KREP

2025

KARNATAKA RADIOLOGY EDUCATION PROGRAM

CASE PRESENTATION

Case of ***ATRT vs ETMR vs choroid plexus carcinoma***

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CLINICAL HISTORY OF THE PATIENT

AGE: 5 month

SEX: Male

Brought to the paediatric OPD with complaints of:

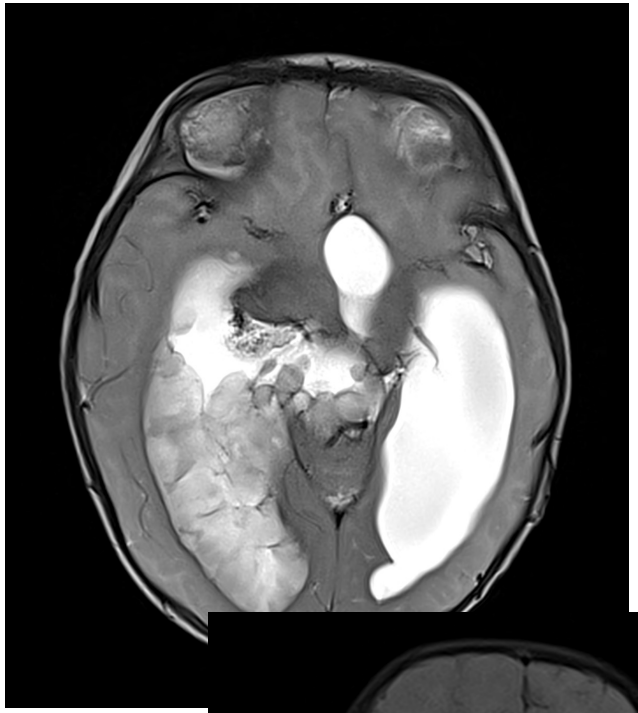
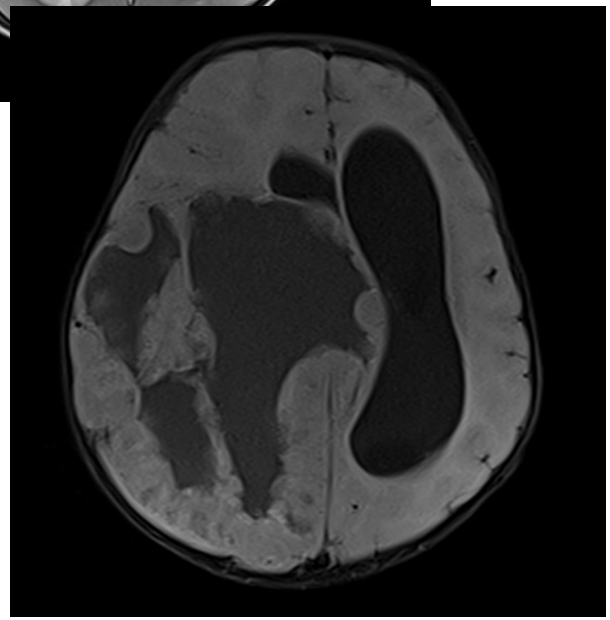
- Vomiting x 8days
- Low grade fever & cold x 20 days (not relieving on medication)

Blood investigations performed : Normal

Anthropometry- head circumference – 47cms (expected 41.5 cm)

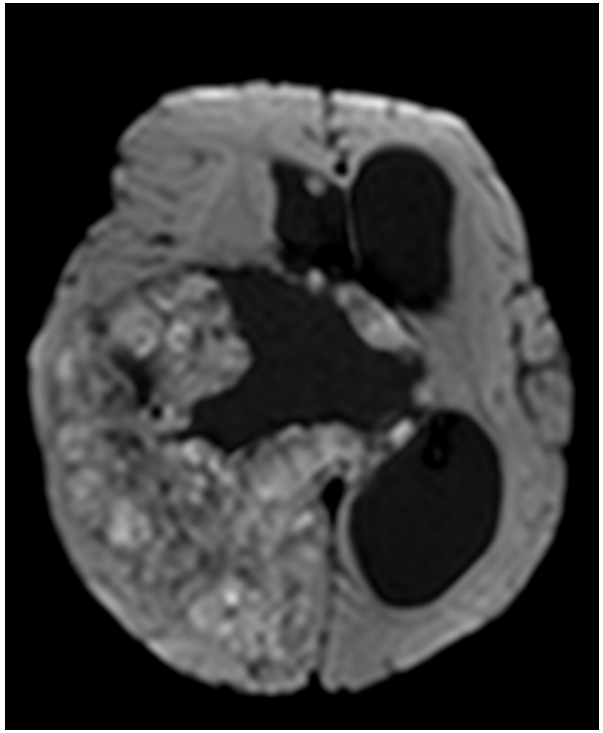
On systemic examination – AF is wide open & bulging

