IndSPN Case of the Month

December 2022

Clinical Presentation

A three years-old boy born out of non-consanguineous marriage brought by parents with complaints of-

- Seizures since 6 months of age
- Undergone herniotomy for right congenital inguinal hernia at the age of five months

Clinical Examination

- Child was alert and playful
- Developmental milestones: Appropriate but limited interaction with peer groups
- Polydactyly in bilateral upper limbs
- Body weight: 16 Kg, Height: 90 cms
- No neurocutaneous markers noted
- Healed and healthy scar over the right groin area

Provisional Diagnosis with Clinical Localization

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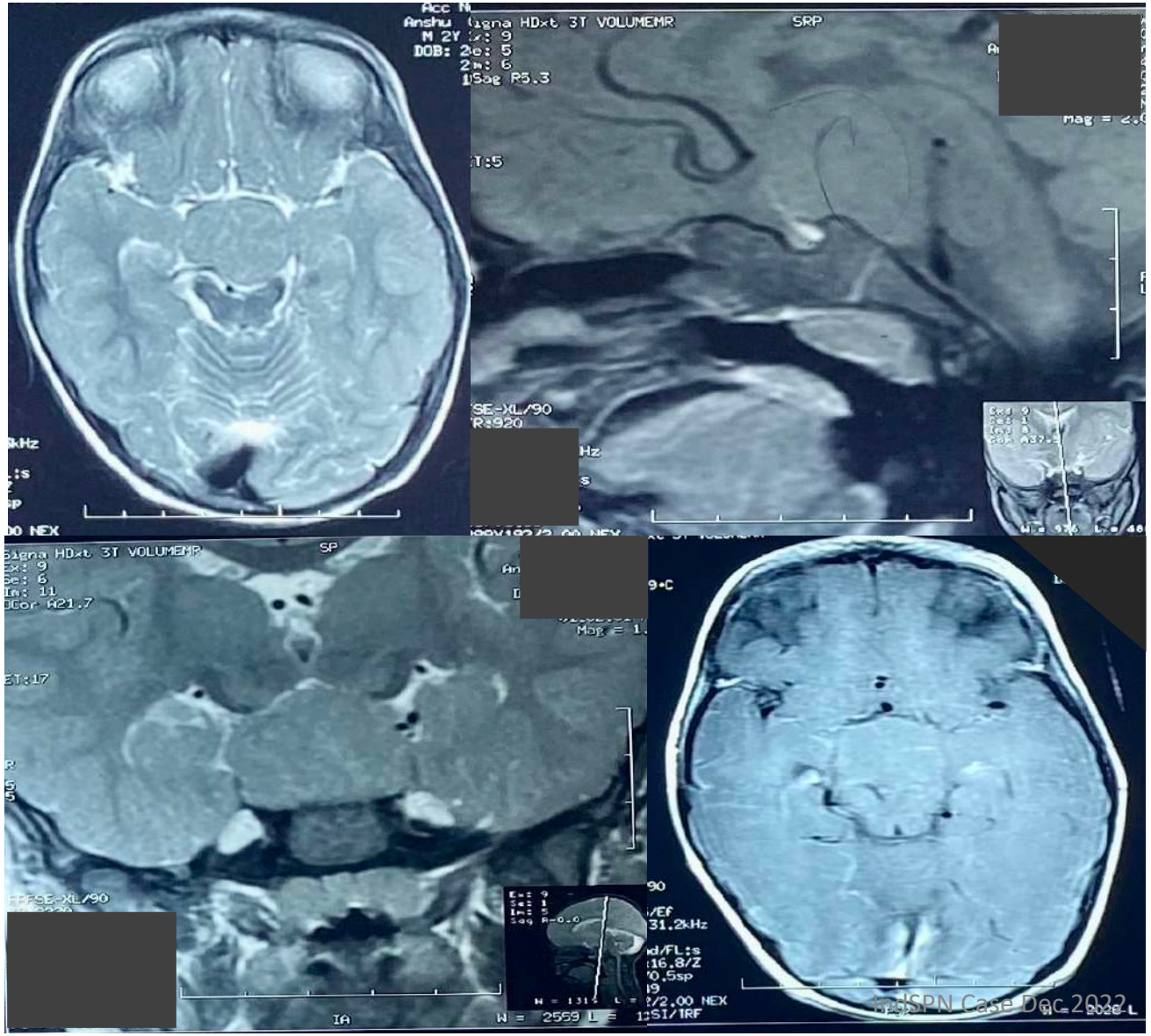
Radiology



MRI Brain
Plain + Contrast

➢MRI Brain (Plain + Contrast)

- The lesion appeared mixed intensity (predominantly hypo- on T1W and T2W sequences) centered in the suprasellar region posterosuperior to pituitary gland
- The lesion did not show contrast enhancement or any diffusion restriction

