IndSPN Case of the Month

January 2024

Clinical Presentation

A 1.5 years-old male child brought by his parents presented with chief complaints of -

- Seizure for 1 year
- Irritability for 3 months
- Altered sensorium for 1 week

Clinical Examination

- Spontaneous eye opening
- Pupil – 2.5 mm bilateral reactive to light
- Rest of the cranial nerves – could not be assessed reliably
- Nutrition – Adequate
- Tone – normal in all 4 limbs
- Power – moving all 4 limbs spontaneously

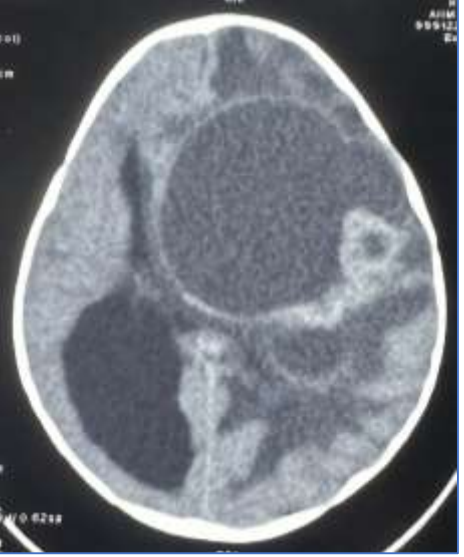
Clinical Examination

- Plantars- B/L flexor
- Sensory- could not be assessed reliably
- Cerebellar/Lobar signs- could not be assessed reliably
- Meningeal/Neurocutaneous signs- absent

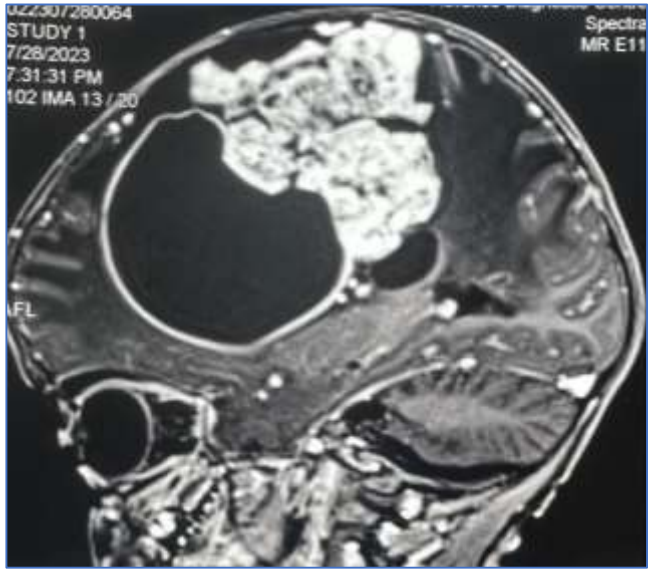
Provisional Diagnosis with Clinical Localization

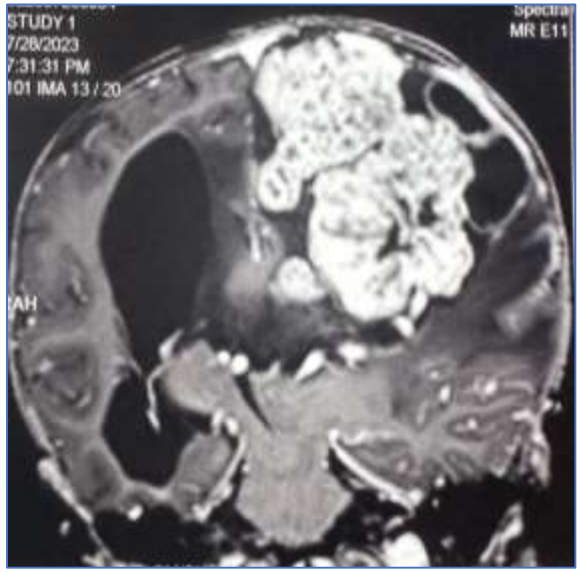
?