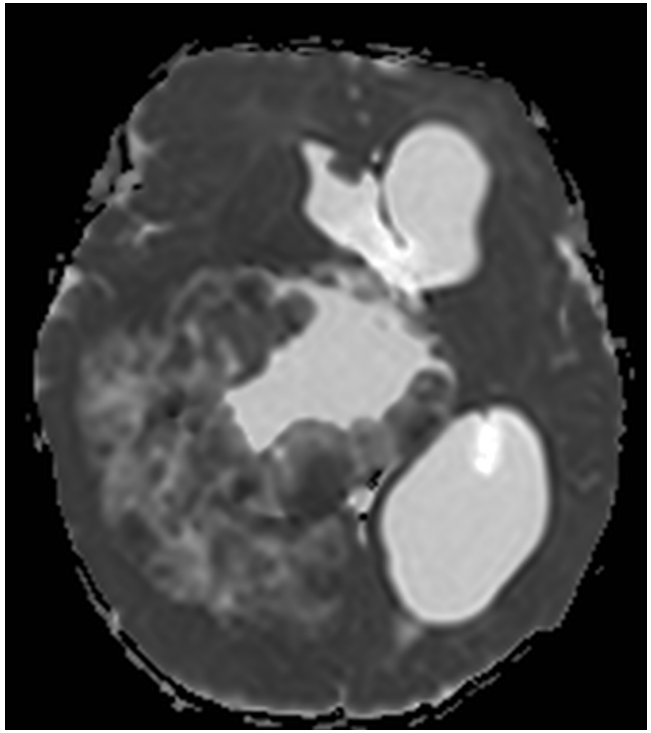


T2 and FLAIR iso to hyperintense **frond like mass** lesion noted in the region of body of right lateral ventricle which is seen to extend into occipital & temporal horn of the right lateral ventricle

The lesion is showing **ill-defined margins with adjacent brain parenchyma** in right high frontal and parietal regions

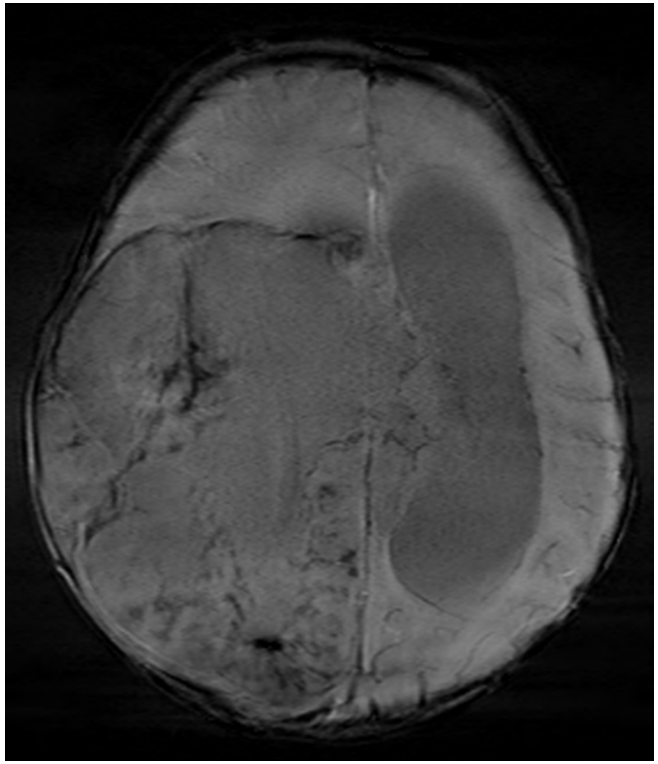




DWI- ADC

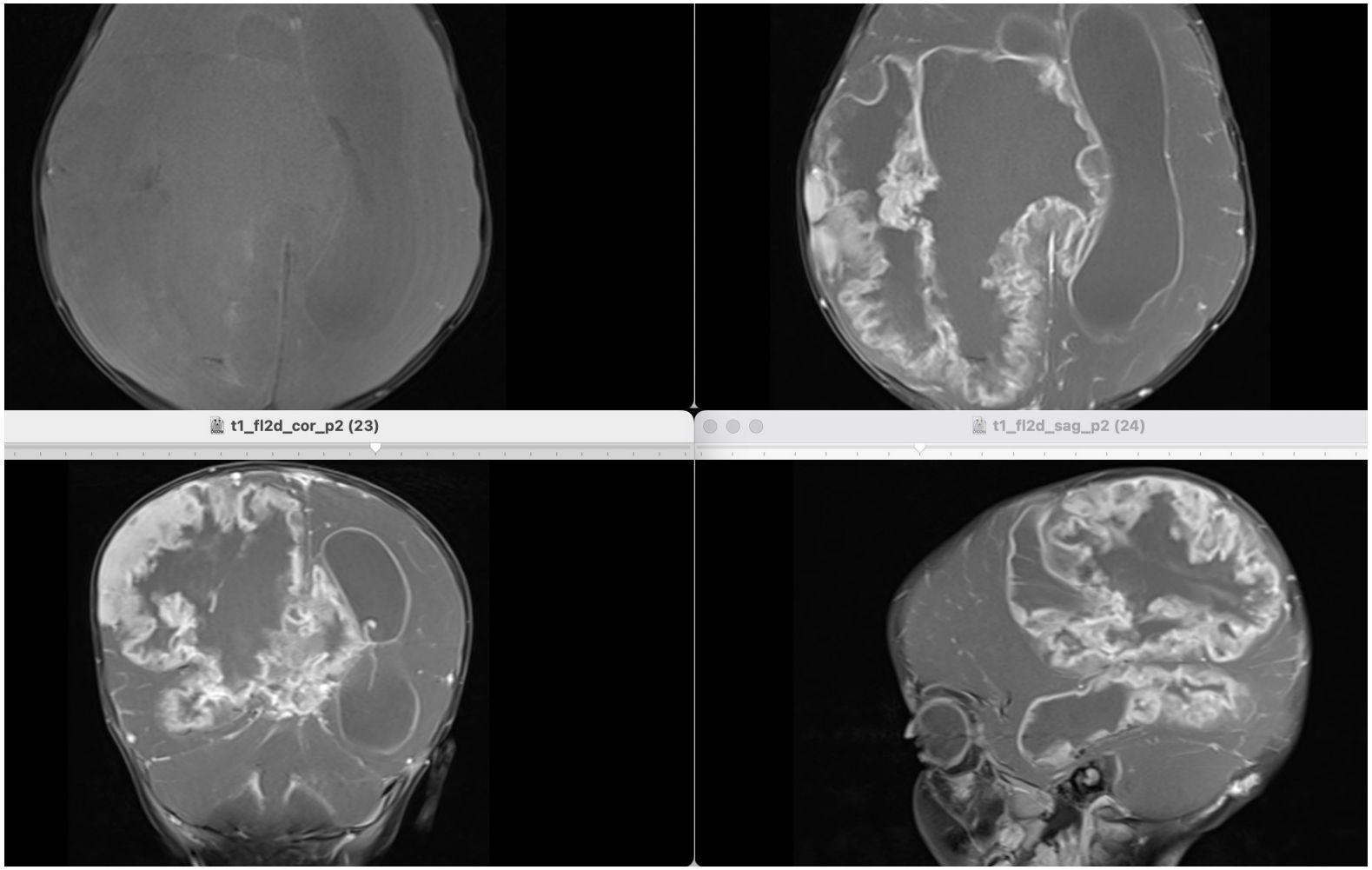
Diffusion restriction on DWI sequence and few areas of blooming on SWI likely hemorrhagic areas

