Extra-axial Cerebellopontine Angle Adult Medulloblastoma: An Unusual Presentation at an Unusual Site

Vishwaraj Ratha[1] Vikram S. Karmarkar[2] Chandrashekhar E. Deopujari[3]

[1]Department of Neurosurgery, SRM Institute of Medical Sciences, Vadapalani, Chennai, Tamil Nadu, India
[2]Department of Neurosurgery, Bombay Hospital Institute of Medical Sciences and Research Center, Mumbai, Maharashtra, India
[3]Department of Neurosurgery, Bombay Hospital Institute of Medical Sciences and Research Center, Mumbai, Maharashtra, India

Address for correspondence Vishwaraj Ratha, MBBS, MS, Mch, Department of Neurosurgery, SRM Institute of Medical Sciences, 100 Feet Road, Jawaharlal Nehru Salai, Vadapalani, Chennai, Tamil Nadu, India, Pin code 600026 (e-mail: thinkvishwaraj@gmail.com).

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Abstract

Medulloblastoma is a rare adult primary brain tumor and its presence in the cerebellopontine angle (CPA) constitutes an even rarer site for origin. There are only 40 reported cases of CPA medulloblastoma in the literature, with most being intra-axial and occurring in the pediatric population. Extra-axial CPA medulloblastoma in adults are extremely rare and only 10 adult cases have been reported in world literature.We report a rare case of primary extra-axial CPA medulloblastoma occurring in an adult. We review the literature for such an unusual presentation of medulloblastoma in adults and discuss its appropriate clinico-radiological features along with its possible pathogenesis and surgical management.With increasing reporting of such tumors, CPA region, medulloblastoma should be considered a differential diagnosis of intra-axial or extra-axial CPA tumor, as their further management and prognosis differ significantly. Adjuvant radiotherapy and combined chemotherapy regimen should be added to the postoperative management.

Keywords
- cerebellopontine angle medulloblastoma
- extra-axial
- differential diagnosis
- adult

Introduction

The term medulloblastoma cerebelli was coined in 1925 for poorly differentiated cerebellar tumors by Bailey and Cushing.[1] It is the most common childhood intracranial tumor, accounting for 25% of all pediatric intracranial tumors and 33% of all posterior fossa neoplasm in children.[1]

In adults, the tumor is uncommon, accounting for approximately 1% of adult primary brain tumors and 6% of posterior fossa tumors, 80% of which occur before the end of the fourth decade.[1,2] The incidence of adult Medulloblastoma is approximately 0.5 per million per year and decreases with increasing age. The published studies on adult medulloblastoma are usually retrospective, owing to its rarity in adult populations, and are mostly midline (cerebellar vermis).[1,2]

We report an extremely rare occurrence of adult cerebellopontine angle (CPA) medulloblastoma. This case underlines the existence of the extremely rare intra-axial adult tumor in this unusual location, mimicking as a purely extra-axial lesion, thereby confounding its diagnosis. It also demonstrates the difficulties encountered when relying on an imaging diagnosis for medulloblastoma, especially in adults. There are only 40 reported cases of CPA medulloblastoma in the literature, with most being intra-axial.[2-5] Extra-axial CPA medulloblastoma is extremely rare and only 10 adult cases have been reported in world literature.[6]

Case Report

A 42-year-old female patient presented with complaints of progressive headache and ataxia over a duration of 2 months. On neurological examination, her vision was normal with no papilledema. She had no cranial nerves deficit or any other focal neurological deficit, except for left cerebellar signs and

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nystagmus. Magnetic resonance imaging (MRI) showed a heterogeneous lesion more hypo-intense than the gray matter on T1-weighted images (T1WIs) and hyper intense on T2-weighted images (T2WIs) (►**Fig. 1A**). There was heterogeneous enhancement of lesion after administration of contrast. Axial and coronal imaging revealed the attachments of lesion to the left posterior petrosal dura and inferior surface of tentorium. On corelating the preoperative clinical and radiological findings, the common extra-axial CPA tumors, such as meningioma and acoustic neuroma, were considered as differential diagnoses.

