBs have been reported in the English-written literature.

Case Presentation: A 23-year-old man presented to our center complaining of a generalized pulsatile headache, imbalance, swallowing impairment, and right-sided hearing loss for the past 20 days. Computed tomography (CT) scan of the brain showed a hyper-dense extra-axial mass lesion (41×37 mm) in the right CPA with a significant compression effect on the fourth ventricle, causing a three ventricular obstructive hydrocephalus. Magnetic resonance imaging (MRI) showed a well-defined heterogeneous extra-axial, lobulated, dural-based mass lesion in the right CPA, hypointense on the T1 sequence, and hyperintense on the T2 sequence compared with the adjacent parenchyma, which has a bright heterogeneous enhancement during gadolinium injection. A significant mass effect was observed on the adjacent parenchyma, brain stem, and fourth ventricle without evidence of parenchymal edema. The patient underwent emergent surgery the next day regarding the mass effect and hydrocephalus. On the postoperative examination in the intensive care unit, no new neurological deficit was detected, and the swallowing and gag reflex significantly improved.

Conclusion: Though rare, clinical considerations, along with early supportive radiologic measures, should be considered in subjects with suspected MB. A total tumor excision approach followed by aggregative chemotherapy/radiotherapy is designed to hinder tumor relapse.

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Highlights

• Cerebellopontine angle (CPA) medulloblastoma (MB) is a rare entity in the adult population.

• The maximal extent of surgical resection combined with chemotherapy/radiotherapy provides the best option for CPA MB.

• The total survival chance of CPAMBs remains similar to other regions, with a five-year survival rate of 25%-70% for MBs.

Plain Language Summary

Medulloblastoma (MB) is regarded as a scarce primary brain neoplasm in adulthood, originating from the fourth ventricle or the cerebellum, and Cerebellopontine Angle (CPA) MB has been described less often in the literature. A 23-year-old man presented to our center complaining of a generalized pulsatile headache, imbalance, swallowing impairment, and right-sided hearing loss for the past 20 days. Physical examination confirmed a bilateral gag reflex impairment, right-sided hemifacial paresis, and right-sided sensory neural hearing loss. The remainder of the physical examination was uneventful. Radiologic evidence was in favor of Cerebellopontine Angle (CPA)-MB, so the patient underwent emergent surgery on the second postoperative day regarding the mass effect and hydrocephalus. On the postoperative examination in the intensive care unit, no new neurological deficit was achieved, and swallowing dramatically improved as well as the gag reflex. A total tumor excision approach followed by aggregative chemotherapy/radiotherapy allows for the best choice for the sufferer.

Background and Importance

Medulloblastoma (MB) is regarded as an aggressive embryonal primitive neuroectodermal neoplasm of the cerebellum of the World Health Organization (WHO) grade IV, arising from germinal matrix cells of the primitive neural tube. The exact reason behind this malignancy is not well identified, but numerous familial cancer syndromes and John Cunningham polyomavirus infections can play a crucial role in the tumor's physiopathology [1-5]. MB frequently occupies the midline of the cerebellum, and cerebellar hemispheres are known as the conventional origin for MBs in adults. MBs can be observed either in the midline of the cerebellum or intra-cerebellar hemispheric or even in the Cerebellopontine Angle (CPA) area, and approximately 80% of childhood MBs tend to arise at the vermis and the apex of the fourth ventricle [2, 6, 7]. Among the molecular subtypes of MBs, group 3 and group 4 mainly involve the fourth ventricle, notwithstanding sonic hedgehog (SHH) MB primarily occupying the cerebellar hemispheric [6].

More importantly, Gross Total Resection (GTR) is preferred for CPA MBs, as it is associated with a significant risk of degenerative impacts on cranial nerve seven (CN-VII) and adjacent components, while the maximal extent of surgical resection is regarded as a standard approach for managing this malignancy [6]. Herein, we sought to outline the general features of CPA MB, its diagnostic dilemma, significant correlated prognostic factors, and multidisciplinary procedure for its management.

Case Presentation

A 23-year-old man presented to our center complaining of a generalized pulsatile headache, imbalance, swallowing impairment, and right-sided hearing loss for the past 20 days. Physical examination showed a bilateral gag reflex impairment, right-sided hemifacial paresis, and right-sided sensory neural hearing loss. The rest of the physical examination was normal.

