Case Report

Uncommon Tumor of Cerebellopontine Angle: Report of an Adult Medulloblastoma

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Abstract

Medulloblastomas are relatively common tumors of posterior fossa during childhood with a frequency of 23 – 30%. However, adult cerebellar medulloblastomas are relatively rare and constitute 0,4 – 1,0% of adult primary brain tumors. Cerebellopontine angle (CPA) is an atypical site for medulloblastoma. We present a 34-year-old male with a CPA medulloblastoma. We recommend appropriate surgical planning whether extending to midline structures or fourth ventricle. CPA medulloblastomas are infrequently reported at the CPA region and have to be considered for differential diagnosis of other intra-axial or extra-axial CPA tumors. Adjuvant radiotherapy and combined chemotherapy regimen should be added at the postoperative management.

Keywords: Pontocerebeller angle, medulloblastoma, adult, surgery

INTRODUCTION

Medulloblastomas are primary neuroepithelial tumours that mainly occurs midline in the posterior fossa of children⁴. Whereas, adult cerebellar medulloblastomas are relatively rare, presenting mostly during the 3rd or 4th decade of life and constitutes 0,4 – 1,0% of all adult primary brain tumors²-⁴. Atypical sites like cerebellopontine angle (CPA) cistern are very uncommon and most of the reported cases were adults with age ranging between 19 and 46 years⁵. Adult medulloblastomas could possibly mimic imaging characteristics of meningiomas or vestibular schwannomas if they extend into the internal auditory canal or extracerebellar localizations³. In the present report, an adult medulloblastoma extending to

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the CPA which appears as an extraaxial tumor was discussed with an emphasis on differential diagnosis.

**CASE PRESENTATION**

A 34-year-old male was admitted with complaints of nausea/vomiting, headache and ataxic gait. On neurological examination, bilateral papilledema and slight left hemiparesis were predominant features. Magnetic resonance imaging (MRI) revealed a heterogeneously enhanced mass at CPA region with a cystic midline component and slight hydrocephalus due to mass effect on fourth ventricle. Axial and coronal MRI confirmed the attachments of the mass to left petrosal dura and inferior surface of the tentorium (Figure 1a–b). The mass was thought to be extraaxial and meningioma or acoustic schwannoma was included in differential diagnosis according to the preoperative radiological and clinical data. At the operation, suboccipital craniectomy was performed for the removal of tumor and adjacent cystic component. Every effort was made to remove the medial extension of the tumor. Postoperative period was uneventful with complete clinical improvement. Histopathological examination revealed a tumor composed of sheets of immature anaplastic cells with abundant mitosis. Tumor cells are pleomorphic small cells, with little cytoplasm and hyperchromatic nuclei that are mostly elongated shaped. Immunohistochemical staining confirmed the diagnosis of medulloblastoma with cytoplasmic synaptophysin and chromogranin-an expression in the tumor cells (Figure 2a–b–c). In the light of these findings, the tumor was identified as a “classical medulloblastoma” (ICD–9 CM; 9470/3) according to WHO classification in 2007.

Adjuvant radiotherapy was given with matching spinal and cranial fields using fraction size of 1.8 Gy per day (totally 54 Gy). Then, combined chemotherapy regimen was administered and consisted of Cyclophosphamide, Vincristine and CCNU at the postoperative period. The patient was neurologically normal at the sixth month of follow-up and MRI showed almost complete removal with a small remnant to the midline (Figure 3a–b).

![Figure 1](image_url)

*Figure 1*: Axial (a) and coronal (b) gadolinium enhanced MRI views show the attachments of the tumor to left petrosal dura and inferior surface of the tentorium
Acoustic neuromas and meningiomas constitutes the majority of CPA tumors however a large spectrum of many unusual lesions form the remaining group of CPA lesions\(^{(3,6)}\). Origin of tumor is the main determinant of differential diagnosis of unusual PCA lesions. Here we present an unusual medulloblastoma localized in CPA with a midline etension.