SWI

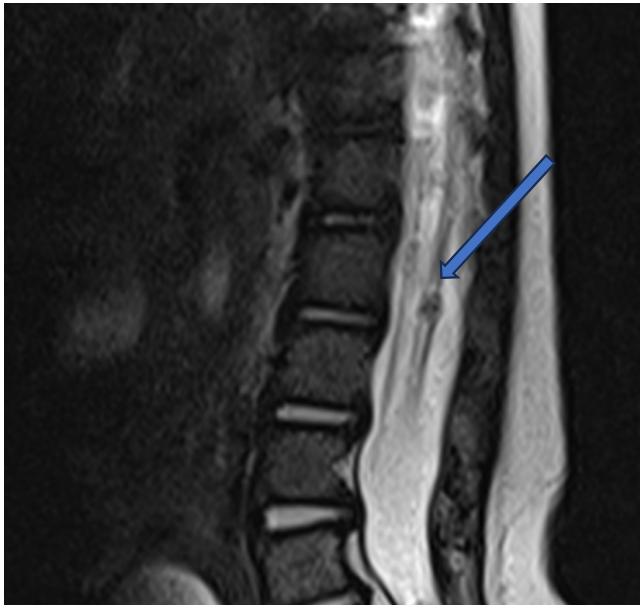


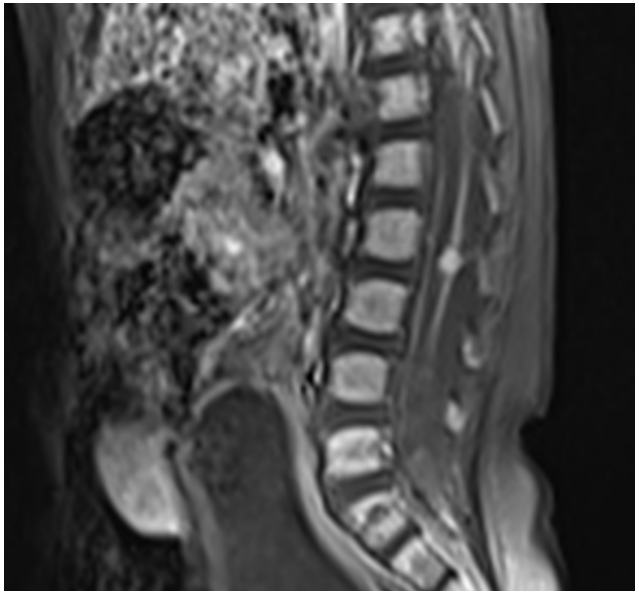
Inferior aspect of the vermis not seen with increased tegmento--vermian angle with dilatation of fourth ventricle likely to be **inferior vermian hypoplasia**.