Radiological Impression

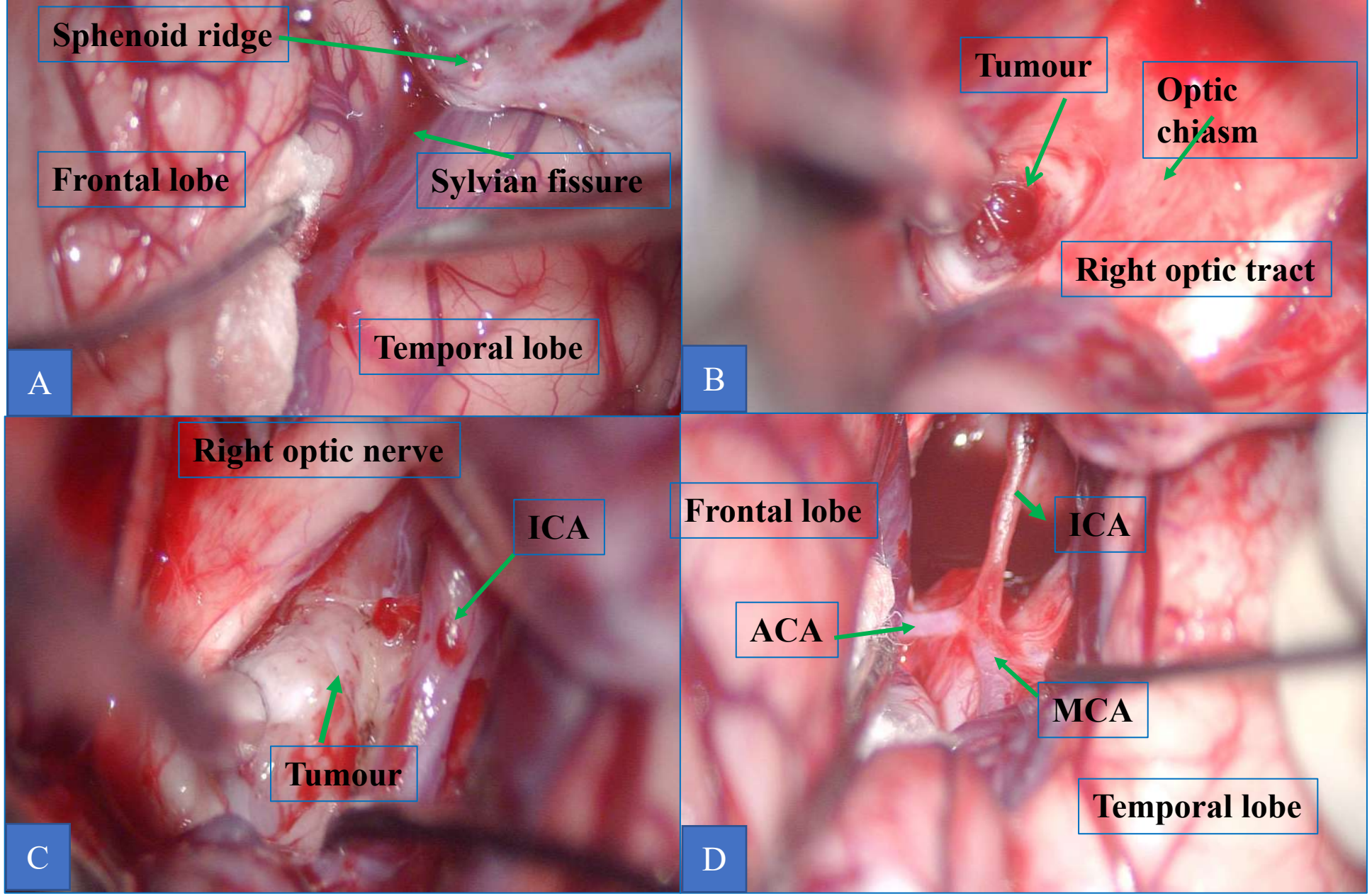
- **Differential Diagnosis**

?