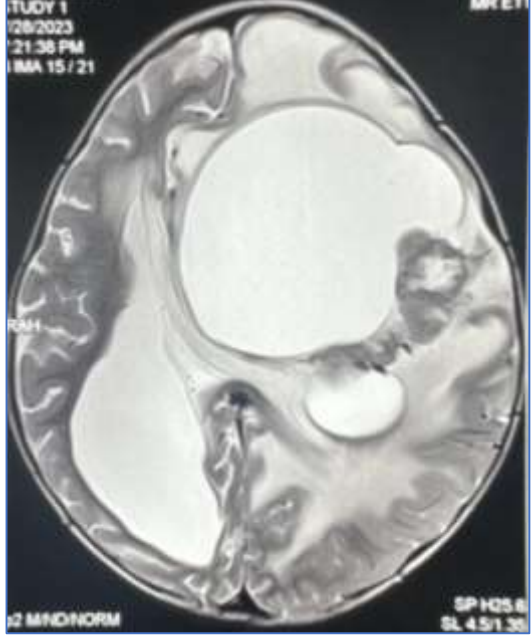
Radiology

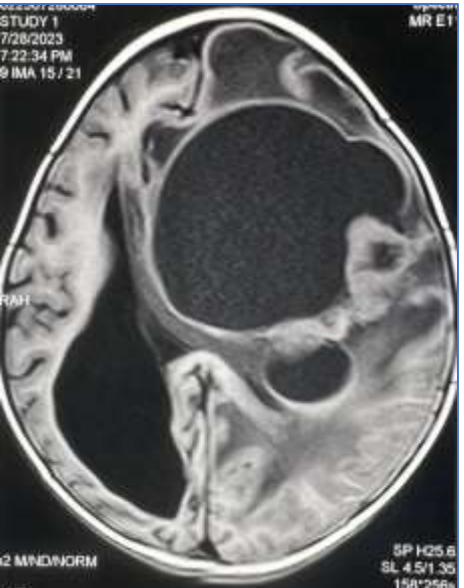


NCCT Head

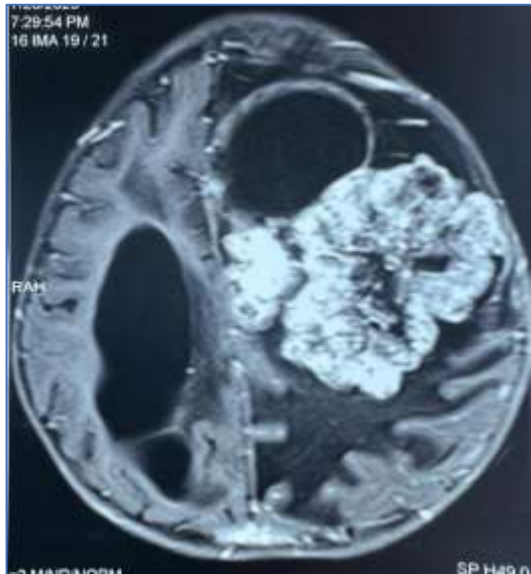








MRI Brain



➢*MRI Brain (Plain + Contrast)*

- There is evidence of a large well defined solid cystic intra-axial space-occupying lesion (100 x 92 x 76mm) involving left frontal lobe. Lesion is T2 heterogeneously hyperintense, T1 hypointense with heterogeneously enhancing solid component and peripherally enhancing cystic component. Moderate perifocal edema is observed extending to adjacent white matter and ipsilateral basal ganglia-capsular region. Significant pressure effect is noted in form of compressed adjacent sulcal-cisternal spaces, compressed-deformed left lateral ventricle with a midline shift of 2.2 cm towards right side. Left sided uncal herniation present compressing over midbrain.