She underwent a standard retromastoid approach and the lesion was exposed through the left CPA. The lesion was grayish white and solid but soft and friable in consistency also, easily suckable. There was a clear plane between the tumor and cerebellum; however, it was also adherent to dura and tent laterally. We could achieve a complete excision of tumor. Hence, our intraoperative inference was that of an extra-axial soft dural based tumor, probably meningioma. Her postoperative period was uneventful. She had no focal neurological deficit and her cerebellar signs also gradually improved. The postoperative CT scan revealed no definite residual or recurrent mass (►**Fig. 1B**).

However, the histopathology (HP) showed a highly cellular tumor composed of rosettes of small round cells, with high nucleus–cytoplasm ratio and increased mitotic figures—suggestive of classical medulloblastoma—WHO grade IV (►**Fig. 2A**). Immunohistochemical study revealed synaptophysin, S-100 protein, and neuron-specific enolase to be positive but GFAP, neurofilament, and cytokeratin were negative (►**Fig. 2B**).

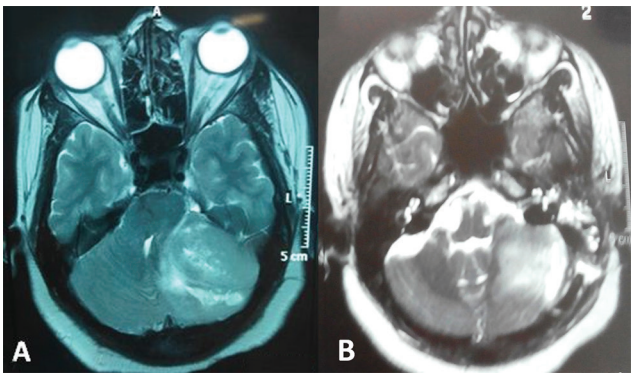

Fig. 1 (**A**) Preoperative MRI brain hyperintense on T2W axial. (**B**) Postoperative MRI brain showing complete excision of the tumor.

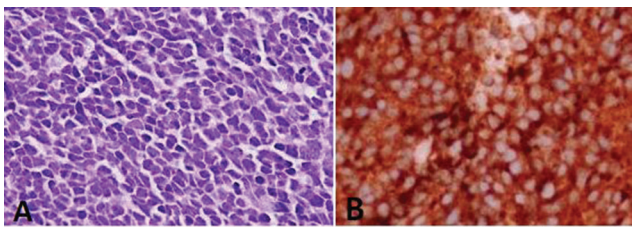

Fig. 2 (**A**) **H&E:** Classic medulloblastoma showing a diffuse pattern of tumor growth with poor cellular differentiation, nuclear molding, and minimal indistinct cytoplasm (**B**) **IHC:** Synaptophysin positivity in medulloblastoma shows as a brown staining of the cells.

In view of the histopathological diagnosis being contrary to our preoperative impression, the specimens were reviewed again by a second pathologist, but the diagnosis remained the same.

After the HP result was reported, MRI of the spinal cord was performed and it revealed no evidence of metastasis to the spinal cord. The patient underwent cranio–spinal radiotherapy. Magnetic resonance images obtained 3 months postoperatively demonstrated no residual or recurrent mass. She has been under regular follow-up for 15 months with no recurrences or metastases yet.

Discussion

In 2007, WHO designated it as a distinct embryonal tumor, distinguishing it from other primitive neuroectodermal tumors (PNETs).[2] Medulloblastoma is predominantly a pediatric tumor accounting for one-fourth of all pediatric intracranial tumors. The most common site is the cerebellar vermis, from where it penetrates the fourth ventricle, resulting in abnormalities in the flow of cerebrospinal fluid (CSF). In 10 to 30% of pediatric cases, it has been observed to spread along the CSF routes and become disseminated within the central nervous system. The radiological features of medulloblastoma are classical: they display an iso- or hypointense signal on T1WI, are heterogeneous on T2WI, and exhibit homogeneous enhancement after addition of gadolinium, sometimes demonstrating a central hemorrhagic zone (our preoperative diagnosis of CPA meningioma was due to age of the patient at presentation and imaging features of well-demarcated, broad-based, extra-axial, homogenously contrast-enhancing lesion). Medulloblastoma in adults is an uncommon entity (1% of adult primary brain tumors) and its presence in CP angle region is exceptionally rare. There have been only 10 reported cases of extra-axial medulloblastoma in the adult literature.[6] The table describes CPA medulloblastomas described in the past, presentation, management, and outcome compared to the present case (►**Table 1**); however, they are likely to be underreported owing to publication bias. This tumor is nearly twice as common in men. The tumors most often occur among patients in their late 20s and early 30s.[6] It has been observed that the two most common sites for extra-axial locations are the tentorial and CPA regions.[3,6-12] Most commonly, they manifest as heterogeneously enhancing lesions upon contrast administration.

