

Extra-axial medulloblastoma in cerebello-pontine angle: A report of a rare case with literature review

Eshagh Bahrami¹, Sahar Bakhti², Seyed-Mohammad Fereshtehnejad³,
Mansour Parvaresh⁴, Mohammad Reza Khani⁵

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Abstract

Medulloblastoma is quite uncommon in the adult population and even rarer in extra-axial site in cerebello-pontine (CP) angle. In this report, a 23-year-old male patient with a two month history of deafness, nausea, vomiting and ataxia is presented. Clinical and radiological findings demonstrated a heterogeneously enhanced extra-axial lesion in the right CP angle. Total excision was performed and the histopathological features of medulloblastoma were confirmed. After surgery, the patient had no neurological deficit and the audiometric findings were improved. In addition, he underwent adjuvant radiotherapy and no sign of metastatic mass was observed in follow-up spinal cord MRI. Although extremely rare, medulloblastoma must be considered in the differential diagnosis of extra-axial CP angle lesions.

Keywords: Medulloblastoma, Extra-axial, Cerebello-pontine (CP) angle, Adult.

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Introduction

Medulloblastoma is a common childhood tumor of the posterior fossa, which accounts for 21.8% of all pediatric primary malignant central nervous system tumors in Iran (1). However, this malignancy is quite rare in the adult population with the incidence rate of 0.4% to 1% among all primary adult brain tumors (2). Most of the cases with cerebello-pontine (CP) angle medulloblastomas are intra-axial, while only a few of them represents extra-axially that makes this site of tumor even extremely rare in adults (3). In the current report, we outline the clinical features, imaging results and surgical treatment of an extra-axial medulloblastoma in CP angle of a young man. Thereafter, relevant literature and the few similar cases are discussed in order to review the available evidences to manage this

rare site of tumor.

Case Report

A 23 year old man presented with a two month history of deafness of the right ear followed by nausea, vomiting and ataxia. Neurological examination yielded normal findings, except for audiometric examination that showed hearing loss and ataxic gait. In particular, audiometric assessment demonstrated a discrete sensorineural hearing loss of 50 dB at 1 KHz to 8 KHz. Magnetic resonance imaging (MRI) revealed a cystic and necrotic lesion in the right CP angle. The lesion was heterogeneously more hypointense on T1 and hyperintense on T2 (Fig. 1). The lesion enhanced on T1 weighted MRI images after injection of gadolinium (Fig. 2). Spinal cord MRI did not reveal any evidence of metastasis.

1. Department of Neurosurgery, Rasool-e-Akram Hospital, Iran University of Medical Sciences, Tehran, Iran. bahrami_eses@yahoo.com

2. Department of Neurosurgery, Rasool-e-Akram Hospital, Iran University of Medical Sciences, Tehran, Iran. dr_sahar_bakhti@yahoo.com

3. **(Corresponding author)** Division of Clinical geriatrics, Department of Neurobiology, Care Sciences, and Society (NVS), Karolinska Institutet, Stockholm, Sweden. sm.fereshtehnejad@ki.se

4. Department of Neurosurgery, Rasool-e-Akram Hospital, Iran University of Medical Sciences, Tehran, Iran. m_parvaresh@yahoo.com