The patient initially underwent a computed tomography CT scan of the brain to investigate intracranial lesions. The CT scan of the brain showed a hyper-dense extra-axial mass lesion (41×37 mm) in the right CPA with a significant compression effect on the fourth ventricle, causing three-ventricular obstructive hydrocephalus (Figure 1).

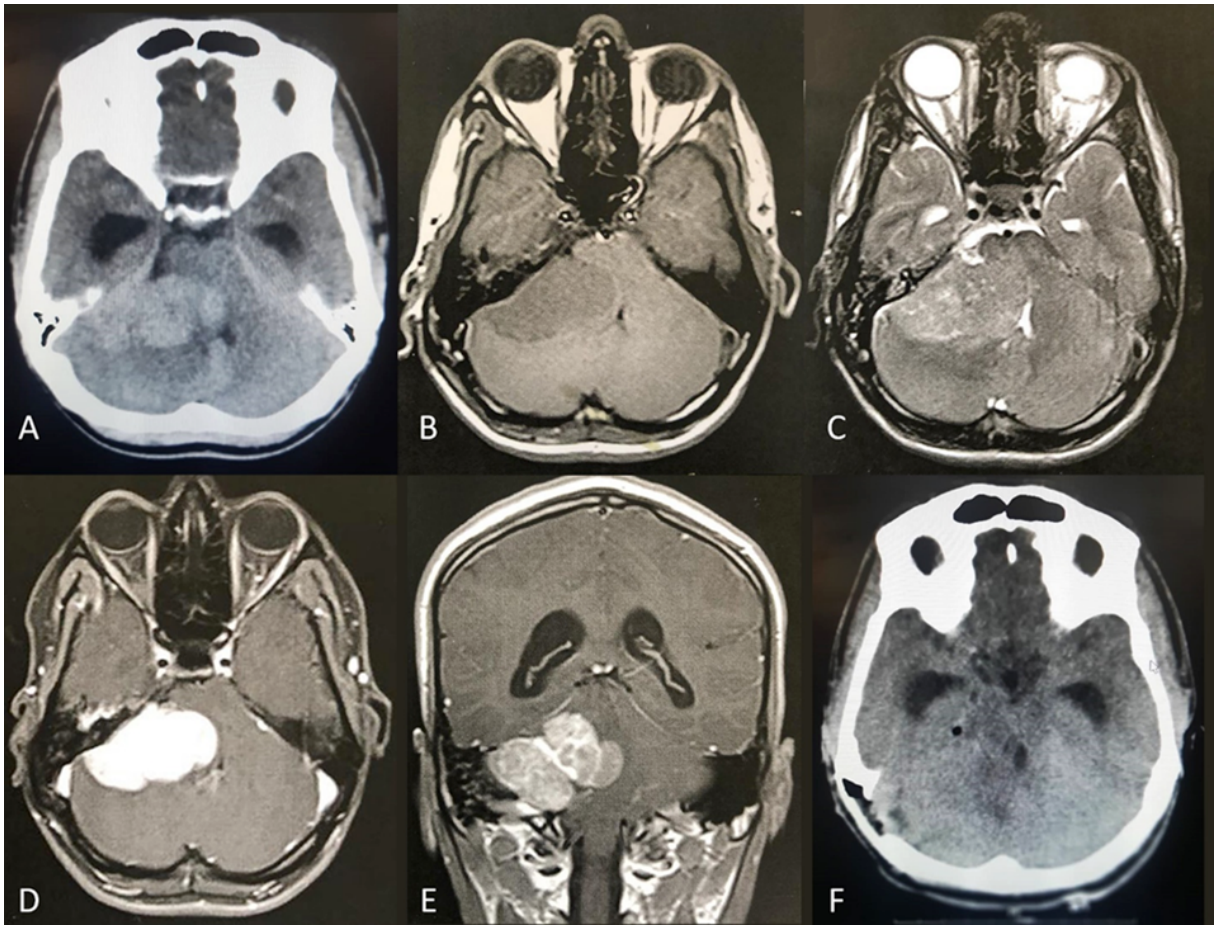

Figure 1. Preoperative Computed Tomography (CT) Scan

A: Magnetic Resonance Imaging (MRI); B, C, D, E: Demonstrating an extra-axial, lobulated, dural-based mass lesion (41*37mm) in the Right Cerebellopontine Angle (CPA), B: Hypointense on T1 and C: Hyperintense on T2; D: With bright heterogeneous enhancement using gadolinium on axial; and E: Coronal View; F: A postoperative CT scan revealing a complete resection of the tumor and a reduction in the mass effect. The mass effect on the brain stem and fourth ventricle causes hydrocephalus.