There are currently two general hypotheses regarding the origin and spread of medulloblastoma. In the first, medulloblastomas are proposed to arise from primitive multipotential cells in the external granular cell layer in the cerebellar hemisphere, mainly the flocculus which faces the CPA. The alternative view is that medulloblastomas arise from multipotential cells in the subependymal region and within the fetal pineal region, giving rise to all PNETs, regardless of location.[1] In other words, medulloblastoma can occur anywhere (including CPA) along the germ cell tumors' normal migration course to lateral side.[1,13]

In our case, both the preoperative and intraoperative findings pointed toward an entirely extra-axial tumor

Table 1 List of various CPA medulloblastomas described in the past, the presentation, course of management, and follow-up

Author and year	Age and sex	Duration of symptoms	Presentation	Treatment	Follow-up
Becker et al 1995[10]	32 F	–	H, NV	–	–
Akay et al 2003[15]	21 M	2 months	H, V, ataxia, B/L PE, HP, HA	PE, CT, RT	18 months
Gil Salu et al, 2004[9]	40 M	–	H, NV, HL, NV inv	TE, AT	–
Fallah et al, 2009[3]	47 M	–	H, NV	TE, RT	–
Furtado et al, 2009[11]	32 M	3 weeks	HNV, ataxia, B/L PE, CS	TE, AT	–
Singh et al, 2011[8]	21 M	1 month	H, NV, ataxia, B/L PE, left VII, IX, X, CS	TE only	Recurrence and metastasis at 15 months
Spina et al, 2013[12]	22 M	3 months	H, HL, ataxia, L Nystagmus	TE, RT	–
Spina et al, 2013[12]	26 F	Chr H	H, HL, ataxia, rt arm weakness, L VII	TE, RT	–
Bahrami et al, 2013[7]	23 M	2 months	HL, NV, ataxia	TE, RT	12 months
Goudihalli et al, 2016[6]	50 M	1 month	L VII, VIII, IX, X, H	PE, abandoned due to bleeding	Vegetative
Present case, 2018	42 F	2 months	H, NV, ataxia, left CS	TE, RT	15 months

Abbreviations: AT, adjuvant therapy; B/L PE, bilateral papilledema; CPA, cerebellopontine angle; CS, cerebellar symptoms; CT, chemotherapy; H, headache; HA, hemianesthesia; HL, hearing loss; HP, hemiparesis; NV, nausea and vomiting; PE, partial excision; RT, radiotherapy; TE, total excision.

without any association with cerebellar tissue; however, subsequent histopathology report of medulloblastoma made our patients' case interesting and rare. After reviewing the literature, the 5-year survival rate has been reported to be around 30% for medulloblastomas in this location after surgery and radiotherapy,[1] while there have been cases that have improved with the use of combination chemotherapy such as vincristine-based regimens of chemotherapy.[10] Few cases have shown improvement with combination of chemotherapy and radiation therapy; however, surgery along with chemotherapy has remained the mainstay of treatment. So far, there is no clear-cut consensus as to whether the CPA medulloblastomas are more aggressive compared with their vermian counterpart.[6,14] We believe this may indicate a probable higher malignant potential of CPA medulloblastomas in comparison to their vermian counterpart. Surgery via the retromastoid route, followed by radiotherapy, remains the main treatment modality to manage these cases.

Conclusion

Over the years, medulloblastomas have shown a remarkable degree of heterogeneity in terms of their presentation, radiological diagnosis and biological behavior. Although considered to be a common pediatric intra-axial tumor, increasing solitary reports of it presenting extra-axially in CPA region mandates it to be included in the differential diagnosis of CPA tumors. This rare presentation at this unusual location may mislead its proper diagnosis and delay the required adjuvant therapy.

Conflict of Interest
None declared.

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