IndSPN Case of the Month

July 2023

Clinical Presentation

A two-years-old male child born out of non-consanguineous marriage brought with complaints of -

- Altered behavior with excessive crying for six months
- Headache associated with vomiting for six months
- Undergone right medium pressure ventriculoperitoneal shunt elsewhere about 2 months ago

Clinical Examination

- Conscious and playful
- Developmental milestones- could not be reliably assessed
- Pupil- 2.5 mm bilateral reactive to light
- Rest of the cranial nerves could not be reliably assessed
- Nutrition- Adequate for age
- Tone and motor power- normal in all 4 limbs

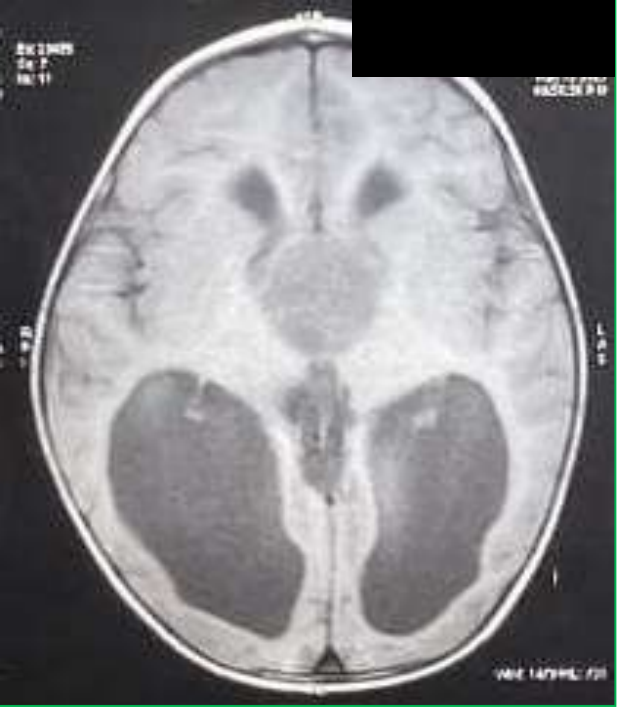
Clinical Examination

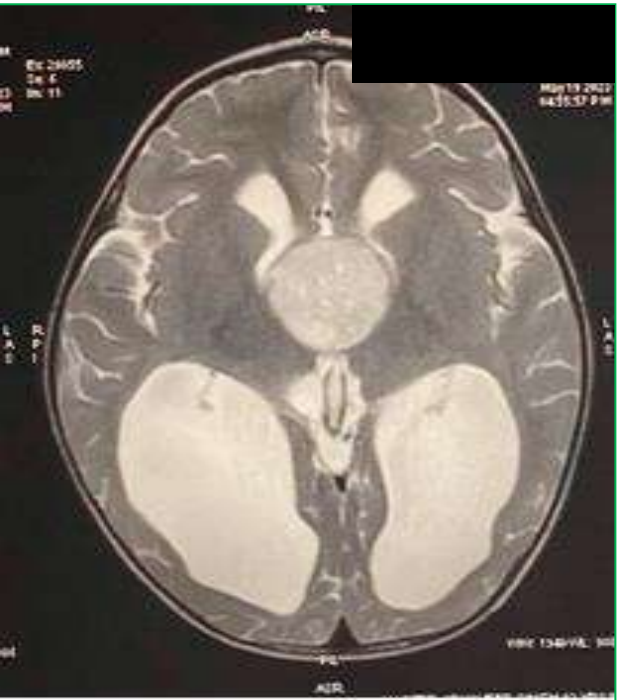
- Deep tendon reflexes- could not be reliably assessed
- Plantars- bilateral extensor
- Sensory- could not be reliably assessed
- Cerebellar/ Lobar signs- could not be reliably assessed
- Meningeal/ Neurocutaneous signs- absent