5. Department of Neurosurgery, Milad Hospital, Tehran, Iran. dr_mr_khani@yahoo.com

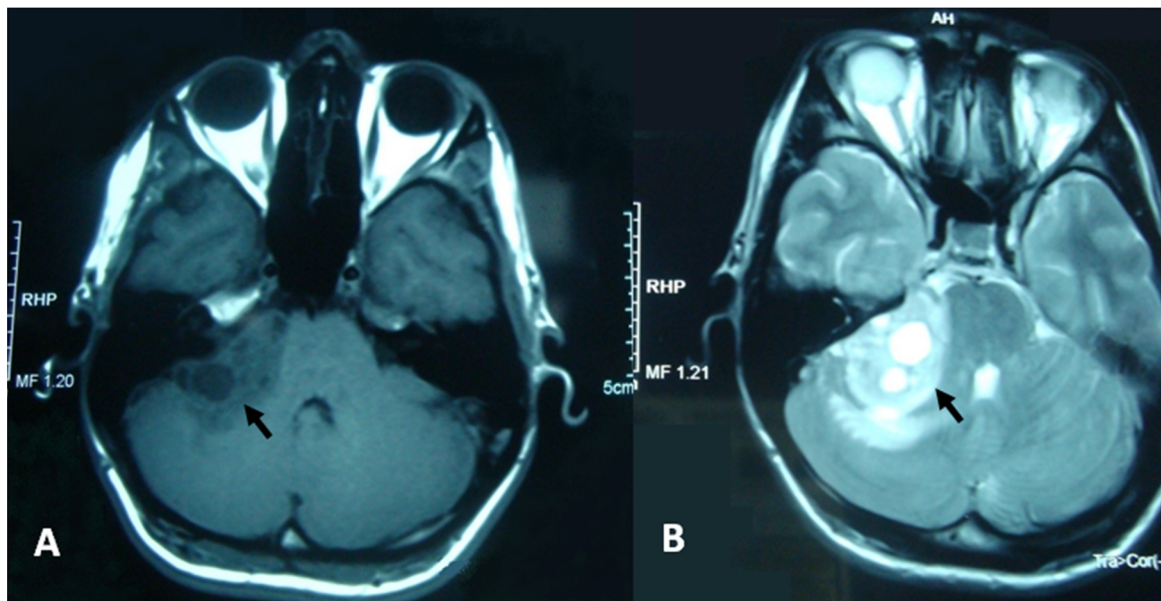


Fig. 1. Axial MRI shows the heterogeneous extra-axial lesion with cystic component on T1 (A) and T2 (B) in the right CP angle.

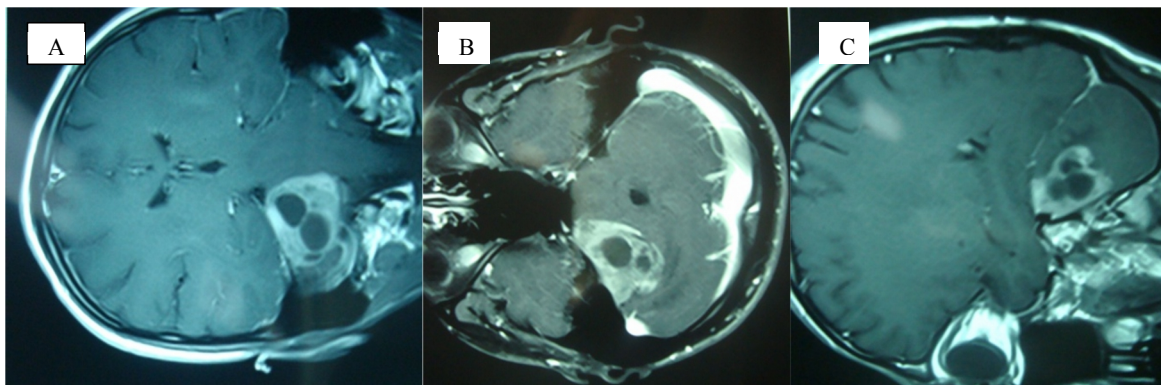


Fig. 2. Brain MRI in coronal (A), axial (B) and sagittal (C); Gadolinium injection shows a right CP angle lesion with heterogeneous enhancement of solid component and compression effect on the 4th ventricle

He underwent right retromastoid craniectomy and gross total excision of the lesion with facial nerve monitoring. There was a clear plane between the tumor and cerebellum, whereas it was adherent to tent laterally. Histopathologic evaluation showed “pale islands” consisting of micronodular, reticulin-free zones with a low magnification appearance similar to follicular lymphoid hyperplasia. The lesion was characterized by reduced cellularity, a rarefied fibrillar matrix and compact undifferentiated area (Fig. 3). After surgery, the patient had no neurological deficit and his audiometric findings were improved. The post-operative MRI showed total extirpation of the tumor (Fig. 4-A). In addition, he underwent radiotherapy as adjuvant treatment.

The latest follow-up was performed one-year post-operation by means of brain MRI, which shows no lesion in the right CP angle (Fig. 4-B).

Discussion

Medulloblastoma is a predominantly pediatric tumor commonly occurring intra-axially in the cerebellar vermis as the most common site of origin (4). Origin of medulloblastoma may be either from germinal cells or their remnants situated at the end of inferior medullary velum or from remnants of the external granular layer, however, the exact origin is not certainly known yet (5-6). Adult medulloblastomas usually arise from the surface of the cerebellum or pons and 50% of these are laterally located (5,