Axial T1w precontrast

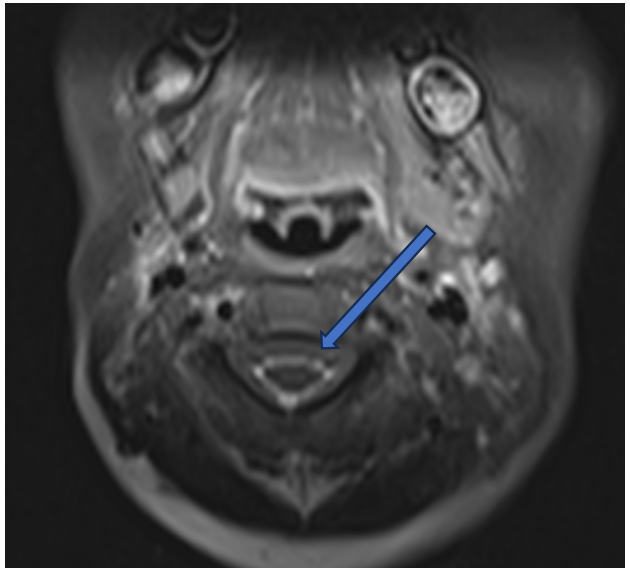


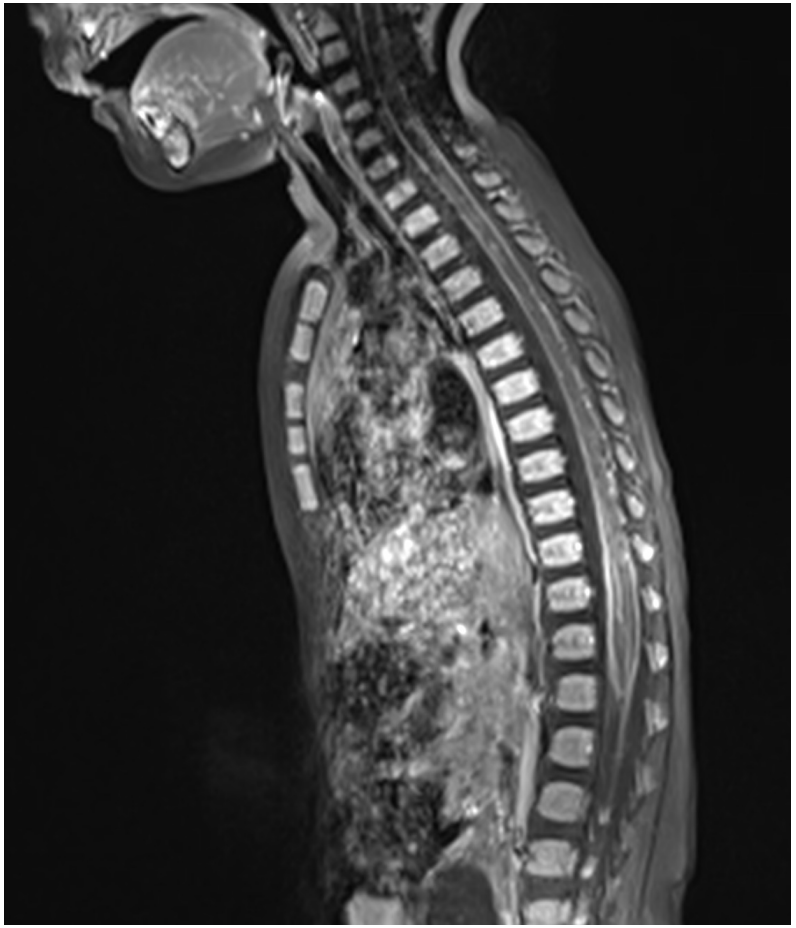
POST CONTRAST – The lesion shows heterogenous enhancement





T2 hypointense lesion involving filum terminale at the level of lower border of L2 vertebra showing contrast enhancement suggestive of- **drop metastases**





Diffuse nodular leptomeningeal enhancement noted along the entire spinal cord s/o **metastasis**

Differentials:

1. *Atypical teratoid rhabdoid tumor- ATRT*
2. *Embryonal tumor with multi-layered rosettes- ETMR*
3. *Choroid plexus carcinoma*

Atypical teratoid-rhabdoid tumour- AT/RT

FAVOUR (+)	AGAINST (-)
Age < 3yrs	
Imaging findings **T1WI** – Iso to hypointense intraventricular mass **T2WI** ➢ Lobulated or irregularly marginated ➢ iso-/hyperintense heterogenous (necrosis & hge) ➢ No peritumoral edema **T1WI C+** ➢ Intense Enhancing solid component, + CSF seeding	Imaging findings – Right choroid plexus not seen separately
Location - lateral ventricle	

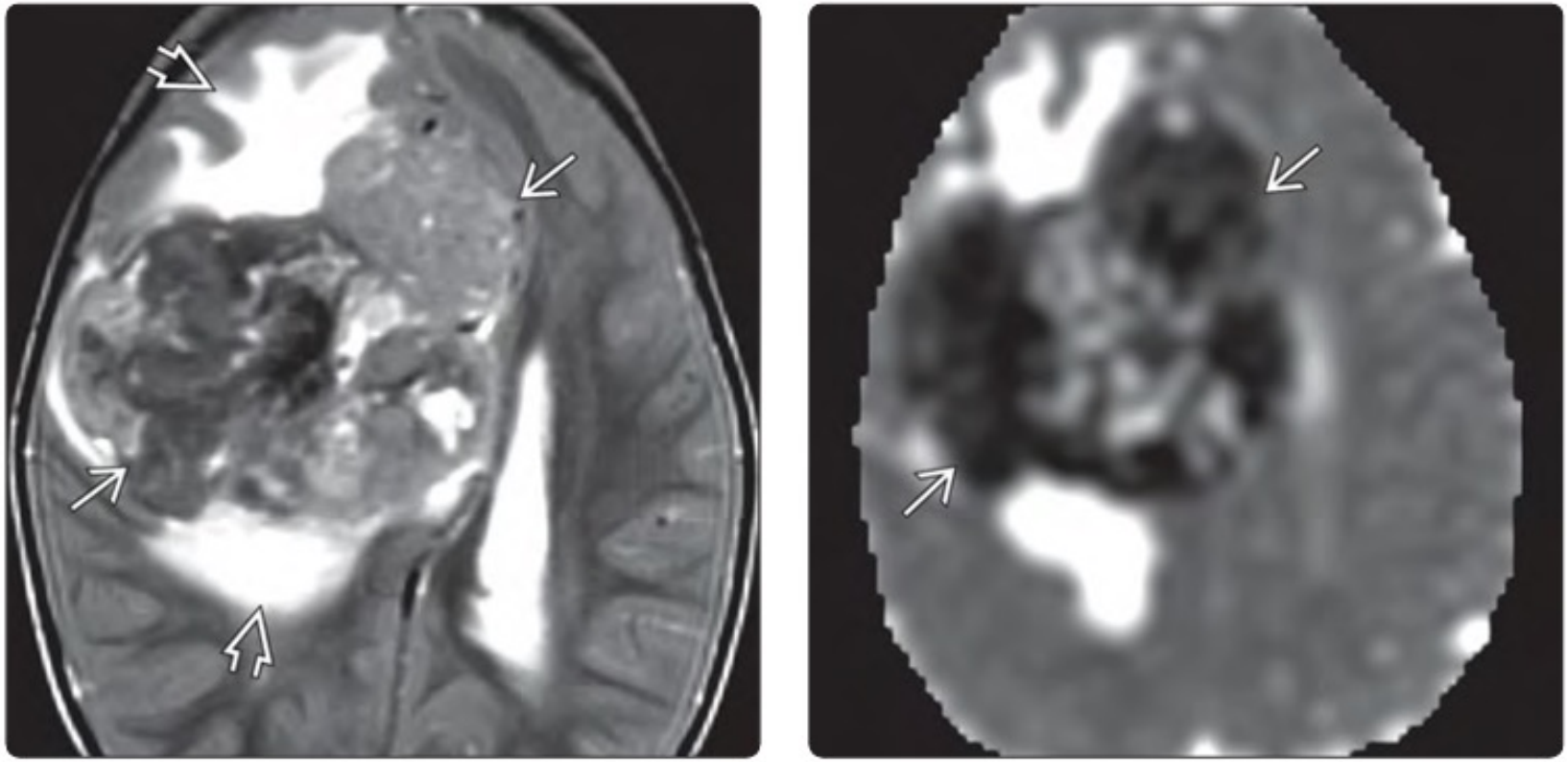
Atypical teratoid-rhabdoid tumour- AT/RT

