IndSPN Case of the Month

June 2023

Clinical Presentation

A 2 years-old male child brought by his parents presented with chief complaints of -

- Multiple episodes of seizure x 2 months
- Reduced speech output x 1 month
- Altered sensorium x 2 days

Clinical Examination

- Drowsy but arousable
- 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

Clinical Examination

- Tone and motor power was grossly normal
- Deep tendon reflexes - could not be reliably assessed
- Plantars - B/L extensor
- Sensory and Cerebellar/Lobar signs - could not be reliably assessed
- Meningeal/Neurocutaneous signs - absent

Provisional Diagnosis with Clinical Localization

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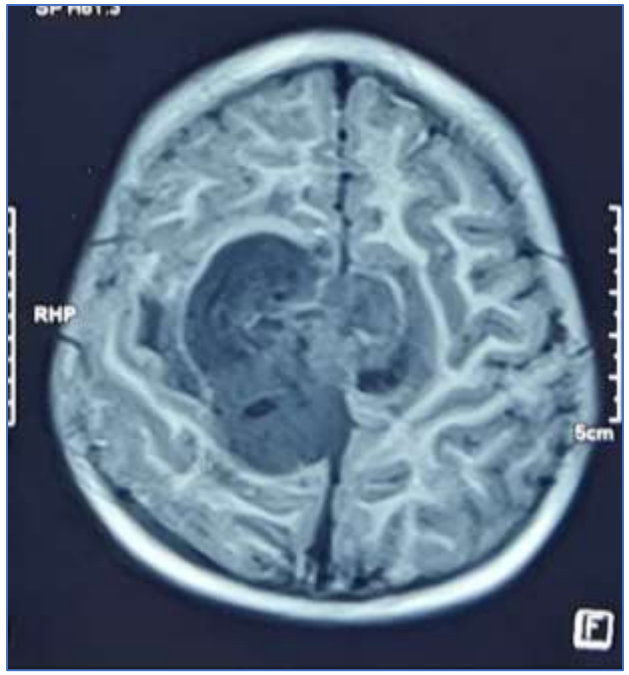
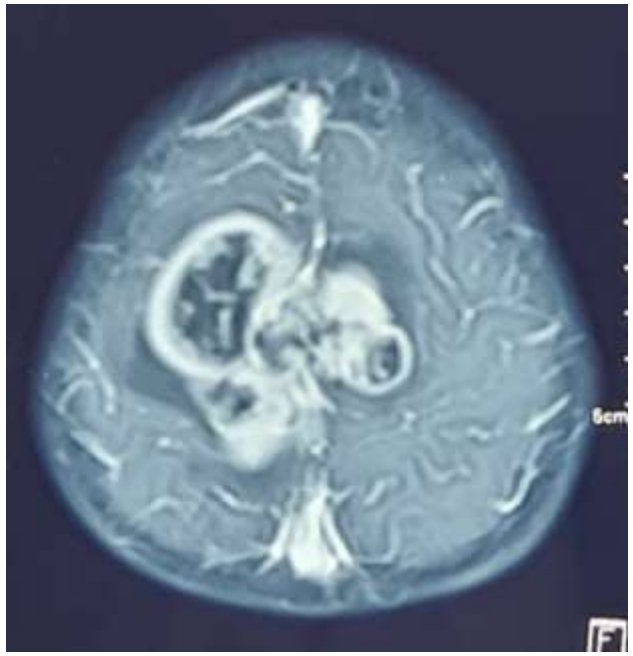
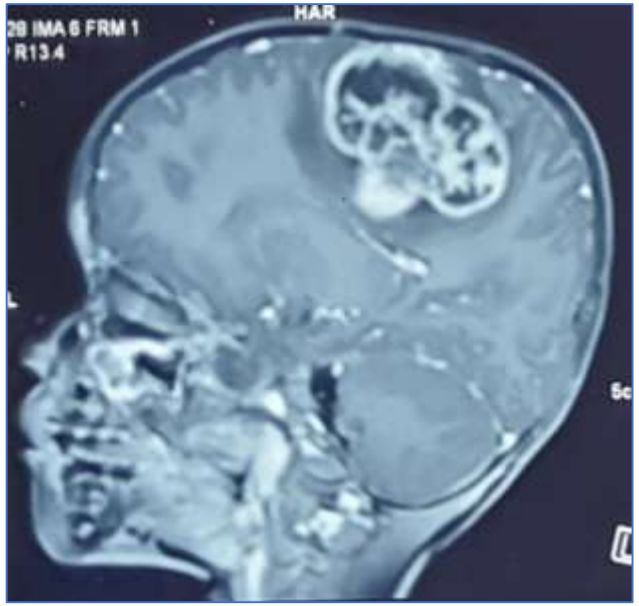
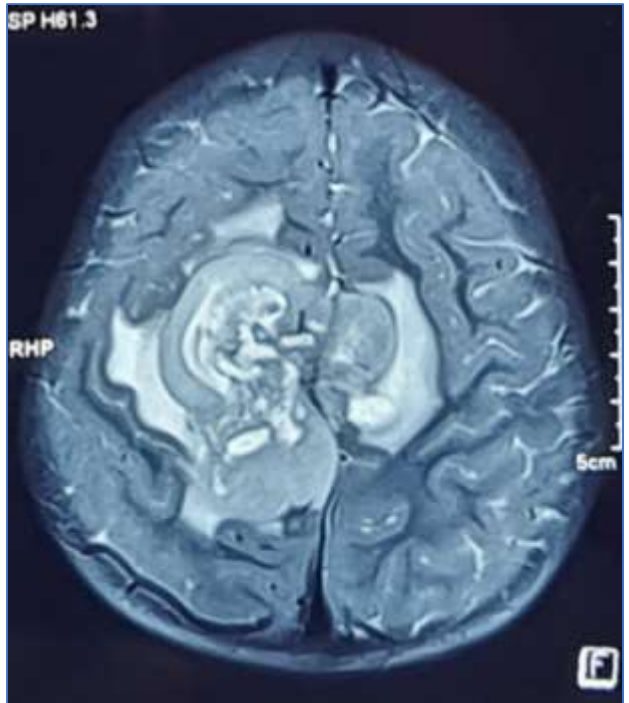
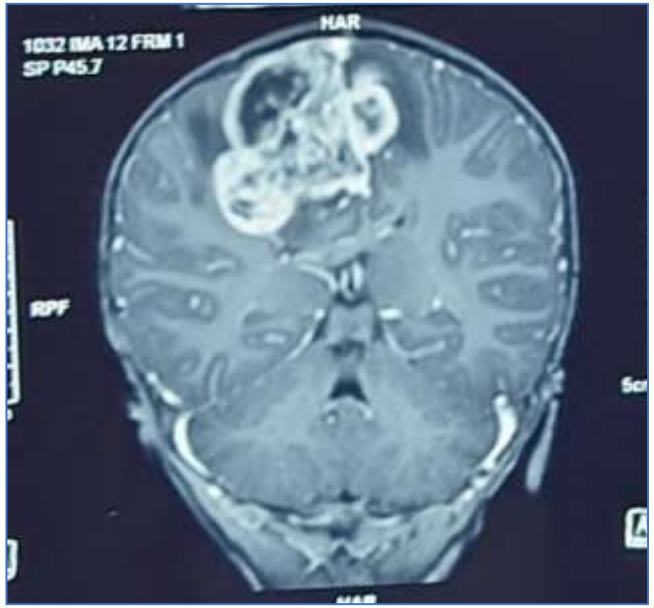
Radiology