Radiological Impression

- **Differential Diagnosis**

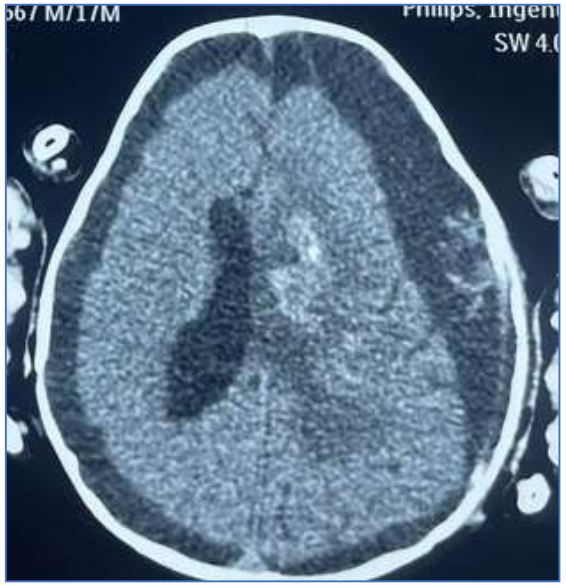
?

Surgery

- **Position –** Supine position
- **Incision –** Horse-shoe incision with anterior limb 2 cm in front of coronal suture and posterior limb 5 cm behind the coronal suture
- **Procedure –** Left frontoparietal craniotomy and near total tumor excision

Post-operative status

- Post-operative course was uneventful
- Conscious, playful
- Sutures removed on post-operative day 7
- Power– moving all 4 limbs spontaneously
- HPE– Desmoplastic Infantile Ganglioglioma



Post-op imaging

Case Summary

A 1.5 years-old male child presented with chief complaints of seizure for 1 year, irritability for 3 months, altered sensorium for 1 week. On examination spontaneous eye opening with spontaneous movement in all 4 limbs was present. Preoperative radiological impression- Infantile Meningioma. During surgery the lesion was noticed to be firm and mildly vascular with a large tumor cyst having xanthochromic fluid within. Biopsy was suggestive of Desmoplastic Infantile Ganglioglioma

Relevant Literature

- Supratentorial brain tumor occurring before 2 years
- WHO grade I tumors, Contributing about 1% of all brain tumors
- Male preponderance present
- Mostly hemispheric in location and without side prevalence

- Multiple lobes are commonly involved with a higher incidence for the frontal and parietal regions. Less frequent in suprasellar region, the posterior cranial fossa, or the spinal canal
- Classical symptoms include rapid head growth (60 % of cases), often associated to a bulging of the anterior fontanel, sunset sign
- These tumors are usually very large at diagnosis with uni-or multiloculated cysts filled with clear or xanthochromic fluid

- Superficial portion is primarily extra-cerebral, involving the leptomeninges and superficial cortex, and is commonly attached to the dura

- CT
  - Solid part appears as isodense or slightly hyperdense with strong contrast enhancement, cystic component is most frequently hypodense or isodense according to the relatively variable protein content; calcifications or hemorrhages are usually absent
- MRI
  - T1 and T2 isointense with strong contrast enhancement of solid part
- *Surgical resection is the first line of treatment*

- When partial resection is achieved, a careful follow-up is mandatory to monitor potential tumor re-growths
- Long relapse-free intervals have been described even after partial resections, suggesting the potential for tumor residual stabilization after a partial tumor removal
- If recurrence or residual tumor growth is seen, if surgery is not an option, chemotherapy might be considered
- Radiotherapy is considered only in patients, aged more than five years, as a last resort in recurring tumors after surgery and chemotherapy

Suggested Readings

- Bianchi F, Tamburrini G, Massimi L, Caldarelli M. Supratentorial tumors typical of the infantile age: desmoplastic infantile ganglioglioma (DIG) and astrocytoma (DIA). A review. Childs Nerv Syst. 2016 Oct;32(10):1833-8. doi: 10.1007/s00381-016-3149-4. Epub 2016 Sep 20. PMID: 27659826.
- Kamoun S, Azouz H, Zemmali M, Haouet S, Kchir N. Desmoplastic infantile ganglioglioma. Pan Afr Med J. 2019 Mar 11;32:113. doi: 10.11604/pamj.2019.32.113.12669. PMID: 31223403; PMCID: PMC6560944.
- Iwami K. Desmoplastic infantile ganglioglioma. Childs Nerv Syst. 2007 Jun;23(6):619-20; author reply 621. doi: 10.1007/s00381-007-0352-3. Epub 2007 Apr 21. PMID: 17450367.