

Medulloblastoma of the Cerebellopontine Angle in a Child: A Rare Entity

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ABSTRACT

Cerebellopontine (CP) angle Medulloblastoma is a quite uncommon presentation in clinical practice, especially in children. We report such a case in a 12 years old female patient with clinical history of headache, altered sensorium, vertigo, gait ataxia, left side facial paresis and hearing loss. Preoperative axial MR imaging showed an ill-defined heterogenous lesion in left CP angle region with involvement of left middle cerebellar peduncle, left cerebellar hemisphere and cerebellar vermis. Post contrast scan showed heterogenous enhancement. Intra operative frozen cytology was suggestive of malignant round cell neoplasm. The subsequent histopathology and immunohistochemistry favoured the diagnosis of Medulloblastoma.


Key Words: Medulloblastoma, Cerebellopontine (CP) angle tumor, Child, MRI.

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INTRODUCTION

Medulloblastoma (MB) is a highly malignant neuroepithelial tumor of the posterior fossa classified WHO grade IV. Medulloblastoma is mainly a brain tumor of pediatric age that account for 10% of all intracranial neoplasms and 29% of all posterior fossa tumors in children whereas they accounts for less than 1% of CNS adult neoplasms.¹⁻³ In children most medulloblastomas (MBs) are encountered in the vermis whereas in adults the cerebellar hemispheres is usual site.¹⁻³

Typical and atypical MRI features have been reported in MBs: mostly present with well-defined margins, isointense signal on T1, hyperintense signal on T2-images, and marked enhancement on T1- postcontrast scan. In rare cases they present with ill-defined margins, hypointensity signal on T2, mild or nodular enhancement.⁴⁻⁶

Cerebellopontine angle (CPA) is an uncommon site and only 39 cases have been reported to date in best of our knowledge. We report a further case of CPA MB in a 12 year old female child.

CASE REPORT

A 12 year old female who presented with a headache, altered sensorium, vertigo, gait ataxia, left side facial paresis and hearing loss around 8 months before admission. BERA examination was indicative of retrocochlear pathology. Non contrast CT scan showed hyperdense mass lesion in left cerebellopontine angle with supratentorial extension causing effacement of 4th ventricle, compression over brain stem with peri-lesional oedema. MRI brain with GD showed presence of large ill-defined heterogenous

intensity lesion of size 53x53x 58 mm noted in the left CP angle region extending in the pons with involvement of left middle cerebellar peduncle. It was causing effacement of 4th ventricle and was involving left cerebellar hemisphere and cerebellar vermis. The tumor was showing heterogenous enhancement on post GD contrast scan. The patient underwent a left suboccipital craniotomy with tumor decompression. On exposure of the left CPA, a whitish grey white mass, soft in consistency was found. The tumor was partially suckable and vascular. The intra operative frozen cytology showed fragments of hyperchromatic cells with round nuclei, high N/C ratio at places arranged in rosette pattern and was suggestive of malignant round cell neoplasm.

We subsequently received several grey white soft tissue pieces measuring 5x5 cm for histopathological examination in our department. The histopathological examination was suggestive of medulloblastoma (WHO Grade IV). The subsequent immunohistochemistry showed synaptophysin positivity and was negative for GFAP. MIB score was 20%, which favoured the final diagnosis of medulloblastoma. The patient died two days after the surgery.

DISCUSSION

The most common cause of childhood oncological death is brain tumors of which medulloblastoma is the most common malignant pediatric brain tumor.⁷ MB is an embryonic neoplasm of central nervous system arising from undifferentiated neuroepithelial cells (primitive neuroectodermal tumor, PNET).⁸ MBs are the most

common posterior fossa pediatric tumor, with approximately 50% of then encountered in the first decade of life. The tumor usually originate from the vermis and extends into the fourth ventricle.^{1,2}

Very rarely MBs develop in the CPA and only 39 cases have been reported in the literature to date. Mean size at time of presentation is of 30 mm.