Magnetic Resonance Imaging (MRI) depicted a well-defined heterogeneous extra-axial, lobulated, dural-based mass lesion in the right CPA, hypointense on the T1 sequence, and hyperintense on the T2 sequence compared with adjacent parenchyma, which has a bright heterogeneous enhancement during gadolinium injection. A significant mass effect was observed on the adjacent parenchyma, brain stem, and fourth ventricle without evidence of parenchymal edema (Figures 2, 3, 4, 5, 6 & 7).

The patient underwent emergent surgery the next day due to the mass effect and hydrocephalus. In a lateral position, his head was fixed using a Mayfield fixator. Concerning the hydrocephalus, we initially placed a posterior external ventricular drainage to control and decrease intracranial pressure during surgery. Under the Electroencephalography (EEG) monitoring of cranial nerves, we approached the patient via the retrosigmoid sub-occipital lateral craniectomy toward the CPA. We drained Cerebrospinal Fluid (CSF) from cisterna magna, then gently dissected the CPA adhesion bands where the tumor presented. At first look, the lesion was creamy and soft. We initially obtained tumor tissue fragments for the frozen section, which revealed large anaplastic cells in favor of MB. The tumor adhered to the VII-VIII cranial nerve complex. It was completely resected with the preservation of the VIII cranial nerve. VII cranial nerve was repaired using a sural nerve graft. All other cranial nerves were intact on direct vision and EEG neuromonitoring.

In the postoperative examination in the intensive care unit, no new neurological deficit was detected, and swallowing significantly improved as well as gag reflex. Histopathologic evaluation revealed hypercellular neoplasm with round cell morphology, frequent mitosis,

