Infantile embryonal tumour with mostly extra-axial extension into cerebellopontine angle: A case report

8-month-old female child with history of excessive crying and irritability.

Imaging Findings:

Mostly extra-axial T2 isointense (Fig. 1) solid mass lesion showing diffusion restriction (Fig. 2) and heterogeneous post contrast enhancement (Fig. 4 and 5) seen in left cerebellopontine angle. No evidence of blooming on GRE to suggest bleeding / calcification (Fig. 3). Mild perifocal oedema seen with mass effect over cerebellum and brainstem. The fourth ventricle is compressed with mild obstructive hydrocephalus. No evidence of extension of the mass lesion into internal auditory canal. The mass was resected with HPE and IHC revealing diagnosis of embryonal tumour NOS.

Discussion:

Embryonal tumours comprise a large subset of paediatric brain tumours and are associated with significant mortality and morbidity [1]. The most common embryonal tumour is medulloblastoma, which most often arises in the vermis and the fourth ventricle. As such cerebellopontine angle tumours are uncommon in children, a predominant extra-axial location of embryonal tumour at the cerebellopontine (CP) angle is very rare [2].

The 2016 World Health Organization classification of tumours of the central nervous system is a practical advance with the use of molecular parameters in establishing the diagnosis. The embryonal tumours other than medulloblastoma have also undergone substantial changes in their classification, with removal of the term primitive neuroectodermal tumour or PNET from the diagnostic lexicon [3].

Since medulloblastoma occurs most often in the midline cerebellum at the level of the fourth ventricle, children present frequently with symptoms and signs of obstructive hydrocephalus and cerebellar dysfunction. A CP angle embryonal tumour does not have specific clinical features and usually presents with features of mass effect over adjacent structures.

On imaging the embryonal tumours, as in other locations, are hyperdense on plain CT with heterogeneous post contrast enhancement. On MRI, the lesions are iso to hyperintense on T2, with restriction on diffusion-weighted imaging and heterogeneous enhancement. About 10 to 20% cases show calcification. There can be necrosis and cystic changes. MR spectroscopy shows elevated NAA and reduced choline with taurine peak at 3.4 ppm.
considered specific for medulloblastoma. Among all childhood brain tumours, embryonal tumours have the greatest
tendency for subarachnoid space seeding and extraneural spread, imaging with contrast of the whole neuraxis is
recommended.
Treatment consists of surgical excision with radiotherapy and chemotherapy. Prognosis depends on complete
surgical excision and the presence of subarachnoid seeding at the time of diagnosis.
**Differential Diagnosis List:** Cerebellopontine angle embryonal tumour NOS, Schwannoma, Meningioma

**Final Diagnosis:** Cerebellopontine angle embryonal tumour NOS

**References:**


medulloblastoma in the cerebellopontine angle: Report of a rare entity with review of literature. Journal of Pediatric
Neurosciences11(4): 331–334

David N. Louis, Arie Perry, Guido Reifenberger et al. (2016) The 2016 World Health Organization Classification of
Description: Axial T2W images show T2 isointense mostly extra-axial mass lesion in left CP angle with perifocal oedema and mass effect. Origin: Siloam Hospitals Lippo village
Description: DWI and ADC maps show restricted diffusion in left CP angle tumour. Origin: Siloam Hospitals Lippo village
Description: Axial GRE image show no evidence of blooming. Origin: Siloam Hospitals Lippo village
Description: Axial post-contrast T1 images show heterogeneous post-contrast enhancement. Origin: Siloam Hospitals Lippo Villge
Description: Coronal post-contrast T1 images show heterogeneous enhancement. Origin: Siloam Hospitals Lippo Village