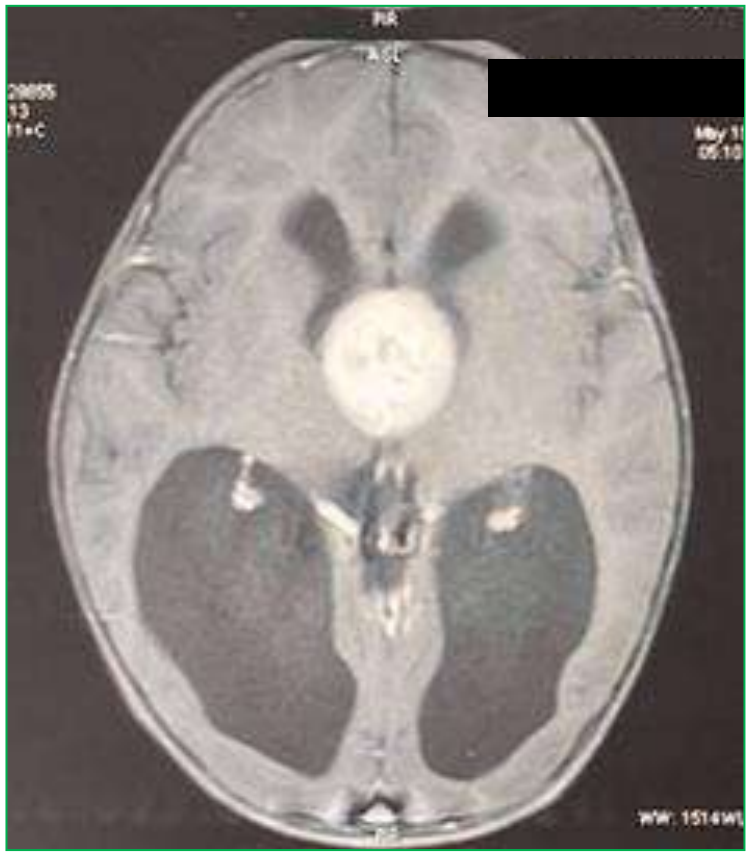
Provisional Diagnosis with Clinical Localization

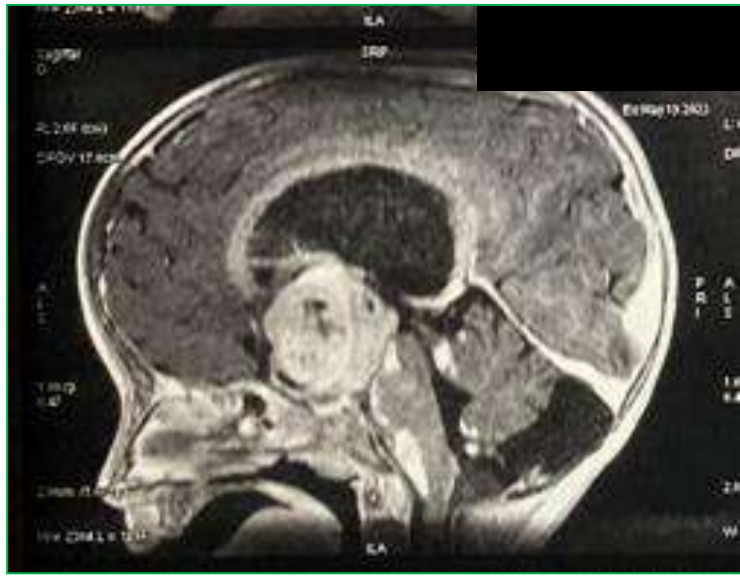
?

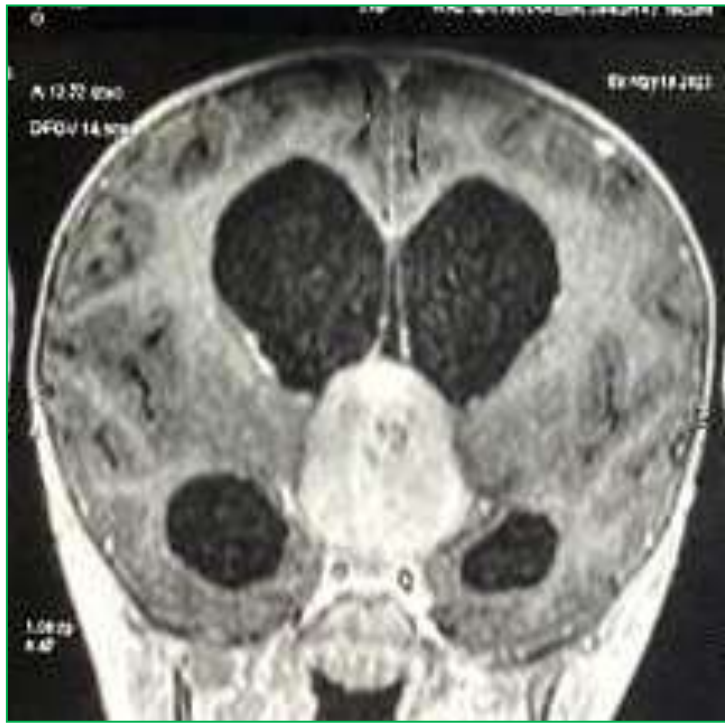
Radiology











➢*MRI Brain (Plain + Contrast)*

- There is evidence of a single ill-defined lesion (4.2 x 3.4 x 4.1 cm) epicentered in suprasellar region and floor of the third ventricle. It is causing dilatation of the 3rd ventricle and obliteration of foramen of Monroe. It is T1 hypointense, T2 heterogeneously hyperintense with intense contrast enhancement with few non-enhancing cystic areas. There is evidence of ill-defined sheath like thickening with nodular meningeal enhancement along basal cisterns, bilateral sylvian fissures, pre-pontine cisterns and bilateral cerebellar hemispheres – likely leptomeningeal seeding