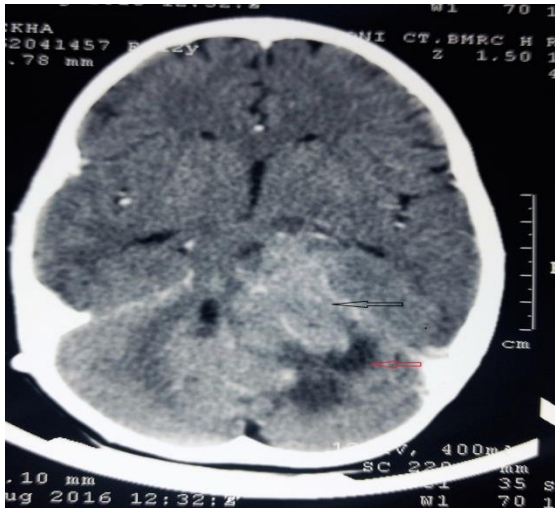


Fig 1: Axial Non contrast CT scan showing hyperdense mass lesion in left cerebellopontine angle (black arrow) causing effacement of 4'th ventricle with peri-lesional edema.(Red arrow)

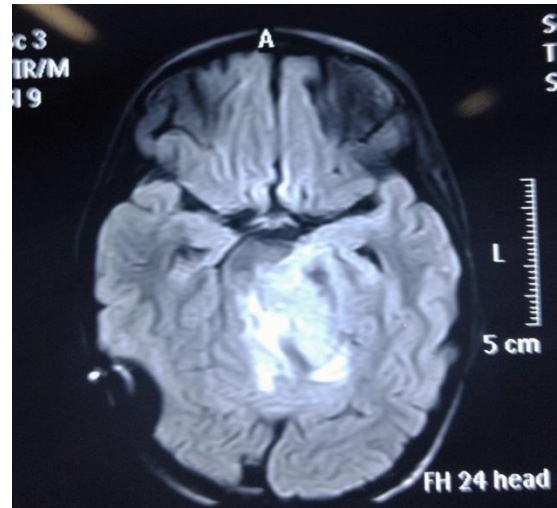


Fig 2: Axial T1 weighted MRI shows illdefined heterogenous intensity lesion in left CP angle extending in the pons with involvement of left cerebellar peduncle.

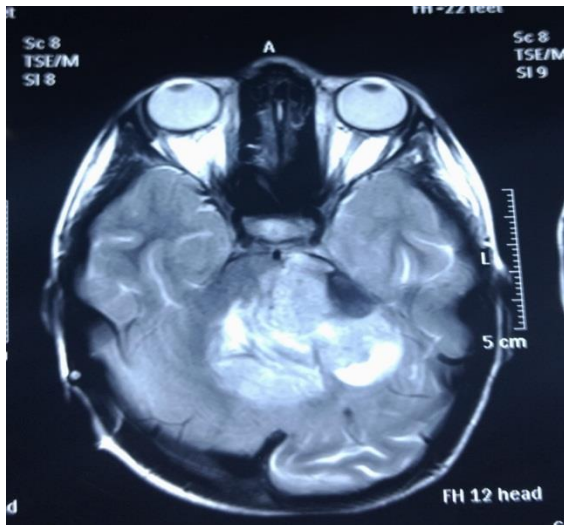


Fig 3: T2 weighted MRI shows illdefined heterogenous intensity lesion in left CP angle extending in the pons with involvement of left cerebellar peduncle.

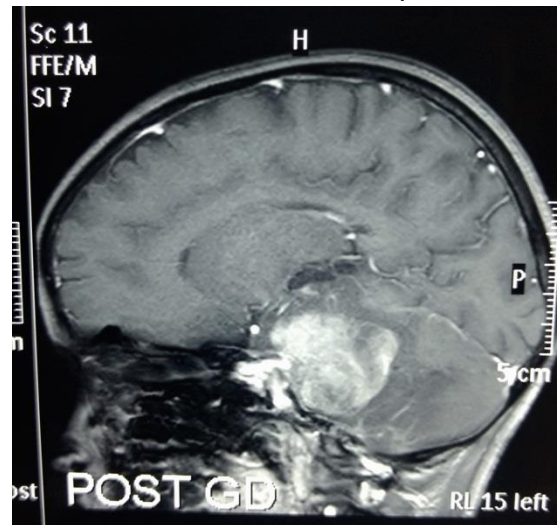


Fig 4: Sagittal view of post GD contrast scans showing heterogenous enhancement.