Most frequent symptoms of posterior fossa tumors in young adults are headache, nausea/vomiting, vertigo/dizziness, tinnitus and hearing loss, however all these symptoms are nonspecific\(^{(5)}\). Besides, CPA medulloblastomas do not present with distinctive clinical, neuro-otological or neuro-radiological findings\(^{(9)}\). Absence of hearing disturbance and VIIth nerve involvement may help to distinguish them from acoustic neuroma\(^{(4,5)}\). Involvement of 7th nerve is usually non-specific and is also a frequent finding of other CPA tumors.

Radiological data is frequently helpful to differentiate the tumor from acoustic neuromas and meningiomas which form the majority of CPA tumors. MR imaging

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**DISCUSSION**

*Figure 2*: a) The sheets of small immature anaplastic cells with hyperchromatic irregular nuclei and scant cytoplasm are typical. Hematoxylin and Eosin Stain (H&E), x400 b) Synaptophysin expression of tumor cells, x200. c) Chromogranin-A expression of tumor cells, x200

*Figure 3a – b*: Gadolinium enhanced axial (a) and coronal (b) MRI views six months after the operation showed a retained midline contrast enhanced tissue without any mass effect.
shows typical features and gives satisfactory knowledge to establish a diagnosis of acoustic neuroma and meningioma\(^{2,3}\). The remaining portion of CPA lesions form a wide spectrum and lesions in cystic nature further complicate the differential diagnosis of CPA lesions\(^2\).

Adult cerebellar medulloblastomas are usually encountered in the third and fourth decades of life and constitute 0.4 – 1.0% of all adult primary brain tumors which is relatively rare\(^{4,5}\). They are unusually located in the cerebellar hemisphere of adults\(^5\). Medulloblastomas are generally located to the midline of the cerebellum. They may also be defined as hyperdense enhancing soft tissue mass with perilesional edema and associated obstructive hydrocephalus\(^2\). The SIOP II trial reviewed 233 cases and showed ‘typical’ features in only 30% of cases\(^8\). The primary lesions were cystic in 59% of the patients, calcified in 22%, cystic and calcified in 15%, ill defined in 7%, not associated with edema in 5% and not accompanied by hydrocephalus in 4%. Eighteen percent of tumours were nonvermal, while 15% were already metastatic at presentation\(^{6,8}\). CPA medulloblastomas appear as round or ovoid tumors with smooth margins, are more common in the cerebellar hemisphere than in the vermis, and often extend to the brain surface with possible exophytic invasion of the CPA. Irregularity of some portions of the tumor-brain interface is a clue to their intraaxial origin\(^2\).

Medulloblastoma arises from germinal cells (or their remnants) anywhere along their migratory path. It was also suggested that because the migratory process normally proceeds in a lateral direction there will be a relatively high frequency of laterally situated tumours in adults\(^{5,6,7}\). However, other sites of origin of CPA medulloblastomas have been proposed, including the lateral medullary velum and the flocculus of the cerebellum\(^6\). Medullablastomas may grow to occupy the CPA through two pathways: lateral extension of fourth ventricle through the foramen of Luschka, or direct exophytic growth from the site of origin at the surface of the cerebellum or pons\(^1\).

In the present case, we performed left suboccipital craniectomy for tumor resection at CPA however the tumor was detected to extend significantly to midline which makes the tumor resection much more difficult without cerebellar retraction. Since the approach was inadequate for total resection, we removed as much tumor as possible. The goal of surgical treatment for CPA tumors is maximum cytoreduction, histopathological diagnosis and restoration of cerebrospinal fluid flow while avoiding brainstem manipulation and cerebellar injury\(^1\). Adjuvant radiotherapy and chemotherapy should be included to the treatment following surgical resection of medulloblastomas\(^4\). This growing trend of adjuvant treatment increases the 5-year survival rates to better rate of 70% for medulloblastomas. In the present report, size of the remnant tumor was reduced after adjuvant therapies at the 6th month of follow-up.

**CONCLUSION**

CPA medulloblastoma is an extremely rare entity and should be considered for differential diagnosis of intra-axial CPA tumors. Surgical planning should be planned according to the extension of the tumor to midline structures or fourth ventricle.

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