- Lethal (usually childhood) cancer with SMARCB1/hSNF5 mutations
- Loss of INI1 protein expression hallmark of CNS AT/RT
- WHO grade IV
- **Always consider AT/RT when large tumor found in child younger than 3 years**

IMAGING

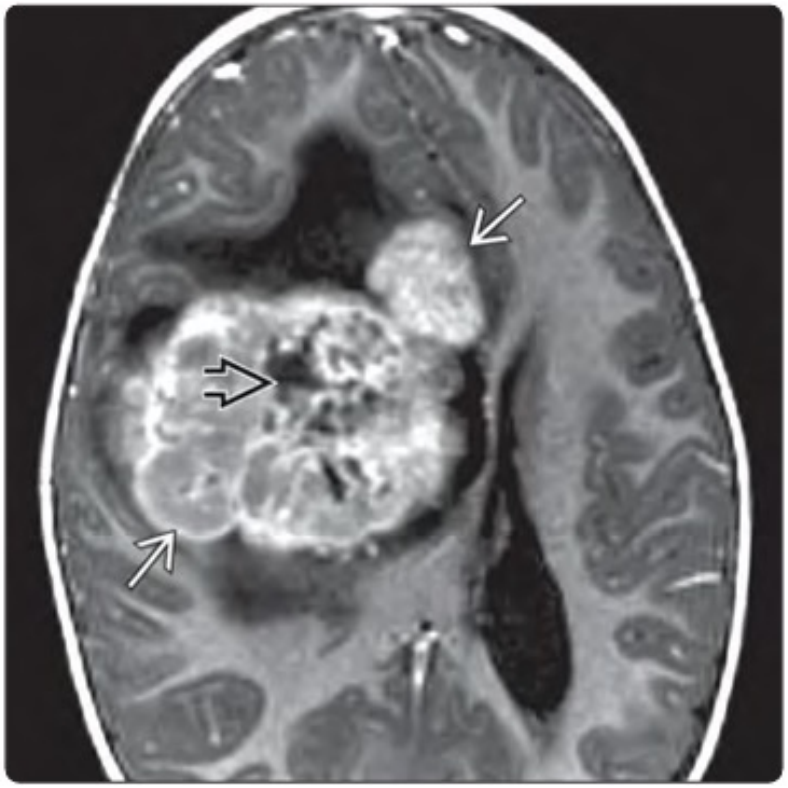
- Heterogeneous intracranial mass in infant and contains cysts or hemorrhage
- Hyperattenuating mass on CT
- Relatively little to no edema for size of tumor
- Intense but heterogeneous enhancement
- Leptomeningeal spread common

Atypical teratoid-rhabdoid tumour- AT/RT



- Axial T2WI MR shows a large, bulky, supratentorial, heterogeneous tumor with central hemorrhage and mass effect. Although some **edema is present , it is less than expected given the size of the tumor.**
- (Right) Axial ADC in the same case shows decreased apparent diffusion coefficient - compatible with the hypercellular nature of the tumor.

Atypical teratoid-rhabdoid tumour- AT/RT



Axial T1WI C+ MR in the **same patient** shows **strong but rather heterogeneous enhancement** .
Some central areas of necrosis can be seen within the generally solid mass.
Sagittal T1WI C+ MR demonstrates "**drop**" **metastases** .

Embryonal Tumor With Multilayered Rosette ETMR

FAVOUR (+)	AGAINST (-)
Age < 2.5 yrs ; M > F	
Imaging findings **T1WI** – Iso to hypointense **T2WI** ➢ Lobulated or irregularly marginated ➢ iso-/hyperintense heterogenous mass (necrosis & hge) ➢ No peritumoral edema **T1WI C+** ➢ Intense Enhancing solid component, + CSF seeding	Imaging findings – Right choroid plexus not seen separately
	Location - lateral ventricle