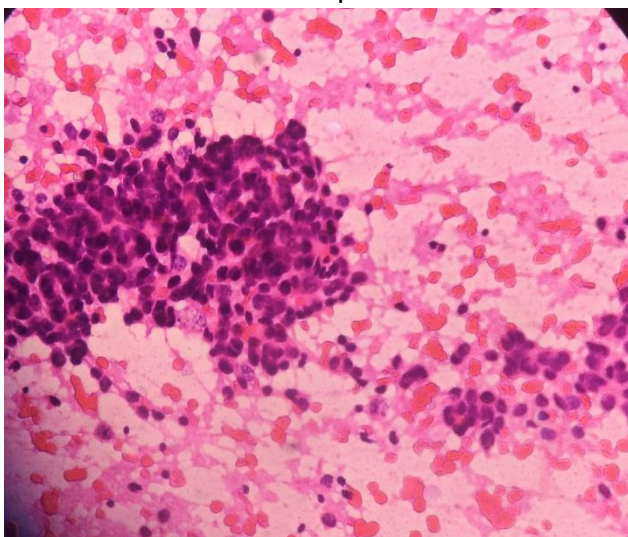


Fig 5: Intra operative frozen cytology showing round hyperchromatic cells with high N:C ratio.

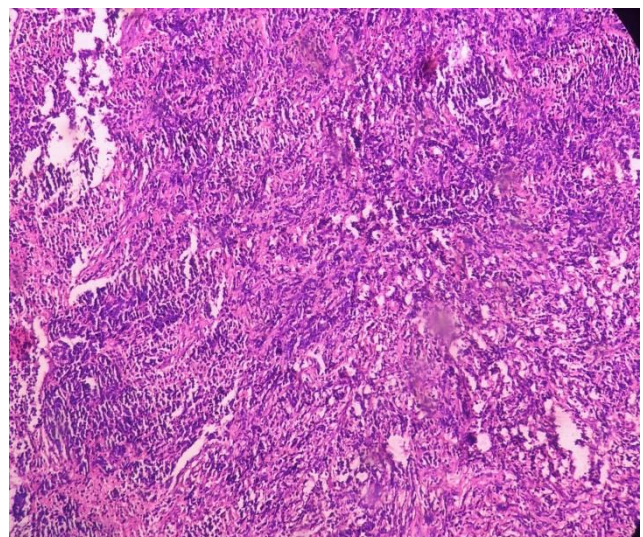


Fig 6: Low power (10X) view of H&E stain showing round hyperchromatic malignant cells infiltrating into the brain parenchyma.

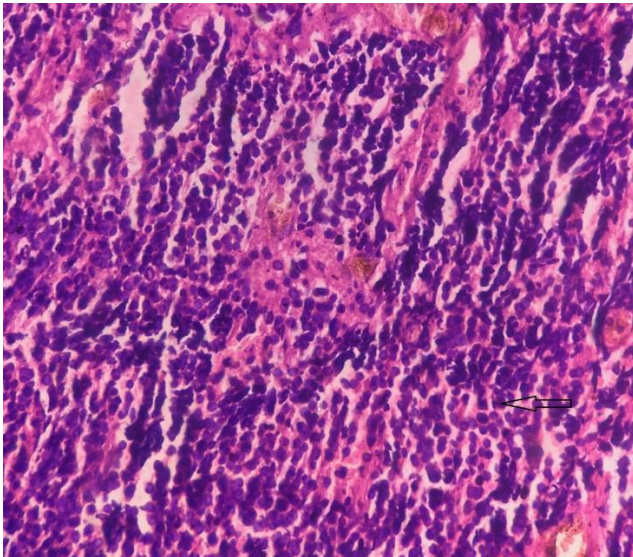


Fig 7: High power (40X) view showing round hyperchromatic cells with high N:C ratio at places arranged in rosette pattern (arrow).