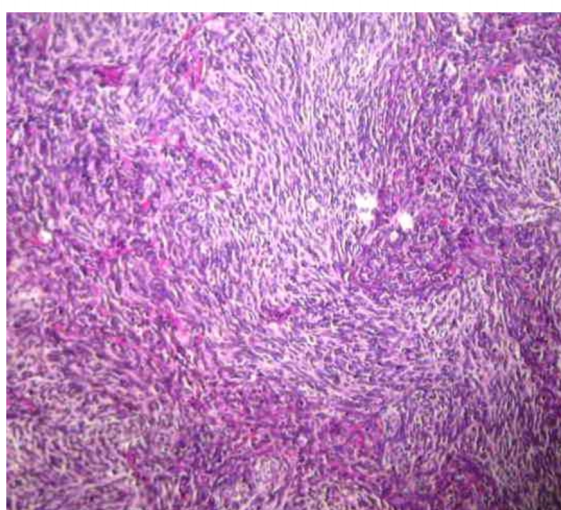
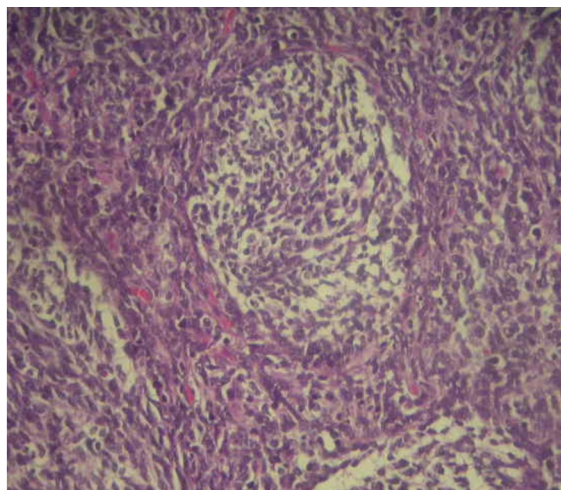


Fig. 3. Desmoplastic/nodular medulloblastoma. Microphotograph of histopathology slide (H and E staining). Micronodular zones of reduced cellularity (pale islands) are a striking feature of this medulloblastoma variant.

7). Extra-axially, the tumors have been

localized in the tentorial region or the CP angle (3, 8). In another point of view, there are only a few cases of CP angle medulloblastomas and most of them are intra-axial, which makes the extra-axial site of this tumor extremely rare. Recently, two misdiagnosed adult cases of medulloblastoma of the CP angle are reported, which were wrongly marked as vestibular schwannoma and petrosal meningioma during the pre-operative radiological assessment (9).

Development of this tumor in the CP angle may be from the remnants of the external granular layer in the cerebellar hemisphere, including the flocculus which faces the CP angle (10-11). It may grow to occupy the CPA through two ways including lateral extension from the 4th ventricle through the foramen of Lushka, or direct exophytic growth from the site of origin at the surface of the cerebellum or pons (6). The lack of association with any cerebellar tissue and extra-axial location in the region of CP angle is an extremely rare phenomenon. Based on the latest literature review, less than 10 cases of extra-axial adulthood medulloblastoma have been reported in the CP angle.

Table 1 summarizes the characteristics, clinical manifestation and treatment work-up for these reported cases. During the adulthood, patient age at diagnosis ranges from 21 to 52 years, mostly often occurring in the late 20s and early 30s (12) including

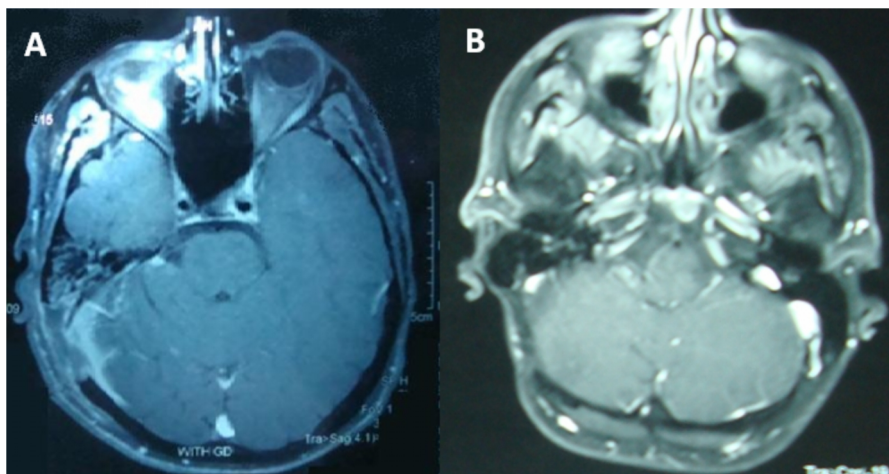


Fig.4. Post-operative MRI shows total resection of tumor with no detectable enhancement in the right CP angle promptly after surgery (A) and one-year post-operation follow-up (B)

Table 1. Characteristics and work-up of published adult cases with extra-axial medulloblastoma in cerebello-pontine angle