Surgery

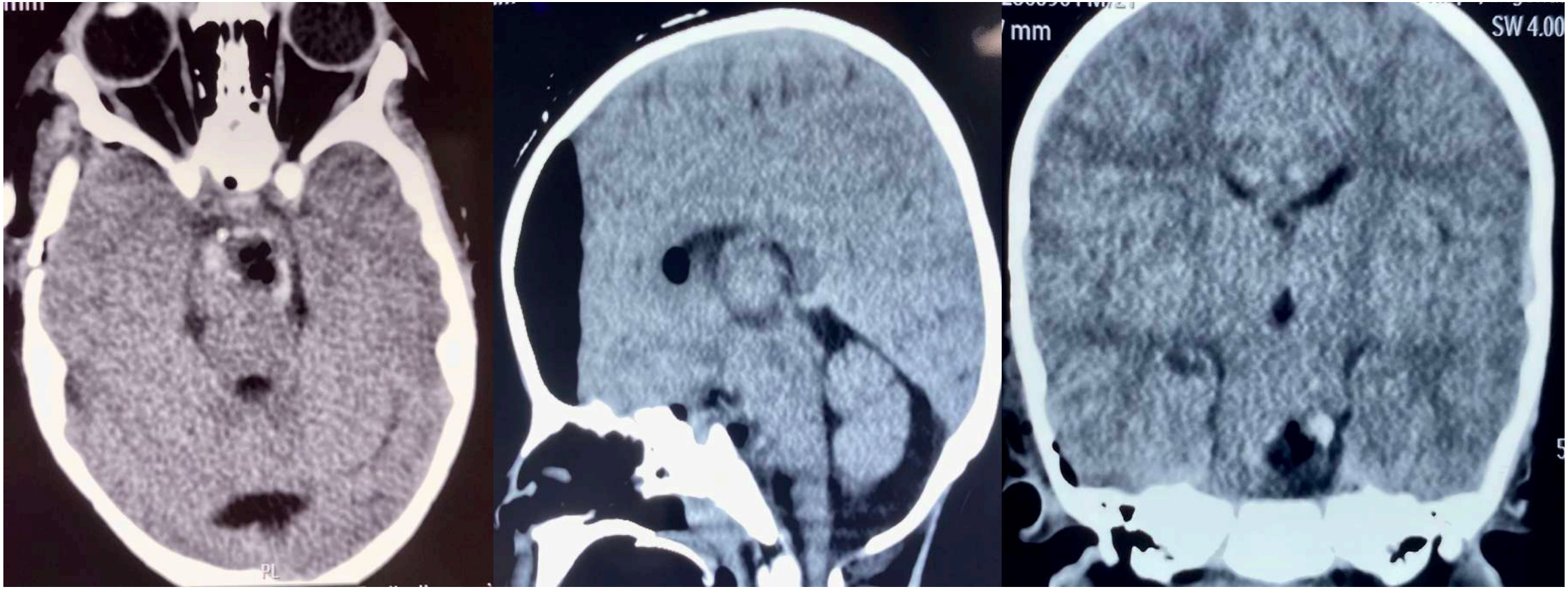
- **Position-** supine position with head turned 30 degrees to left side; rigid fixation on Mayfield head frame
- **Craniotomy-** Right fronto-temporal craniotomy
- **Approach-** Trans-sylvian approach

Intraoperative Images



A– Trans sylvian approach, Surgical corridors for tumor decompression (B & C); B– Inter-optic and trans lamina terminalis C– optico-carotid, D– Post tumor decompression

Post- operative Images



HPE report: Hypothalamic Hamartoma

Case Summary

- This 3-year-old boy was brought with complaints of seizure since six months of age. Non localizing seizure semiology was noticed with polydactyly in both the hands. Radiological work up was suggestive of suprasellar lesion with mixed intensity posterior to the pituitary without enhancement on contrast
- Right Pterional craniotomy with trans sylvian approach was performed and sub-total tumor decompression was achieved using optico-carotid, inter optic and trans lamina terminalis corridors. Intraoperatively the pituitary was identified separately from the lesion. Histopathology report turned out to be Hypothalamic hamartoma
- The child underwent tracheostomy and was discharged in two weeks

Relevant Literature

- Hypothalamic hamartomas (HH) are rare, basilar developmental lesions with widespread comorbidities often associated with refractory epilepsy and encephalopathy
- HHs develop in a region rostral to the mammillary body. The lateral tuberal nucleus (LTN), a cell group in this region is hypothesized to be the origin of HH
- Imaging allows for early detection of HH with high-resolution fetal MRI. Since the lumen of the third ventricle is not fully established until 26–28 weeks gestational age, so it is difficult to diagnose HH until the second trimester. A common misdiagnosis at this time are inter-hypothalamic adhesions, which are horizontally oriented bands that connect the medial walls of the third ventricle

- Genetic studies suggest mutations in *GLI3* and other patterning genes are involved in HH pathogenesis
- The Delalande classification system groups HH into 4 types based on location and is in turn related to surgical trajectory/approaches and surgical morbidities
- Endoscopic resection/disconnection can be performed by means of bipolar, radiofrequency, or laser coagulation and achieves a high level of seizure control (up to 78% Engel I + II) but can have an overall complication rate up to 8.3%
- Gamma Knife radiosurgery (GKS) has good long-term safety/efficacy data, but requires a substantial time (up to 2 years) before positive effects are seen, and is not recommended for larger HH

Suggested Readings

- Alberto JR. Leal, José P. Monteiro, Mário Forjaz Secca, Constança Jordão. Functional brain mapping of ictal activity in gelastic epilepsy associated with hypothalamic hamartoma: A case report. Epilepsia. 2009;50:1624–31.
- Ng YT, Rekate HL, Prenger EC, Wang NC, Chung SS, Feiz-Erfan I et al. Endoscopic resection of hypothalamic hamartomas for refractory symptomatic epilepsy. Neurology. 2008;70:1543-8.
- Delalande O, Fohlen M. Disconnecting surgical treatment of hypothalamic hamartoma in children and adults with refractory epilepsy and proposal of a new classification. Neurol Med Chir. 2003;43:61-8.