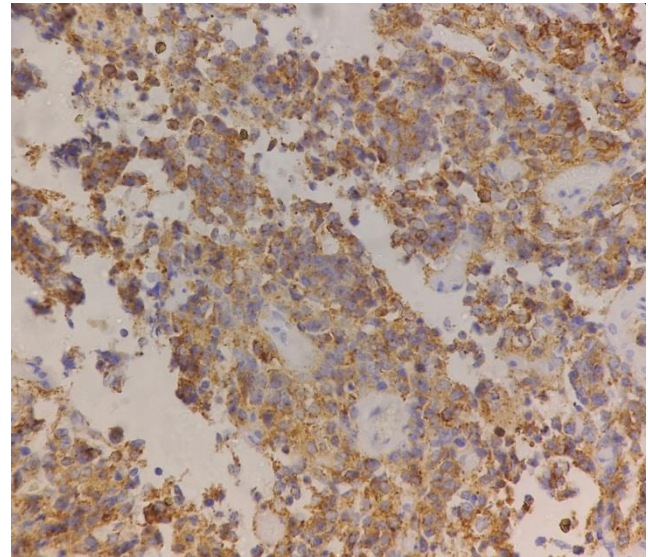


Fig 8: Malignant cell showing synaptophysin positivity

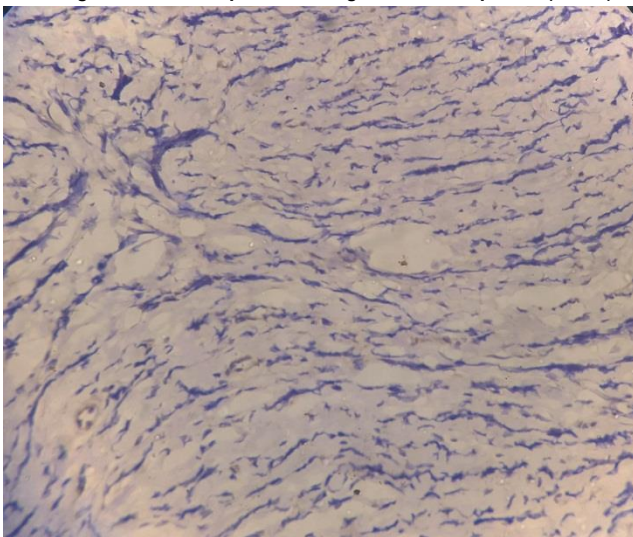


Fig 9: Tumor show negative immunostaining for GFAP.

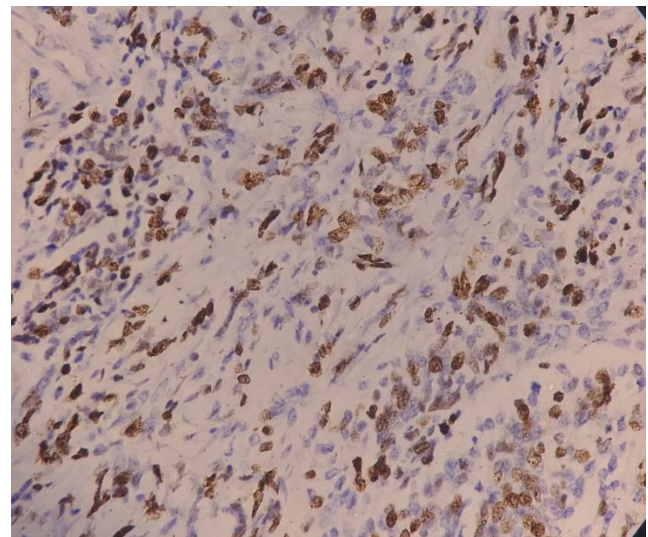


Fig 10: MIB-1 score 20% high power view.

The symptoms usually shorter in duration than other posterior fossa tumors. Some clinical features that may help distinguish them from other CPA lesions: Vth, VIth, VIIth, VIIIth and lower cranial nerve involvement, and signs of cerebellar dysfunction are commonly note in CPA lesions. Hearing impairment and VIIth nerve involvement is usually less common and a late feature of CPA MB.

Only 5 of the 38 CPA MBs published so far presented with hearing impairment as initial symptom. Classic and desmoplastic MBs are the most common histological variants. There are no pathognomonic MRI criteria for differentiating them, and the only characteristic found is an iso- or hypointensity on T2/FLAIR sequence.⁸⁻¹⁵ MB has variable appearances on MRI in both children and adults.⁴

In most reported cases, CPA MBs have a homogeneous hypointensity on T1-, a heterogeneous hyperintensity on T2/FLAIR and enhance heterogeneously and strongly after contrast. In most cases classic MB had a high signal on T2 and a low signal on T1. In half cases of desmoplastic MB the lesion had an isointense signal on T2. In all variants the enhancement was in most cases intense and homogeneous.^{6,9} Very infrequently, MBs involving the CPA may present with widening and deep invasion

of the IAC, simulating an acoustic neuroma, or with a dural-based appearance, without bony involvement or hyperostosis, simulating a petrosal meningioma.^{12,13,17}

In our reported case the tumor had heterogenous hyperintensity on T2 WI and heterogenous enhancement on post contrast scan. Intra-axial tumors that may have an exophytic mass projecting into the CPA are astrocytoma and ependymoma. Occasionally, brain stem glioma, metastasis and lymphoma result in similar findings, but metastases are not likely children tumors.¹¹

CONCLUSION

Cerebellopontine angle is a very rare location of medulloblastoma, especially in children and mostly presents intra-axially. However, it is important to keep in mind the rare possibility of medulloblastoma presenting extra-axially. Its occurrence in the CP angle can confuse the nature of the lesion. Long term survival in these patients is very poor.

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