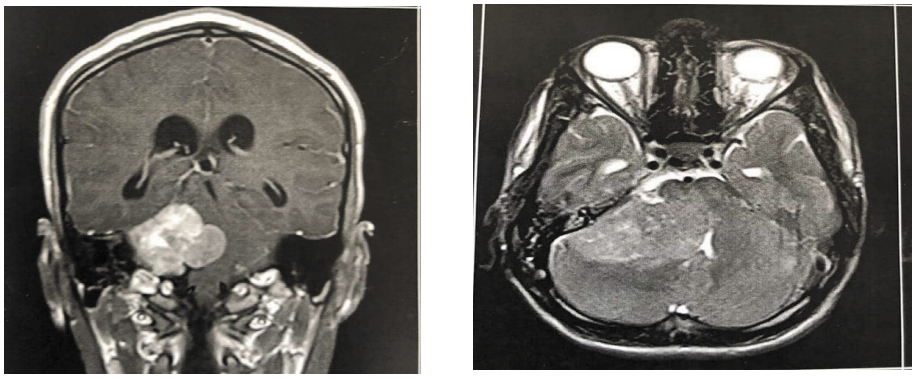

Figure 2. T1 with gadolinium-coronal view, T2-axial view

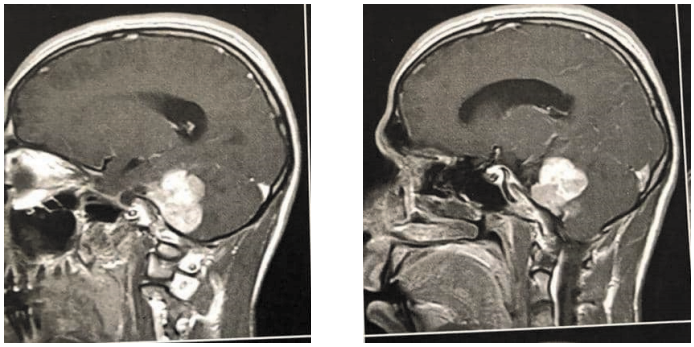

Figure 3. T1 with gadolinium-sagittal view

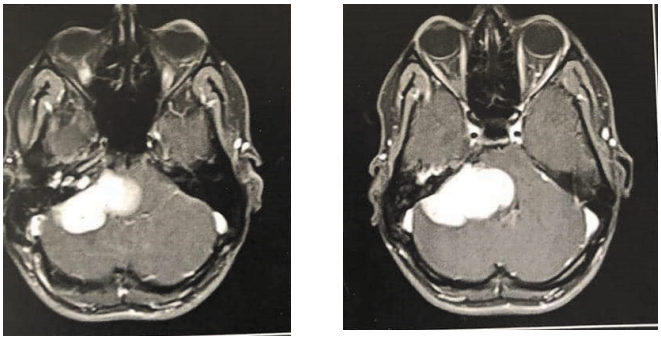

Figure 4. T1 with gadolinium

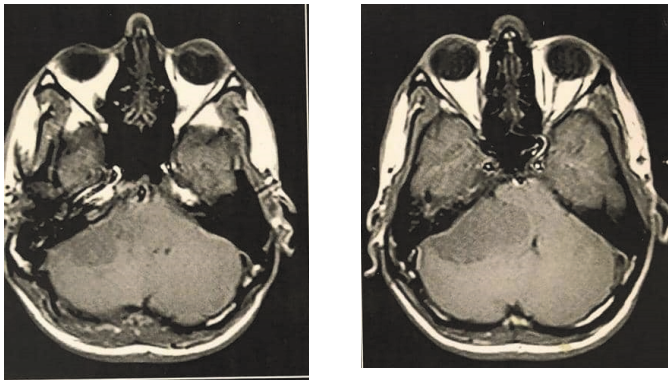

Figure 5. T1 without gadolinium-axial view

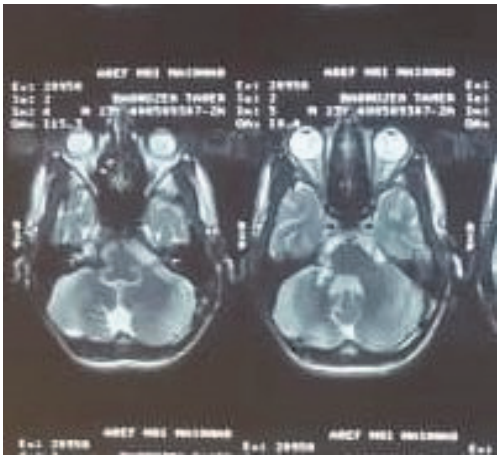

Figure 6. A postoperative axial view T2-weighted brain MRI of the patient, which demonstrates a total removal of the lesion

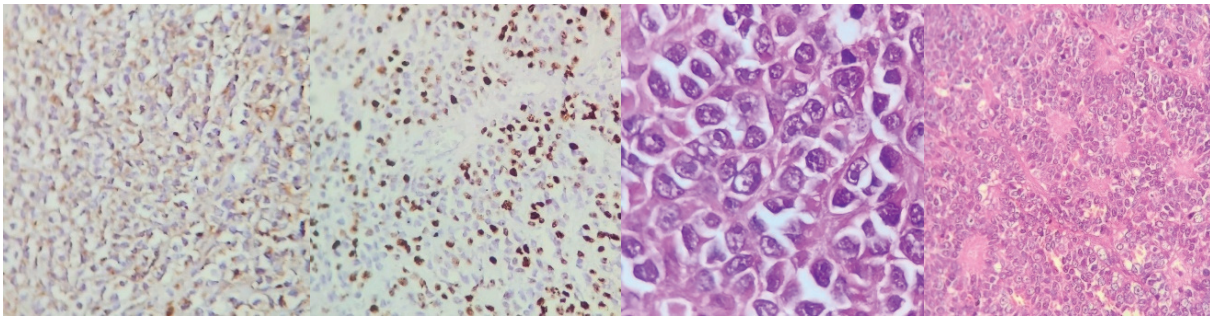

Figure 7. Synaptophysin is positive, Ki67 is highly increased, large cells revealed vehicular nuclei and prominent nucleoli, hematoxylin and eosin (H and E) stain,×400, hypercellular neoplasm with round cell morphology and homer wright rosettes, hematoxylin and eosin (H and E) stain,×100.

