

# Extra-axial cerebello pontine angle medulloblastoma: A rare site of tumor

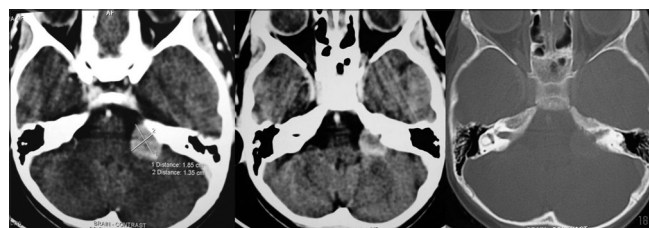
Sir,

Medulloblastoma is a common tumor of the posterior fossa, representing 20–25% of all pediatric neoplasms.<sup>[1]</sup> The tumor often occurs in the cerebellar vermis and at the apex of the fourth ventricle.<sup>[1,2]</sup> There are only a few reported cases of cerebellopontine (CP)-angle medulloblastoma in the literature, with most being intra-axial. The extra-axial site of this tumor remains a rarity.<sup>[1,3]</sup>

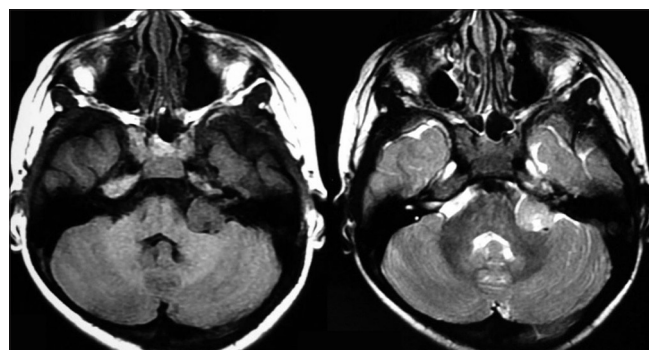
This 4-year-old girl presented with left hemicranial headache followed by facial asymmetry with deviation of angle of mouth for 1 month. There was no other significant history.

On clinical examination, higher intellectual functions were normal, both pupils were equal and reacting to light, visual acuity/visual fields were normal, fundus—no papilloedema, left lower motor neuron facial paresis, and left-sided sensory neural hearing loss, other cranial nerves normal. No stigmata of neurofibromatosis was noted. A computerised tomography (CT) scan of the brain showed contrast enhancing extra-axial lesion in the left CP angle centered around internal acoustic meatus [Figure 1]. CT bone window did not show enlargement of the internal acoustic meatus or hyperostosis [Figure 1]. Magnetic resonance imaging (MRI) of the brain showed CP angle lesion which was hypointense on T1W and hyperintense on T2W image [Figure 2]. The lesion was brilliantly enhancing with contrast, and no dural tail or canalicular component noticed [Figure 3]. She underwent left retromastoid craniectomy and total excision of the lesion. It was grayish, moderately vascular, and soft. There was a clear plane between the tumor and cerebellum, whereas it was adherent to dura and tent laterally. The HPE was confirmed as desmoplastic medulloblastoma [Figure 4] with the high MIB-1 labeling index and S-100 negativity.

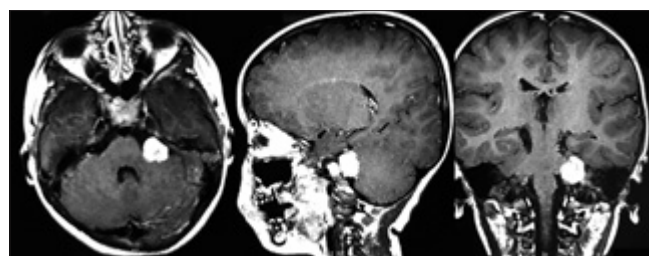
Medulloblastoma usually occurs in inferior medullary velum in the midline.<sup>[2]</sup> However rarely it may occur laterally in the cerebellar hemisphere in the pediatric and adult age group<sup>[1,3-5]</sup> with most being intra-axial. The extra-axial site of this tumor remains a rarity.<sup>[1,3]</sup> Origin of medulloblastoma may be either from germinal cells or their remnants situated at the end of the posterior medullary velum or from remnants of the external granular layer.<sup>[3,6]</sup> Their development in the CPA may be from the remnants of the external granular layer in the cerebellar hemisphere, including the flocculus which faces the CP



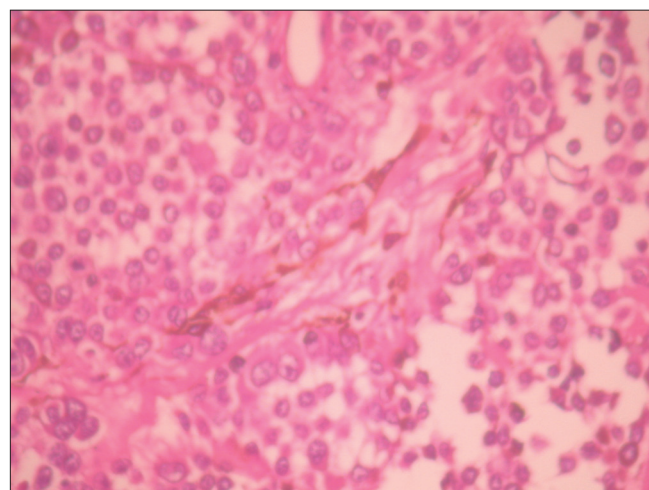
**Figure 1:** CT scan brain plain with contrast shows left extra-axial contrast enhancing lesion



**Figure 2:** MRI of the brain showed the CP angle lesion which was hypointense on T1W and hyperintense on T2W image



**Figure 3:** MRI of the brain contrast study axial, sagittal, and coronal section shows contrast enhancing extra-axial lesion



**Figure 4:** HPE suggestive of desmoplastic medulloblastoma

angle.<sup>[3,6]</sup> In the CP angle, medulloblastomas though fifth, sixth, and eighth cranial nerves are frequently involved, these nerves were spared in this patient.<sup>[5,7]</sup> CP angle medulloblastomas are very rare with nearly 36 cases published in the literature<sup>[1,3,5]</sup> of which only 10 are in adults.<sup>[1,3,5,7]</sup> The lack of association with any cerebellar tissue and the extra-axial location of the tumor made our patient's case quite rare. However, they are likely under-reported owing to publication bias and must be considered in the differential diagnosis of extra-axial CP angle lesions.

*Goutham Cugati, Manish Singh, Nigel Peter Symss,  
Anil Pande, Vasudevan Madabushi Chakravarthy,  
Ravi Ramamurthi*

*Department of Neurosurgery, Voluntary Health Services,  
Adyar, Chennai, Tamil Nadu, India*

**Address for correspondence:**

Dr. Manish Singh,  
Department of Neurosurgery  
Voluntary Health Services, Adyar,  
Chennai - 600113, Tamil Nadu, India  
E-mail: manish007singh@yahoo.com

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**Contact Persons**

Dr. Lalit Kumar, Organizing Secretary, Email: [lalitaiims@yahoo.com](mailto:lalitaiims@yahoo.com),  
Jose Pulickal, Email: [Jose.pulickal@ranbaxy.com](mailto:Jose.pulickal@ranbaxy.com)