➢ *MRI Brain (Plain + Contrast)*

- There is evidence of a single well-defined lesion (5.7 x 5.6 x 4.0 cm) epicentered predominantly in right frontoparietal region with extension across the midline to opposite side. It is extending anteriorly till prefrontal cortex on the right side and till premotor cortex on left side and posteriorly till superior parietal lobule. It is T1 predominantly hypointense with a few specks of hyperintensity, T2 heterogenously hyperintense with irregular peripheral contrast enhancement with central necrotic areas. There is presence of significant perilesional edema
- On DWI, diffusion restriction present. On SWI blooming seen. The lesion is abutting the superior sagittal sinus with effacement of right lateral ventricle. On MRS, Choline : NAA ratio is significantly increased

Radiological Impression

- **Differential Diagnosis**

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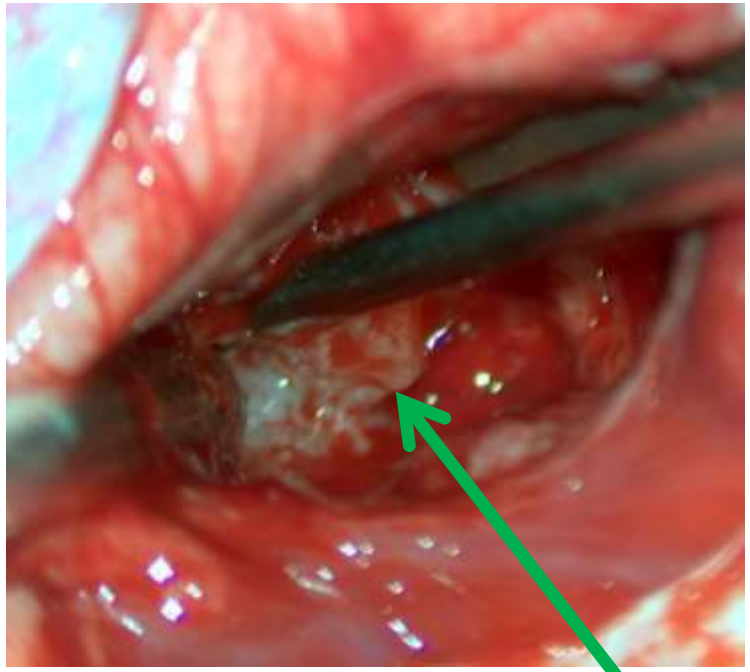
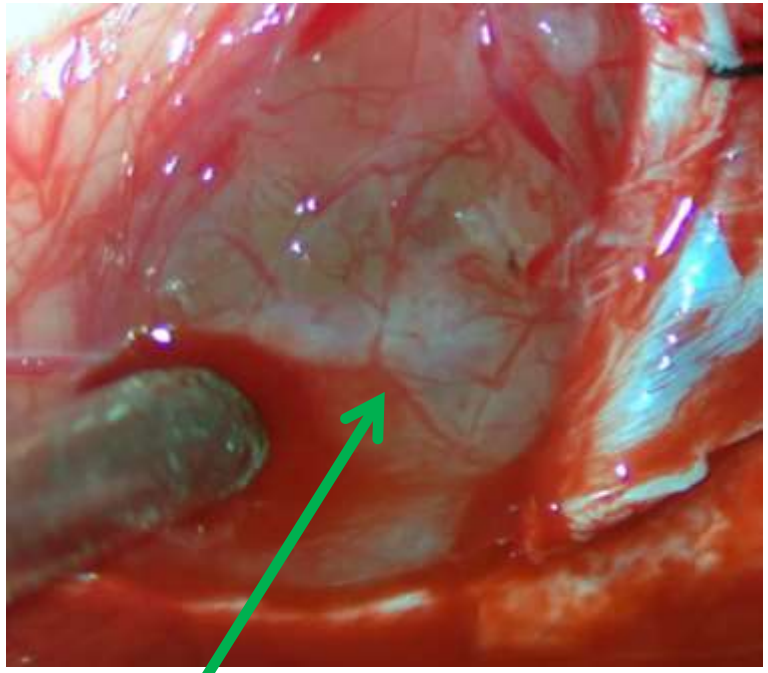
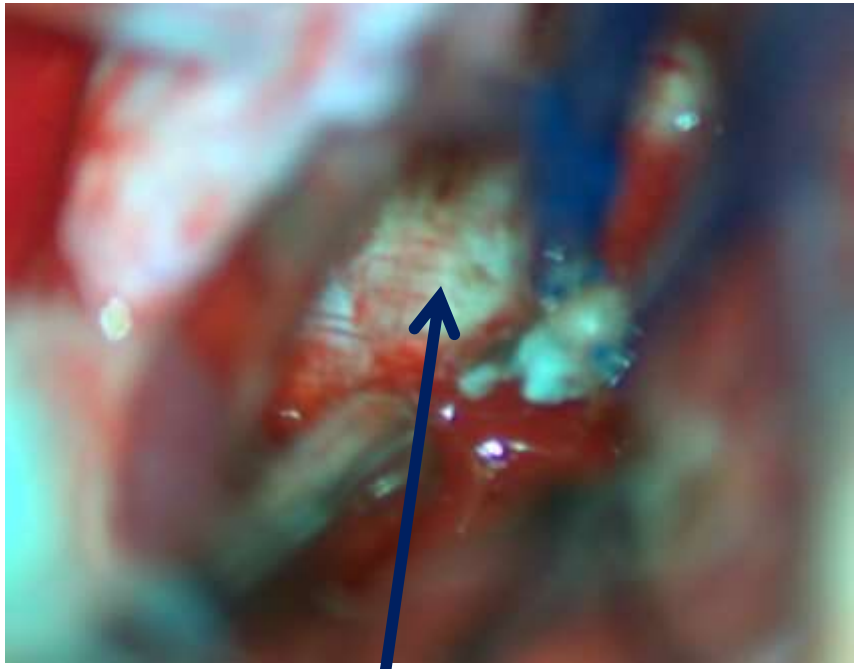
Surgery