and areas of necrosis. Areas of large cell transformation with vehicular nuclei and prominent nucleoli are evident. Immunohistochemical (IHC) stain is diffusely positive for synaptophysin, focally positive for Glial Fibrillary Acidic Protein (GFAP) and Epithelial Membrane Antigen (EMA), and negative for cytokeratin (CK), Leukocyte Common Antigen (LCA), and Olig2. KI67 showed a high proliferative index. The final diagnosis was MB, a large cell variant, WHO grade IV (Figure 7). In the histologic and immunohistochemistry evaluation of the tumoral tissue, GFAP was positive, EMA was focally positive, synaptophysin was patchy positive, and Ki-67 was 60% positive in large anaplastic cells, while Olig-2 was reported negative. The above markers and histologic re-evaluation confirmed the diagnosis of MB grade VI. The patient underwent a radiochemotherapy regimen and was discharged in good clinical condition on the sixth postoperative day.

In the histologic and immunohistochemistry evaluation of the tumoral tissue, GFAP was positive, EMA was focally positive, synaptophysin was patchy positive, and Ki-67 was 60% positive in large anaplastic cells, while Olig-2 was reported negative. The above markers and histologic re-evaluation confirmed the diagnosis of MB grade IV. Table 1 provides more details regarding the previously reported CPA-MBs in the literature.

Discussion

MB is among the posterior fossa's most common childhood malignant tumors, accounting for 16%-20% of all pediatric tumors [8]. It also represents approximately one-fourth of all neoplasms of central nervous systems and 40% of posterior fossa tumors and is known as the second most prevalent inter-cranial tumor after pilocytic astrocytoma [6, 9]. The CPA MB is more common in adults, with a male sex ratio (1, 3:1) [6, 10, 11]. Six to eight-year-old juveniles consist of most case reports of this malignancy [6, 11, 12]. MB predominantly occurs as an intra-axial phenomenon arising from the subependymal matrix or external granular layer of the inferior medullary vellum of the vermis with an intrinsic proclivity to leptomeningeal diffusion [13-15]. The most common variant in the adult group of MB is SHH. It accounts for approximately 60% of all patients in adulthood [16].

Table 1. The literature review of the extra-axial cerebellopontine angle (CPA) medulloblastomas (MBs)

Author	Tumor Characteristics/Variant (According to WHO)	Age/Gender	History	Sign	Approach/ Outcome	Citation
Eshagh Bahrami et al. (2013)	Extra-axial MB in the CPA Pale islands comprised of micronodular/reticulin-free zones Desmoplastic/ nodular	23-year-old man	Nausea, vomiting and ataxia, and deafness of his right ear in the past 2 months	Impaired hearing in audiometry Hypointense on T1 weighted images and hyperintense on T2 weighted images	Right retromastoid craniotomy and gross total excision No neurological deficit after surgery No metastasis after radiotherapy	[15]
Spina et al. (2013)	Classic extra-axial MB in the CPA Case A: Classic type with nodular aspects Case B: Classic type of MB infiltrating the arachnoid	Case A: 22-year-old man Case B: 26- year-old woman	Case A: Headache, ataxia, hearing loss in her left ear, dizziness, and tinnitus as of the last 3 months Weakness of the right arm, slight Left nystagmus and a mild peripheral deficiency of the left VII-CN nerve. Case B: Chronic headache	Case A: Hyperintense T2-weighted images, hypointense T1-weighted images Case B: Hypointense in T1-weighted images and isointense in T2-weighted images Case B: Edema in the anterior superior right cerebellar hemisphere	Case A: retrosigmoid craniectomy and gross total tumor resection and radiotherapy. Case A: postoperative personality changes and neurobehavioural abnormalities. Case B: right retrosigmoid craniectomy with gross-total resection. Case B: Radiation therapy Case B: No evidence of postoperative metastasis or recurrence was observed.	[10]
Singh et al. (2011)	Extra-axial MB in the CPA Rosettes of small round cells The high nucleus-cytoplasm ratio increased mitotic figures(classical MB, grade IV)	22-year old man	Headache, ataxia, vomiting and left VII-CN failure as of last month Lack of superficial reflexes Brisk deep tendon reflexes and bilateral extensor plantar reflexes 3/5 motor power in his upper limbs, 2/5 in his lower limbs Sensory impairment below D4 dermatome A background of obstructive hydrocephalus/right ventriculoperitoneal shunt Papilloedema Paralyzed left IX and X-CN, as well as left cerebellar signs	Lower left motor VII CN deficit, paralyzed Left X-CN and IX-CN along with papilloedema in fundus examination and left cerebellar Signs	Left retromastoid craniectomy and gross total excision declined to receive craniospinal radiotherapy; subsequent progressive quadriparesis/sensory impairment, as well as bladder and bowel impairment, was presented.	[27]
Ninh Ba Doan et al. (2018)	Extra-axial midline tentorial MB grade IV SHH type Desmoplastic	29-year-old man	Syncopal episode and headache for the last 2 weeks	Normal neurological exam MRI: Tentorial dural-tail sign	Unremarkable postoperative period Radiation and adjuvant chemotherapies (vincristine, cisplatin, and cyclophosphamide)	[2]