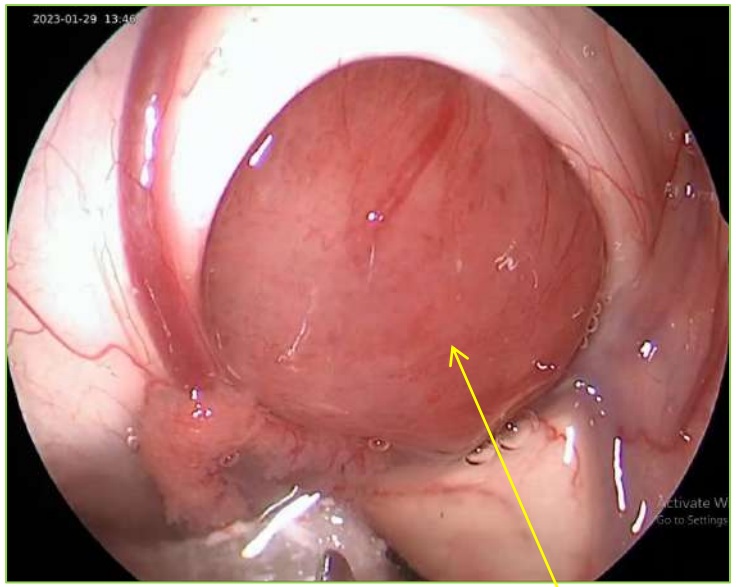
Radiological Impression

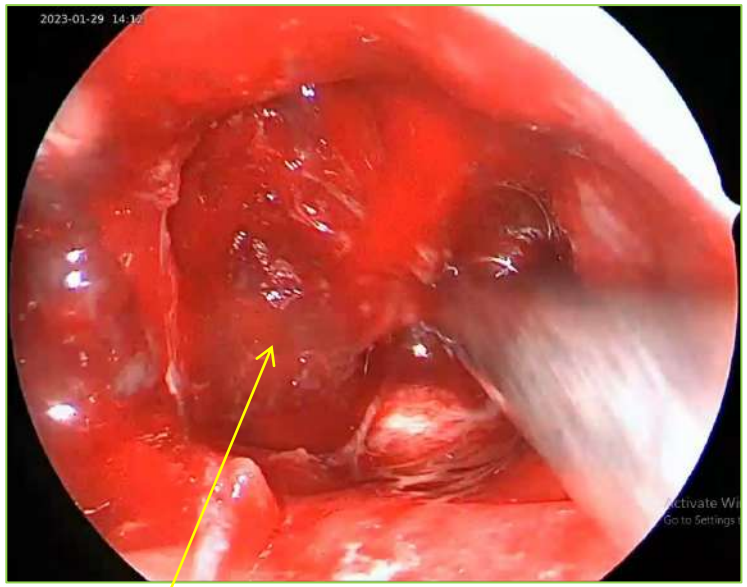
- **Differential Diagnosis**

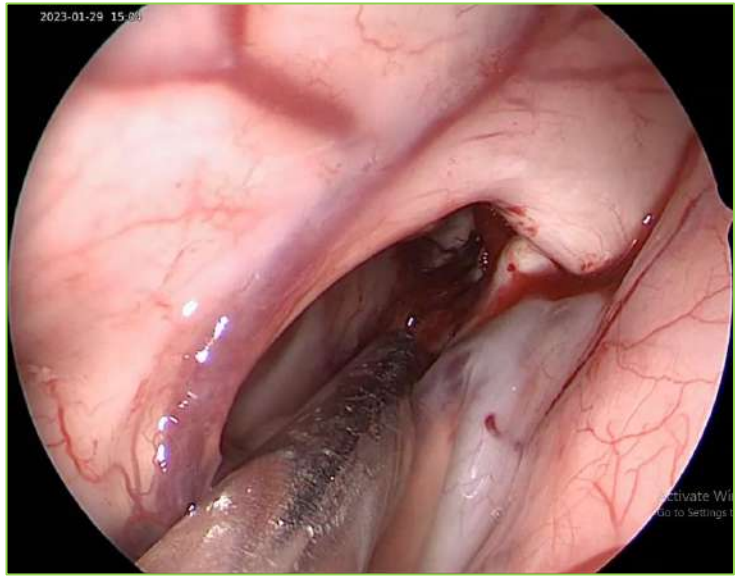
Surgery

- **Position-** Supine position
- **Incision-** Curvilinear incision of 4 cm size given in mid-pupillary line just in front of coronal suture on the right side
- **Procedure –** Right frontal pre-coronal key-hole craniotomy and endoscope assisted gross total tumor excision