Embryonal Tumor With Multilayered Rosette ETMR

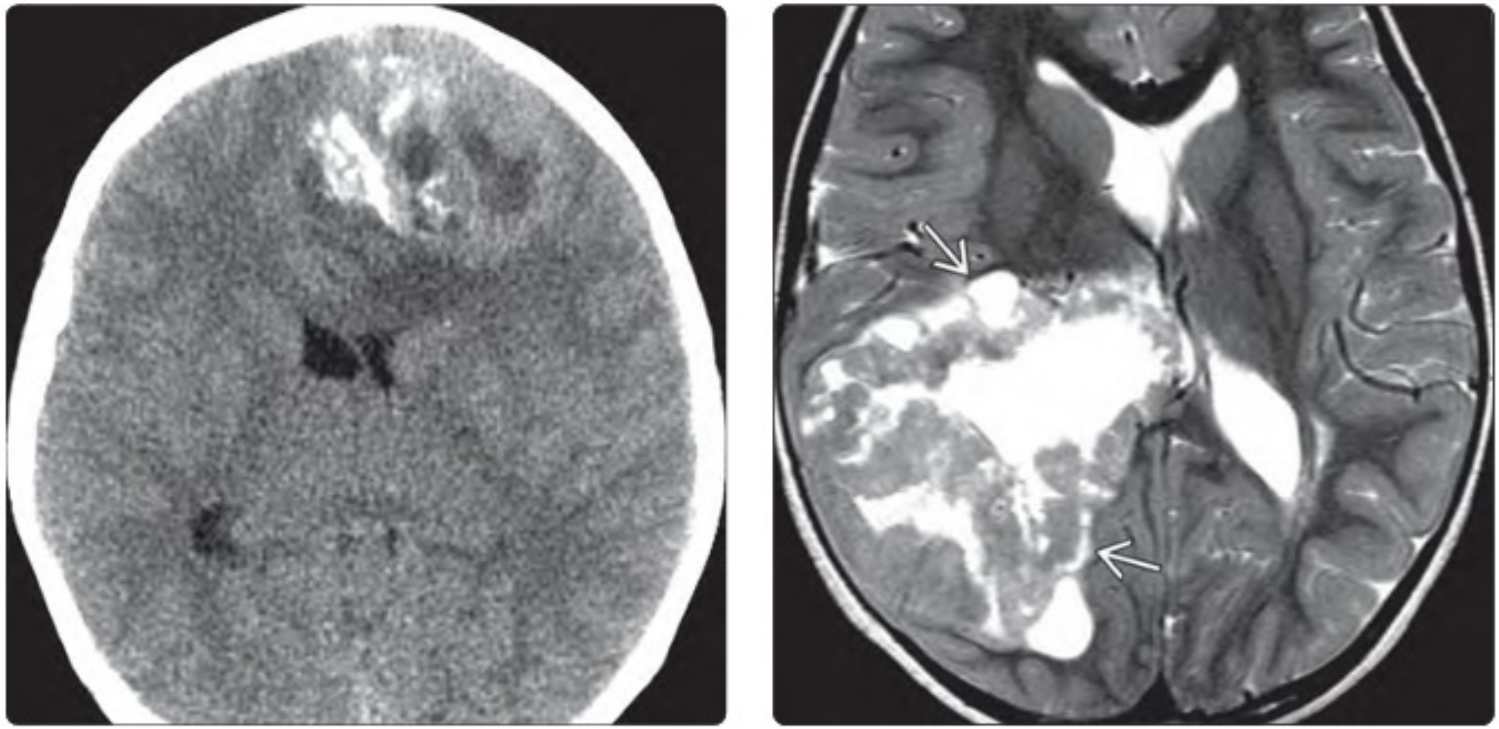
Embryonal tumor composed of undifferentiated neuroepithelial cells

If large bulky hemispheric mass with sparse edema in infant, think ETMR or AT/RT

IMAGING:

- Variable size, based on location
- Hemispheric PNETs larger (mean diameter: 5 cm)
- Sharply delineated to diffusely infiltrative
- Minimal to no peritumoral edema
- **Calcification common (50-70%)**
- Heterogeneous density/signal intensity, enhancement

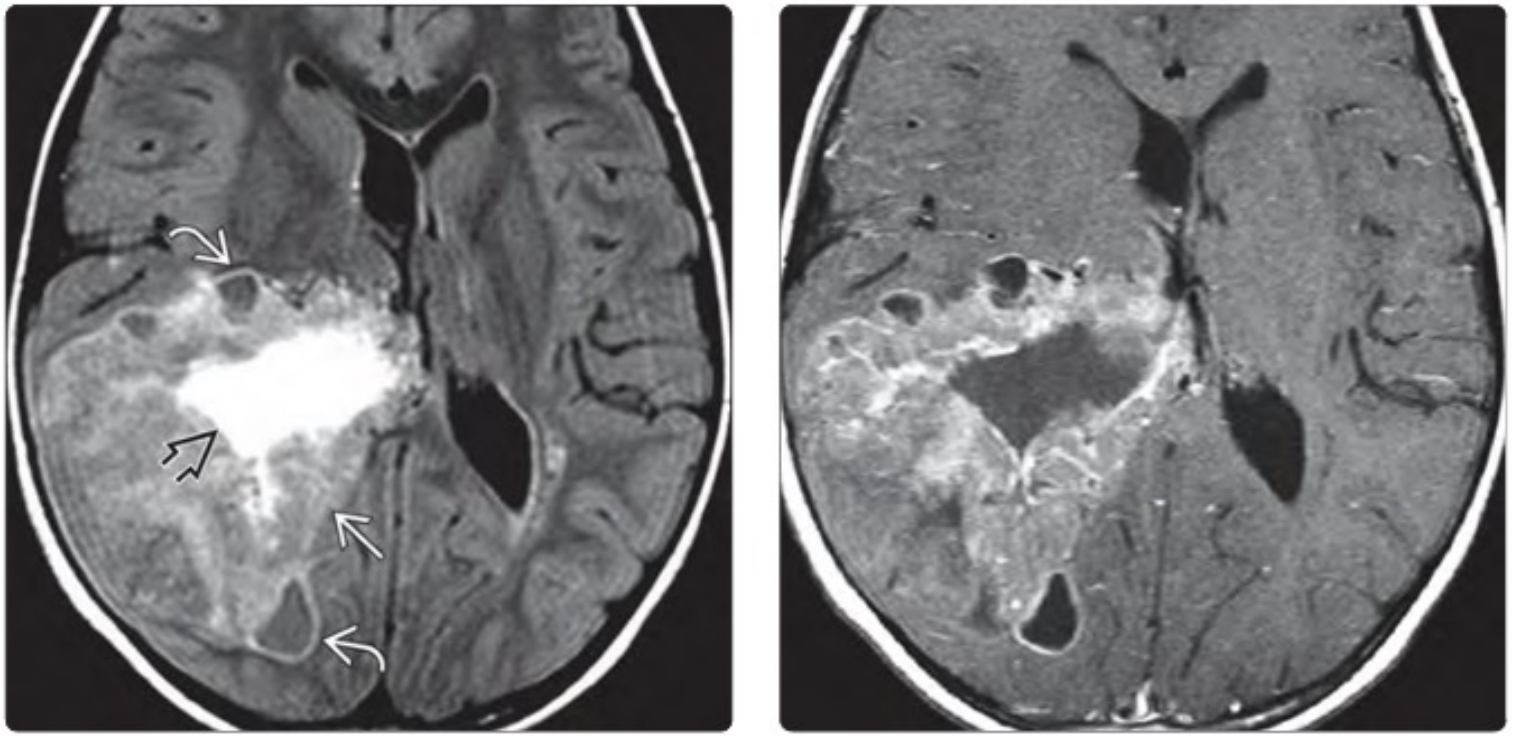
Embryonal Tumor With Multilayered Rosette ETMR