Author	Tumor Characteristics/Variant (According to WHO)	Age/Gender	History	Sign	Approach/ Outcome	Citation
EUI jin Chung et al. (2014)	Extra-axially MB in the cerebellar Hemisphere Desmoplastic/nodular type (grade IV) low nuclear-cytoplasmic ratio with neuronal maturation	5-year-old man	Nausea, vomiting, and gait disorder in the past 5 days.	Dysmetria, terminal ataxia, and tandem gait Upstream obstructive hydrocephalus iso-SI with a gray matter on T1-weighted and T2-weighted imaging Parenchymal edema Dilatation of the 3rd and both lateral ventricles	Emergency external ventricular drain and a left lateral suboccipital, infratentorial craniotomy, as well as gross total surgical excision, were employed. No postoperative evidence of tumor remnant Radiotherapy regimen	[19]
Ishita Pantet al. (2016)	Extra-axial MB in the cerebellopontine angle Minimally vascular grayish-white, soft, and suckable mass. Carrot shaped hyperchromatic nuclei with scanty cytoplasm, brisk mitosis, and neuroblastic rosettes Desmoplastic/ nodular	15-year-old man	Sharp-shooting headaches amid last 3 months. Gait disturbance in his last month.	Ataxia with no other neurologic deficit Hydrocephalus Heterogeneous signal intensity, as well as necrosis/ cystic degeneration Mild perifocal edema	Retromastoid suboccipital craniectomy and gross total excision Uneventful postoperative period He refused radiotherapy and lost the follow-up.	[4]
Mukesh Kumar Bhaskar et al. (2017)	Extra-Axial CPA MB hyperechoic on ultrasound, soft to firm, moderately vascular, partially suckable, grayish-brown Densely packed cells arranged in pseudorosettes/ a trabecular pattern Round to oval, hyperchromatic nuclei, as well as scanty amounts of cytoplasm in tumor cell	1-year-old woman	Vomiting, decrease in feeding and intermittent fever period in the past 30 days, asymmetry of VII-CN nerve with a 15-day history of deviation of the angle of the mouth Changed sensorium	Unconscious status along with pallor Bilateral coarse crepitations Hydrocephalus Reacting only to localized painful stimulations Paralyzed VII-CN associated with a deficiency in cough reflex lobulated the left CP angle MRI: Hypointense on T1-weighted, isointense on T2-weighted images	Left retromastoid suboccipital craniectomy and gross total excision Ventilator support and adjuvant CT/ radiotherapy were employed. Succumbed on the 20th day of the operation following a low Glasgow Coma Scale state	[20]

Classic and desmoplastic have higher incidence rates among the histological subgroups of CPA MBs [7]. Despite growing bodies of literature justifying the exact cause and origin of MB tumors, it remained controversial among investigators. In this regard, some researchers emphasize that CPA MBs can arise from proliferating remnants in the posterolateral medullary velum or residue of the external granular layer of the flocculus hemispheres. Others accentuate the assumption that is based on the upward and lateral migration of germinal cells from the roof of the fourth ventricle during the formation of the external granular layer of the cerebellum, a theory that confirms the CPA localization of MB. Albeit 80% of MBs are located in the cerebellar midline at the inferior vermis and typically project into the fourth ventricle, they can also be present in the sidewall of the cerebellum's hemisphere or disseminate exophytically into the C.P. angle zone [3, 4, 6, 9, 14, 15, 17-20]. The extra-axial manifestation of MB in other sites is regarded as a very unexpected phenomenon [4]. Extra-axial incidence

of the MBs can probably be attributed to residues of the neural crest cells, which persisted particularly from where MB arose.