Author	Year	Patient's Age	Patient's Sex	Presentation	Neurological Examination	Treatment
Becker et al. ^[8]	1995	32 yr	Female	Headache, vomiting	-	-
Akay et al. ^[6]	2003	21 yr	Male	Headache, nausea, vomiting, ataxia	Bilateral papilloedema, hemiparesis, hemihypesthesia	Partial excision, radiotherapy, chemotherapy
Gil-Salú et al. ^[12]	2004	40 yr	Male	Headache, vomiting, hearing difficulties	One-side trigeminal 1st and 2nd nerves deficit	Total excision, adjuvant therapy
Fallah et al. ^[3]	2009	47 yr	Male	Headache, nausea, vomiting	Normal	Tumor resection, radiotherapy
Furtado et al. ^[5]	2009	32 yr	Male	Headache, Vomiting, gait unsteadiness	Papilloedema, right-sided dymetria, dysdiadokokiniasia, gait ataxia	Total excision, referred to radiation oncologist for further management
Singh et al. ^[18]	2011	21 yr	Male	Headache, vomiting, ataxia, left facial weakness	Papilloedema, left lower motor neuron facial paresis, left IX and X cranial nerve paresis, left cerebellar signs	Total excision, failed to receive radiotherapy (recurrent lesions and metastasis after 15 months)
Spina et al. ^[9]	2013	22 yr 26 yr	Male Female	Headache, hearing loss, tinnitus, dizziness and ataxia	Weakness of the right arm, slight left nystagmus and a mild peripheral deficit of the left facial nerve	Total tumor resection, radiotherapy
Present Study	2013	23 yr	Male	Deafness, nausea, vomiting, ataxia	Hearing loss in audiometry, ataxic gait	Total excision, radiotherapy

the current case in our report with 23 years of age. It seems that a male preponderance is also seen for the reported cases. Regarding the clinical manifestations, no specific features have been ascribed to CP angle medulloblastomas. However, there are some characteristics that may help distinguish them from other CP angle lesions, mainly the most common tumor, acoustic neuroma followed by meningiomas, primary cholesteatomas and epidermoid tumours (3, 13). Involvement of the Vth, VIth, VIIth, VIIIth and lower cranial nerve and signs of cerebellar dysfunction are commonly noted in CP angle lesions (14). Early onset of progressive cerebellar signs and gait ataxia may indicate an axial origin of tumor, whereas positional nystagmus may be an early sign suggestive of an acoustic schwannoma (11). Hearing impairment and VIIth nerve involvement is usually less common and a late feature of a CP angle medulloblastoma; however, this feature has been even reported as initial symptom in a

few cases (11) including ours that also complained from diminution of hearing as initial symptom. As shown in Table 1, headache, nausea, vomiting and ataxia are among the most common symptoms reported in these cases. Related findings could be found in neurological examination such as papilloedema, hemiparesis, gait disturbances, facial paresis and etc.

As we also found in our reported case, MRI assessment often shows heterogeneously gadolinium-enhancement lesions (8). However, there are some previous reports where the tumor has demonstrated a homogenous enhancement pattern, which may lead to misdiagnosis (3, 12).

Histopathologically, desmoplastic variant of medulloblastoma is more common in adults than the classical type (5). This variant is hemispheric in location and is more often associated with cysts and necrosis in comparison with classical medulloblastomas; however, there are no pathognomonic MRI criteria to differentiate between these

two types (15-16). Medulloblastomas are known to metastasize through cerebrospinal fluid into the spinal canal, leptomeninges, and supratentorial regions. Metastasis in medulloblastomas varies between 38 and 60% in various series, with the spinal canal being the commonest site with approximately 58% (17). Spinal metastasis from CP angle medulloblastoma is very rare and until today just one case was reported with involvement of spine where the patients failed to receive post-surgical adjunctive radiotherapy (18). We followed the patient for probable metastasis by spinal cord MRI and no metastasis is reported yet.

Adult medulloblastomas are treated similar to pediatric medulloblastoma (14). A highly suspicious medulloblastoma should be purposed in atypical posterior fossa lesions in adults (11). A pre-operative diagnosis based on clinical assessment and radiological findings supports complete resection followed by beneficial adjuvant therapy.

Conclusion

Conclusively, although extra-axial site of adulthood medulloblastoma is extremely rare, this tumor must be considered in the differential diagnosis of extra-axial CP angle lesions. Appropriate diagnostic and surgical work-up should be performed including total resection and adjuvant therapy. Any neurological deterioration seen in followed-up patient must be evaluated for metastasis.

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