Intra-operative Images







At Foramen of Monro

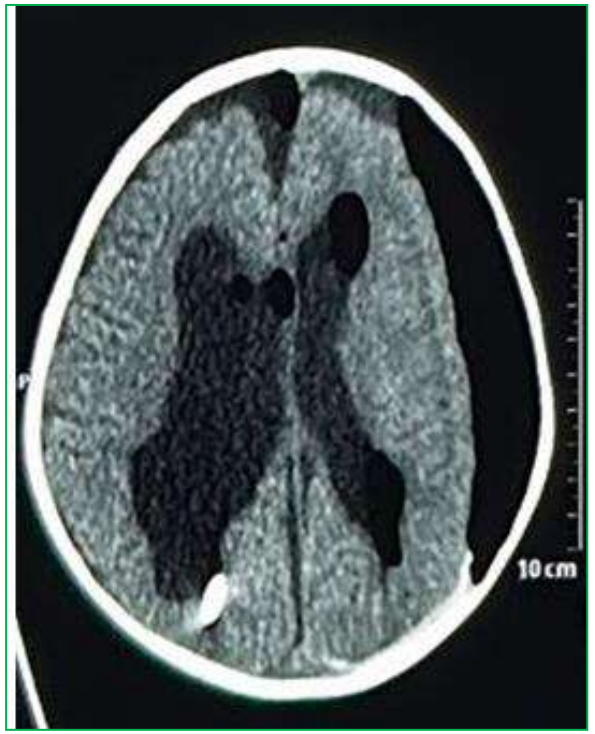
Third ventricle

Tumor visualised

Ventricle seen after tumor excision

Post-operative status

- Post-operative course was uneventful
- Wound healthy
- Conscious and playful
- No new neurological deficits
- HPE report- Ependymoma (WHO grade II)



Post-op imaging

Case Summary

A two-years-old male child brought by parents with chief complaints of altered behavior, excessive crying, headache associated with vomiting for six months. Underwent CSF diversion about two months ago. At present on examination, the child is conscious and playful. Radiological evaluation was suggestive of third ventricle tumor..? Germ cell tumor. Underwent endoscopic assisted tumor excision. Intra-operative impression- Germ cell tumor. But biopsy turned out to be ependymoma (WHO grade II)

Relevant Literature

- Ependymomas are the third most common primary third ventricular tumor found in children
- Constitute about 8- 15% of the neoplasms
- Arise from the ependyma and are also seen with neurofibromatosis type II
- Composed of cells with regular, round to oval nuclei
- It is a neuroepithelial glial cell tumor

- Up to 30% of intracranial ependymomas arise from the lateral or third ventricles

- Bimodal peaks of occurrence- five years and 35 years of age
- Clinical presentation
  - Severe headache, visual loss (secondary to papilledema), vomiting, drowsiness, gait change (rotation of feet when walking), constipation, back pain, and abnormal flexibility of the back
- MRI
  - Imaging modality of choice
  - T1- isointense to hypointense, T2 hyperintense , blooming on SWI
  - Heterogeneous enhancement
- Surgical resection is the first line of treatment

- Adjuvant radiotherapy can be beneficial, but the ideal volume of irradiation remains controversial
- Use of chemotherapy remains uncertain
- Prognosis for Pediatric ependymomas remains relatively poor as compared to other brain tumors
- Intraventricular ependymomas remain a surgical challenge due to their high rate of incomplete tumor resection and permanent neurological complications linked to their removal
- Incomplete tumor removal accompanied by radiotherapy helps in long-term progression-free survival in a few cases

Suggested Readings

- Ahmed SI, Javed G, Laghari AA, Bareeqa SB, Aziz K, Khan M, Samar SS, Humera RA, Khan AR, Farooqui MO, Shahbaz A. Third Ventricular Tumors: A Comprehensive Literature Review. Cureus. 2018 Oct 5;10(10):e3417. doi: 10.7759/cureus.3417. PMID: 30542631; PMCID: PMC6284874

- A systematic review of treatment outcomes in pediatric patients with intracranial ependymomas: A review. Cage TA, Clark AJ, Aranda D, et al. *J Neurosurg Pediatr.* 2013;11:673–681

- Surgical treatment of intraventricular ependymomas and subependymomas. Nowak A, Marchel A. *Neurol Neurochir Pol.* 2012;46:333–343