The lateral recess of the fourth ventricle contains a portion of the Lower Rhombic Lip (LRL), which is strongly susceptible to Wingless Tumors (WNT) expression. Subsequent empirical investigations clarified the hypothesis that WNT MB may originate from the midline of the fourth ventricle; thereby, they potentially tend to lateralize in the CPA [5, 21].

Paul Gibson et al. compared genes expression, marking human WNT and SHH MBs in the Upper Rhombic Lip (URL) and LRL, and their investigations bring SHHMBs tumor up as a neoplasm originating from the glutamatergic granule neural precursor cells (GNPCs, cells forming the external granular layer of cerebellum)of the URL whereby they are often present in the convexity of cerebella's hemispheres and vermis; however, WNT-MBs tumors developed from cells of the dorsal brainstem demonstrating that brainstem could be regarded a potential source of WNT MBs by infiltrating of WNT to the embryonic brainstem leading to the high incidence of WNT-MBs genes expression in LRL and embryonic dorsal and lateral brainstem [21]. The inability of SHH MBs to engage the fourth ventricle floor in the CPA region is consistent with recent findings of Tao Wu et al. [6]. Several lines of literature identify SHH MBs as the only type among all MBs subgroups in which the tumors are located in the intra-hemispherical position. Hence, to the best of the authors' knowledge, the first reported case of extra-axial midline tentorial adult MB followed by a dural-tail sign by Doan NB et al. suggests an unconventional inter-hemispheric SHH MBs [2].

In contrast to midline MBs in most pediatric populations, almost half of MBs in adult groups are located laterally in the cerebellar hemispheres, reflecting miscellaneous etiology and pathogenesis pathways [1]. In this respect, desmoplastic MBs are much more common in the adult population, with approximately 71% of them accompanied by the lateral cerebellar hemisphere manifestation, contrary to 12.5% occurrences of the mostly midline-restricted characteristics of the classical MBs variant [2]. The non-specific triad of vomiting, lethargy, and headache is observed in most pediatric patients with MB [14].

Rapid progressive intracranial hypertension-related symptoms (headache, nausea, vomiting, and papilledema), cerebellar ataxia and diplopia, and nystagmus due to cranial nerve involvement are the most common clinical presentations of MBs. Other manifestations, such as gait problems, backache, and bowel and bladder failures, can include spinal metastatic MBs [8, 10, 11, 22].

Generally, trigeminal, abducens, facial, and vestibulecochlear impairments, lower cranial nerve problems, and symptoms of cerebellar malfunction often highlight CPA neoplasms. Insofar as an initial and aggressive behavior of cerebellar tumor symptoms and gait disorders, including progressing gait ataxia propose the probable presence of the CPA MB lesions, early impairment of facial and vestibule-cochlear nerve, and positional nystagmus reflect an early sign of a probable existing vestibular-acoustic schwannoma, respectively [15]. Despite the intrinsic microscopic appearance of pink or gray MBs observed either in calcified or hemorrhaged regions, histopathological characteristics of the tumor in the CPA region and lateral cerebellar cortex include heterogonous isomorphic sheets of densely round to oval hyperchromatic cells surrounded by scanty cytoplasm along with the dramatically nucleocytoplasmic ratio [4, 8]. Regretfully, neither a reliable pathognomonic nor radiographic approach is recommended as an assistant for different MBs from other subjects with a high probability of developing in the CPA, including vestibular schwannoma and meningioma, which collectively account for 85%-90% of all CPA tumors in adults followed by the meningioma, primary cholesteatomas, and epidermoid tumors [6, 9, 22, 23]. Notably, CT characteristics of CPA MBs include prominent hyperdense homogeneous mass in most assessments and in 10% of cases, it may show calcification [10]. Despite MBs, often presenting as poor-demarcated soft friable mass and central compacted lesions, other tumors of postcranial fossa, including epidermoid cysts, are attenuated on non-enhanced CT due to sufficient cytoplasm and less dense matrix, and they demonstrate well-demarcated lesions concerning their high lipid content, such as cholesterol [8, 14, 24]. No exclusive radiological features exist that favor a definitive diagnosis of CPA MBs malignancies.