- **Position –** Prone position on Mayfield head frame
- **Incision** – Horizontal incision given extending from one parietal eminence to another
- **Procedure –** Bi parietal craniotomy and near total tumor excision

Intra operative Images





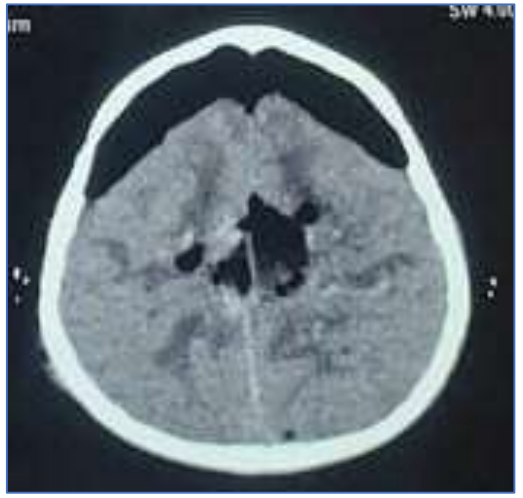


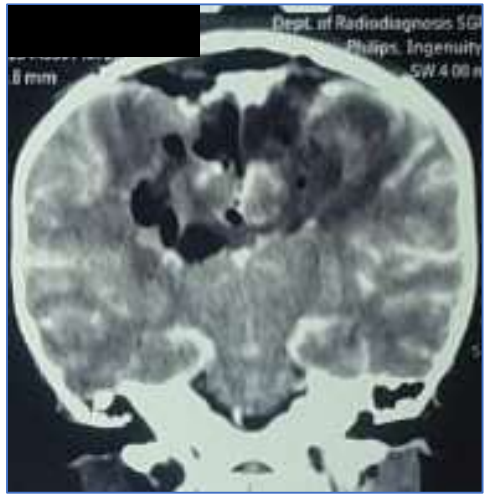
Tumor visualised

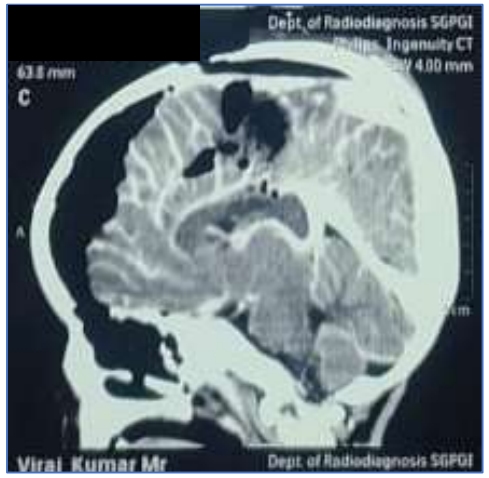
Tumor attached to falx cerebri

Post-operative status

- Post-operative course was uneventful
- Operative wound was healthy
- Conscious, irritable with no post operative deficits
- **Histopathology Report-** Atypical Teratoid Rhabdoid tumor







Post-op imaging

Case Summary

A 2 years-old male child brought by parents with chief complaints of seizures, reduced speech output and altered sensorium. On examination patient was drowsy but arousable

- Radiological work up was suggestive of fronto parietal high-grade lesion
- Intra-operative impression – Atypical Teratoid / Rhabdoid Tumor
- Biopsy – Atypical Teratoid Rhabdoid Tumor

Relevant Literature

- Atypical Teratoid / Rhabdoid Tumor (AT/RT) constitutes one of three major CNS embryonal tumors
- WHO Grade IV tumor
- Predominantly observed in children younger than 3 years
- Reported in all CNS locations, including the cerebellopontine angle cistern, meninges, cranial nerves, spinal canal, and extra dural location

- **Clinical presentation**
  - Symptoms of raised intracranial tension, lethargy, failure to thrive, regression of developmental milestones, irritability, and macrocephaly in very young children
  - Involvement of cerebellar hemisphere may lead to symptoms of ataxia, head tilt, and nystagmus
  - Cranial nerve palsies (cranial nerves VII and VIII) [Cerebellopontine angle]

- **Investigations**

➢Computed tomography of brain demonstrates hyperdense lesion attributable to the high cellularity of the tumor and heterogeneous enhancement on postcontrast images. Calcification may be seen in up to 40% of tumors

➢MRI is the imaging modality of choice. Heterogeneous iso-intense signal on both T1- and T2-weighted MR images. Variable enhancement (heterogeneous, peripheral nodular, intense, and mild). Peripherally located cystic components are commonly demonstrated on MR images and are a useful distinguishing feature.

- **Management**

➢Management includes surgical resection followed by adjuvant chemoradiation

➢Survival in patients with AT/RT has been dismal, with a reported median survival of ~1 year

Suggested Readings

- Biswas A, Kashyap L, Kakkar A, Sarkar C, Julka PK. Atypical teratoid/rhabdoid tumors: challenges and search for solutions. *Cancer Manag Res*. 2016;8:115-125. Published 2016 Sep 16. doi:10.2147/CMAR.S83472
- Calandrelli R, Massimi L, Pilato F, et al. Atypical Teratoid Rhabdoid Tumor: Proposal of a Diagnostic Pathway Based on Clinical Features and Neuroimaging Findings. *Diagnostics (Basel)*. 2023;13(3):475. Published 2023 Jan 28. doi:10.3390/diagnostics13030475
- Major K, Daggubati LC, Mau C, Zacharia B, Glantz M, Pu C. Sellar Atypical Teratoid/Rhabdoid Tumors (AT/RT): A Systematic Review and Case Illustration. *Cureus*. 2022;14(7):e26838. Published 2022 Jul 14. doi:10.7759/cureus.26838