Axial NECT shows a typical, intraaxial, frontal lobe cerebral PNET --with heterogeneity, and little or no peritumoral edema. The tumor is largely hyperattenuating with scattered calcifications.

Another case-- Axial T2WI in a 2 year old boy with seizures shows a large heterogeneous mixed iso- and hyperintense mass in the right parietal lobe. Note the absence of peritumoral edema

Embryonal Tumor With Multilayered Rosette ETMR



FLAIR scan in the same patient shows a **hyperintense area of central necrosis** , solid portions of the tumor and **several intratumoral cysts** that do not suppress completely.

T1 C+ MR in the same patient shows **heterogeneous enhancement in the solid portions** of the mass and around the intratumoral cysts. The central necrotic area does not enhance. PNET was found at surgery

Choroid Plexus Carcinoma

FAVOUR (+)	AGAINST (-)
Age < 2yrs	
Imaging findings **T1WI** – Iso to hypointense intraventricular mass **T2WI** ➢ Lobulated, frond- like appearance ➢ iso-/hyperintense solid-cystic lesion **T1WI C+** ➢ Intense Enhancing solid component, ± CSF seeding and ependymal invasion ➢ Right choroid plexus not seen separately	Imaging findings – Absense of peritumoral edema Absence of prominent flow voids
Location - lateral ventricle	

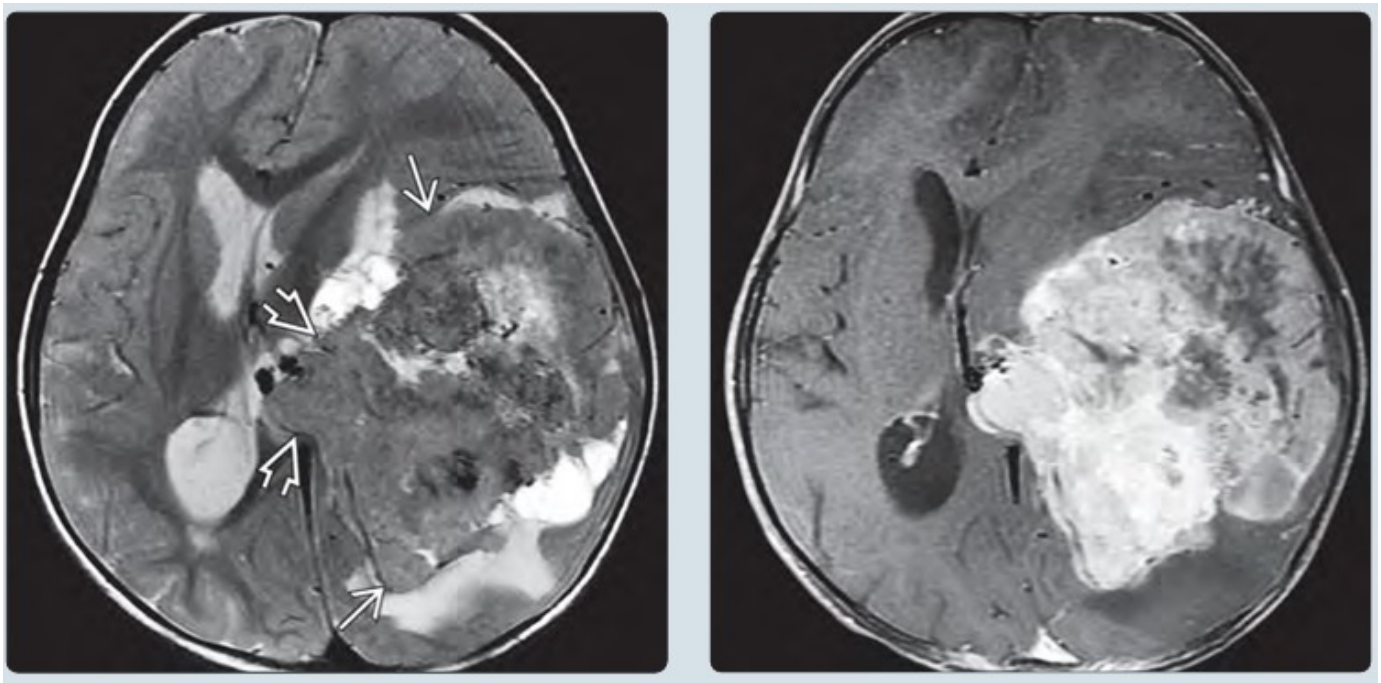
Choroid Plexus Carcinoma

- Malignant tumor originating from epithelium of choroid plexus (WHO grade III)