MBs arising either from CPA or intraventricular region characterized by iso-hypointense to gray matter on T1-Weighted Images (T1W1) with a predominantly heterogeneous contrast enhancement paradigm after the addition of gadolinium, most likely due to excessive tumors cellularity, inadequate blood supplies, or necrosis, howbeit their appearance ranges from hypointensity to hyperintensity to gray matter on T2-weighted (T2W2) sequences due to the incremented nucleus to cytoplasm ratio of neoplasms cells or diminished free water [9]. Although most MRI assessments reveal heterogeneous Gd-enhancement agents, MBs may vary

from uniform to patchy enhancements, and 10%-15% of cases are not allocated to heterogeneous aspects, which canaled to misdiagnosis [5, 9, 20]. Normal integrity and symmetry of the internal auditory canals rather than other CPA lesions, particularly schwannoma, confirm MBs. However, the dural-tail-sign regrades as an unspecific hallmark mostly denoting meningioma, Ninh Ba Doan et al. represented an extra-axial midline tentorial MBs in adults who were associated with this indicator [2, 4, 10]. To the best of the authors' knowledge, only 12 prior cases of extra-axial CPA-MBs have been reported in the English-written literature [12].

Abbreviations: CPA, cerebellopontine angle; MB, medulloblastoma; CN, cranial nerve; MRI, magnetic resonance imaging; WHO, World Health OrganizationIt is well established that MBs have a marked propensity to metastasize to the leptomeninges, spinal canal, and supratentorial section via CSF and subarachnoid space, with an overall incidence of 38%-60% of all patients and a range of 20% to 40% of pediatrics populations. The spinal canal is the most common pathway at approximately 58%. Moreover, to the best of our knowledge, spinal metastasis from CPA MB has been reported just in one patient, followed by refusing postoperative radiotherapy. Recently, 2 patients suffering MB type SHH (desmoplastic and anaplastic) have been reported, manifesting leptomeningeal tumor dissemination [4, 10, 15, 18, 25-29]. The total survival chance of CPA MBs is similar to other regions, and with timely surgery and radiotherapy, the 5-year survival rate for MBs is 25%-70% [9, 22]. In almost half of the cases, recurrence occurs, and they survive for about 15 months after relapse [22]. Our patient did not observe residue or recurrences during the first postoperative year.

Microsurgery for total gross resection using the retro mastoid approach accompanied by craniospinal irradiation and chemotherapy is recommended as a safe and effective mainstay in children and adults to prohibit tumor growth [9, 10]. The operation aims to determine the histological diagnosis and deal with probable hydrocephalus. Approximately 10% to 30% of patients with MBs require postoperative ventricular shunting [14, 22].

Chemotherapy is less endured in adults and seemingly did not provide complete and valuable outcomes compared with the pediatric groups. Due to the lack of subsisting randomized control trials, a good understanding of differentiation between radiotherapy with or without adjuvant chemotherapy remained uncovered; consequently, chemotherapy is strongly recommended for adults in the high-risk group. In that respect, our patient underwent radiochemotherapy and was discharged in good clinical condition on the sixth day after the operation. Supplementary immunological procedures that have recently gathered momentum in adult cranial neoplasms represent Programmed Death-1 (PD-1) and the Cytotoxic T-Lymphocyte-Associated Antigen 4 (CTLA-4) as the agents, diminishing immune responses in both MBs and gliomas [12].

Conclusion

Regardless of the rare presentation of extra-axially CPA, neurosurgeons should be aware of this scarce entity. Clinical considerations and early radiologic evidence should be considered in subjects with suspected MB. A total tumor excision approach followed by aggregative chemotherapy/radiotherapy is designed to hinder tumor relapse.

Ethical Considerations

Compliance with ethical guidelines

Written informed consent was obtained from the patients for publication of their clinical details and clinical evidence.

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Authors' contributions

Conception and design: Kaveh Ebrahimzadeh; Data Analysis and Interpretation: Hesameddin Hoseini Tavassol; Drafting the article: Mohammad Mirahmadi Eraghi; Critically revising the article: Omidvar Rezaeimirghaed, Mohammad Hallajnejad; Reviewing the submitted version of the manuscript and approving the final version of the manuscript: All authors. Kaveh Ebrahimzadeh and Mohammad Mirahmadi Eraghi contributed equally to this work.

Conflict of interest

The authors declared no conflict of interest.

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