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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.[1] The tumor often occurs in the cerebellar vermis and at the apex of the fourth ventricle.[1,2] 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.[1,3]

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.[2] However rarely it may occur laterally in the cerebellar hemisphere in the pediatric and adult age group[1,3-5] with most being intra-axial. The extra-axial site of this tumor remains a rarity.[1,3] 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.[3,6] 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

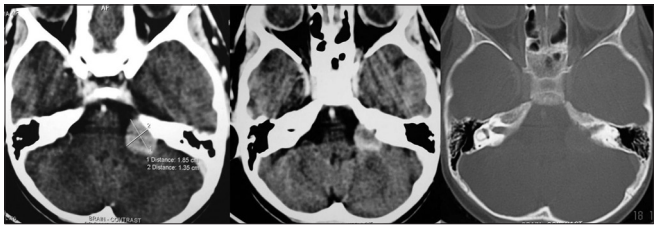

Figure 1: CT scan brain plain with contrast shows left extra-axial contrast enhancing lesion

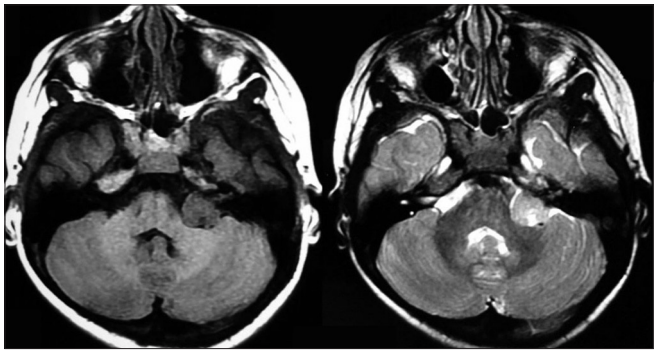

Figure 2: MRI of the brain showed the CP angle lesion which was hypointense on T1W and hyperintense on T2W image

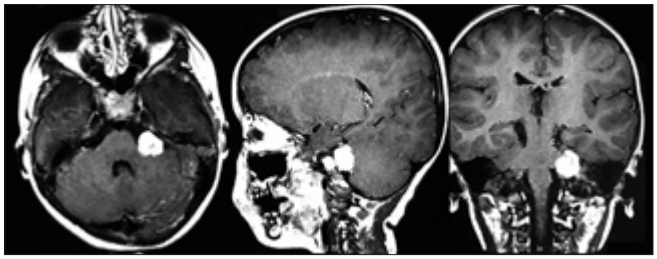

Figure 3: MRI of the brain contrast study axial, sagital, and coronal section shows contrast enhancing extra-axial lesion

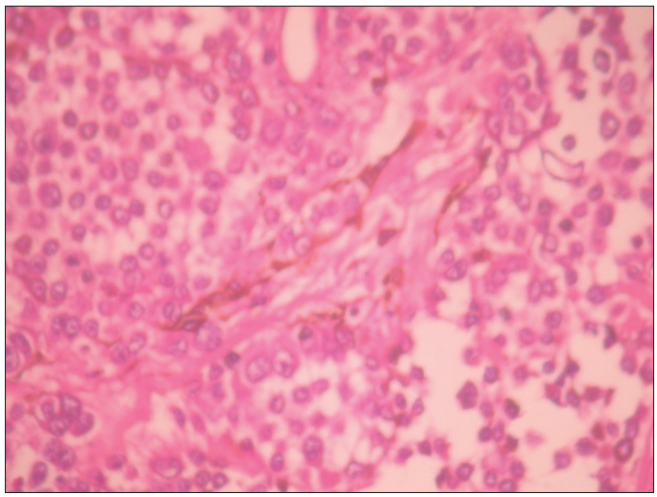

Figure 4: HPE suggestive of desmoplastic medulloblastoma

angle.[3,6] In the CP angle, medulloblastomas though fifth, sixth, and eighth cranial nerves are frequently involved, these nerves were spared in this patient.[5,7] CP angle medulloblastomas are very rare with nearly 36 cases published in the literature[1,3,5] of which only 10 are in adults.[1,3,5,7] 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.

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