IMAGING

- Best imaging clue: Child < 2 years with enhancing intraventricular mass and ependymal invasion, ± prominent flow voids
- Asymmetric periventricular white matter edema suggests invasion
- MR may not distinguish papilloma from carcinoma - Heterogeneity, brain invasion, CSF spread favors CPCa
- Important to image spine prior to surgery

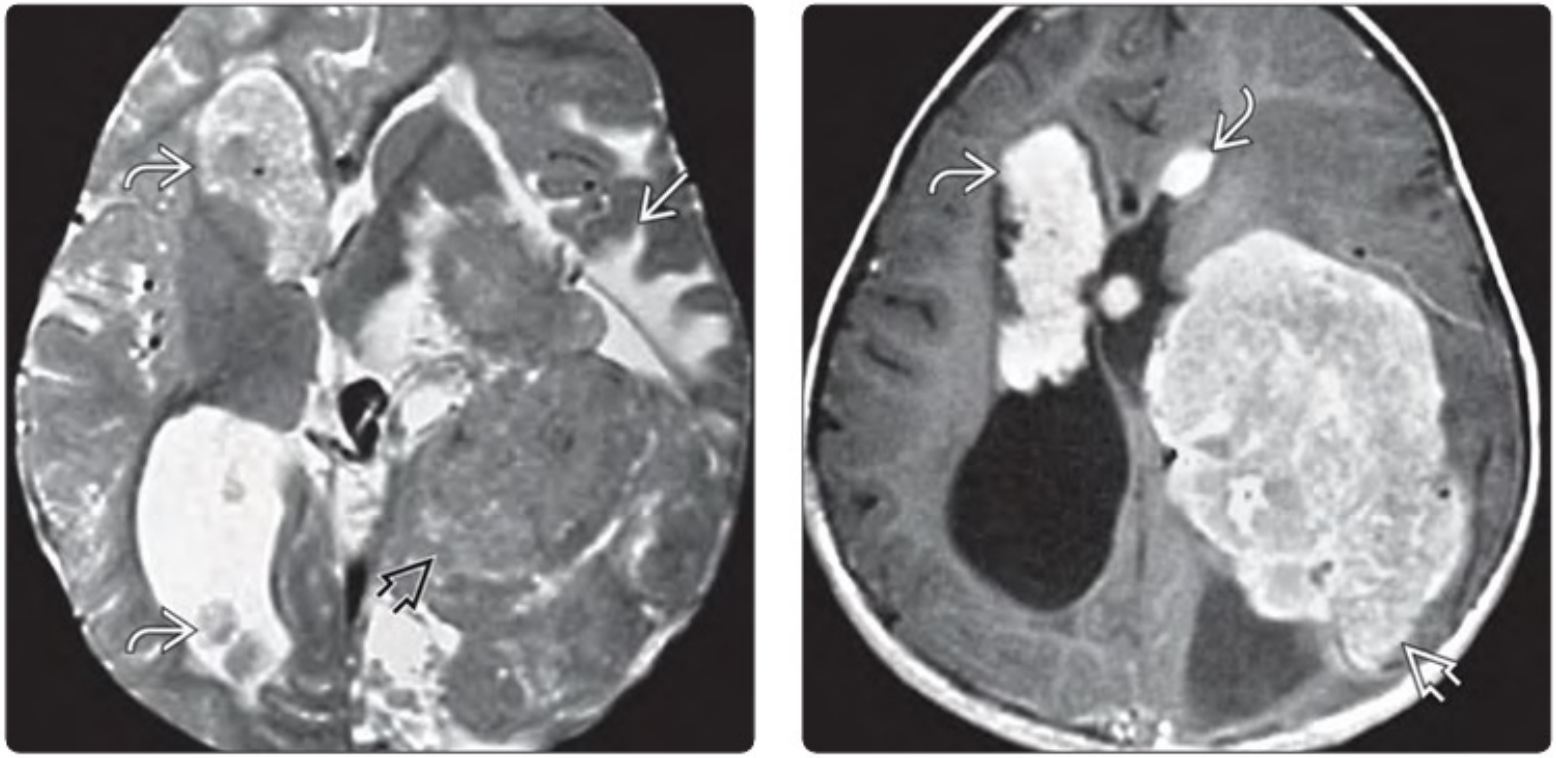
Choroid Plexus Carcinoma



(Left) Axial T2WI shows the lobulated mass is extremely heterogeneous in signal intensity. The lesion appears to arise within the atrium of the left lateral ventricle and invades the adjacent brain extensively

(Right) Axial post-contrast T1WI shows the mass enhances strongly but heterogeneously. Choroid plexus carcinoma (WHO grade III) was found at surgery.

Choroid Plexus Carcinoma



(Left) Axial T2WI MR shows a massive left lateral ventricle choroid plexus carcinoma with multiple nodules of metastatic CSF spread.
Note the asymmetric periventricular T2 hyperintensity related to brain invasion.

(Right) Axial T1WI C+ MR in the same patient shows marked enhancement of the lateral ventricle choroid plexus carcinoma with ependymal invasion and multiple nodules of metastatic CSF spread

FOLLOW UP

- Patient did not undergo any treatment and succumbed to the disease after 1 month