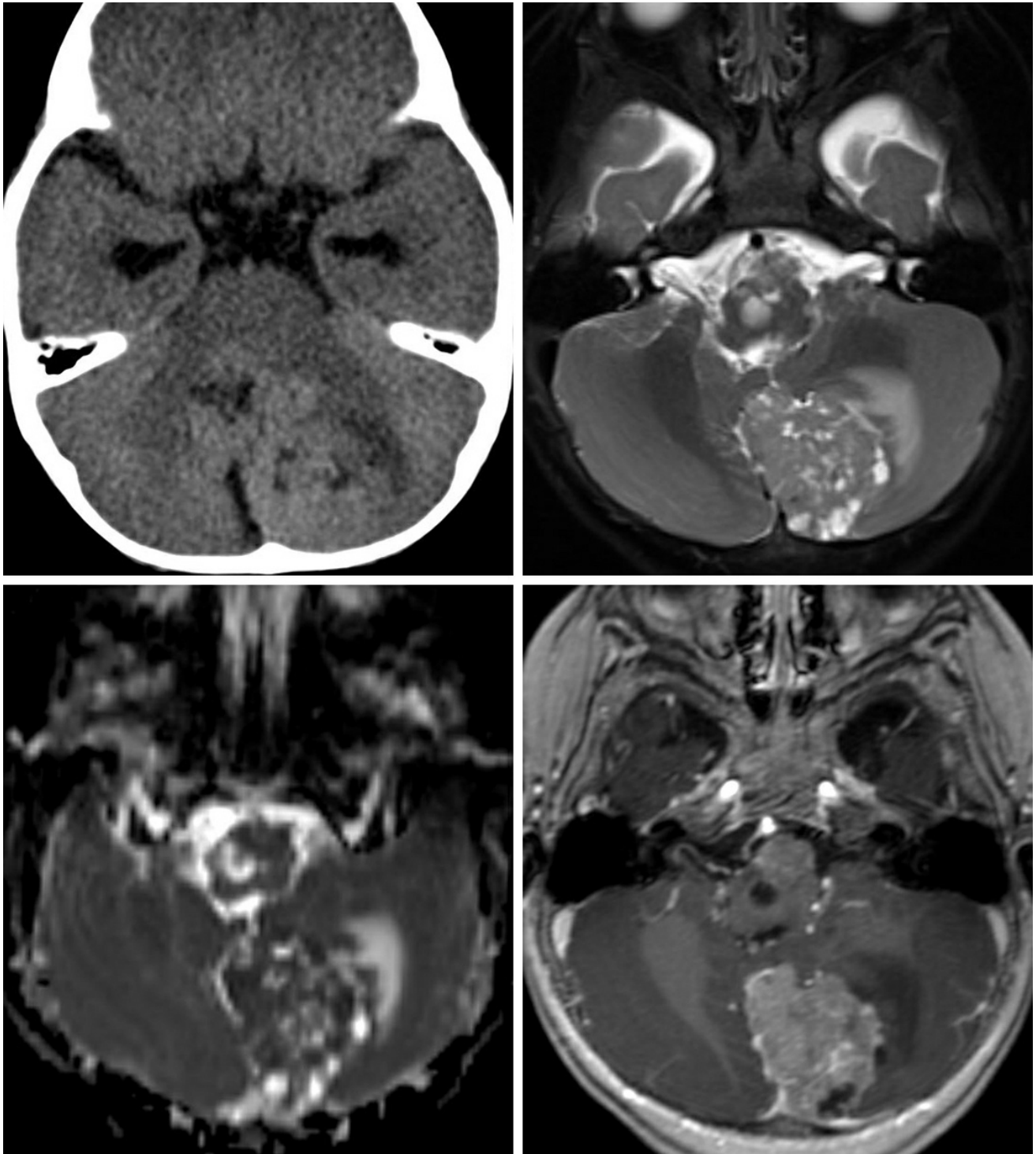




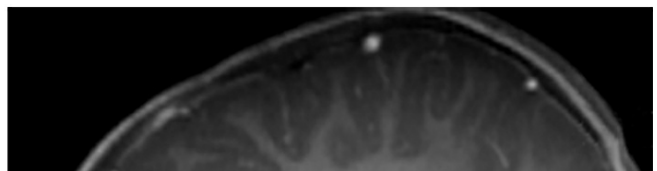
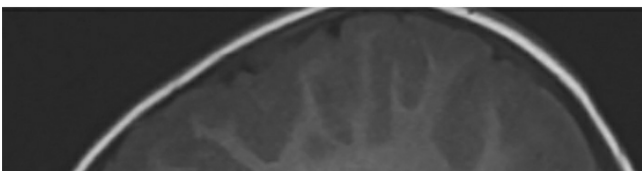
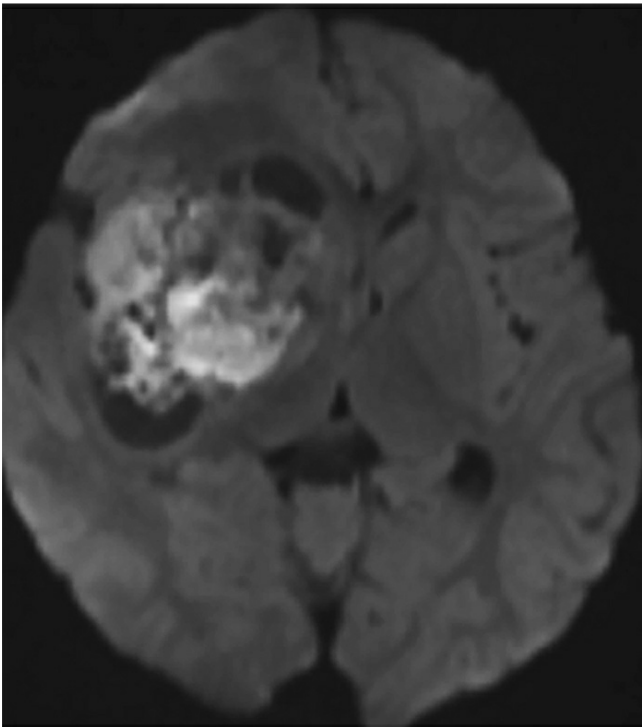
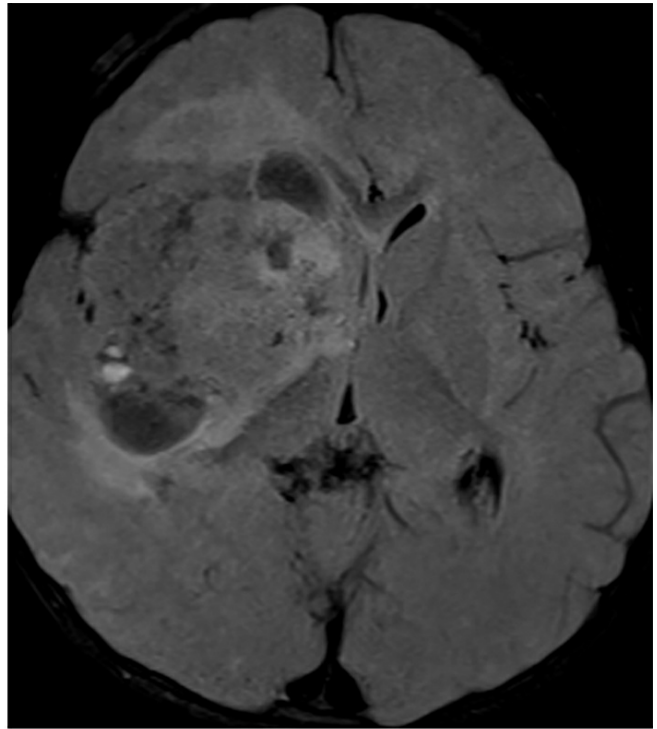
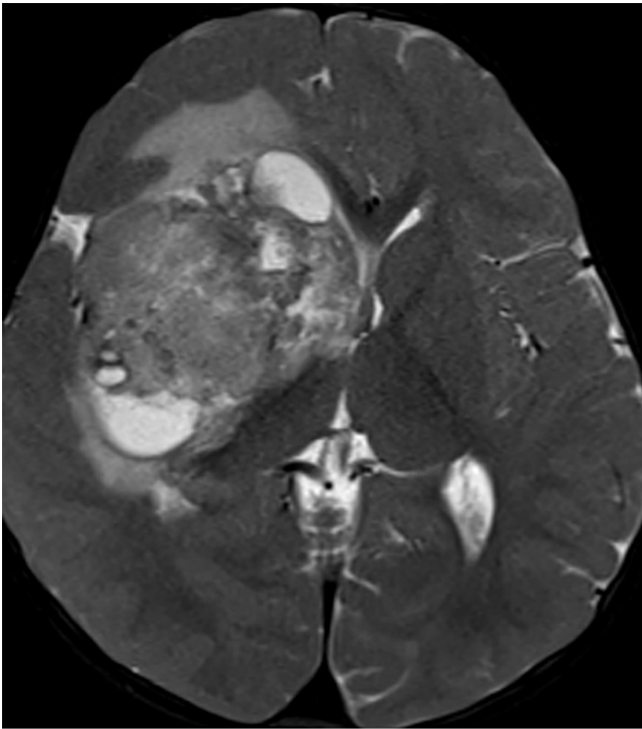
## Atypical Teratoid-Rhabdoid Tumor

*Last Updated: May 2, 2021*



**Figure 1:** (Top Left) Axial nonenhanced CT image demonstrates a heterogeneous, mildly hyperdense AT/RT in the posteromedial left cerebellum. There is associated mass effect on the cerebellum and fourth

ventricle. (Top Right) Axial T2WI shows cystic change within this tumor. Abnormality in the medulla represents leptomeningeal metastasis of tumor with associated invasion. (Bottom Left) Apparent diffusion coefficient (ADC) image shows dark restricted diffusion of hypercellularity within the tumor. (Bottom Right) Axial T1WI postcontrast image shows the avid enhancement and cystic changes of this high-grade tumor.



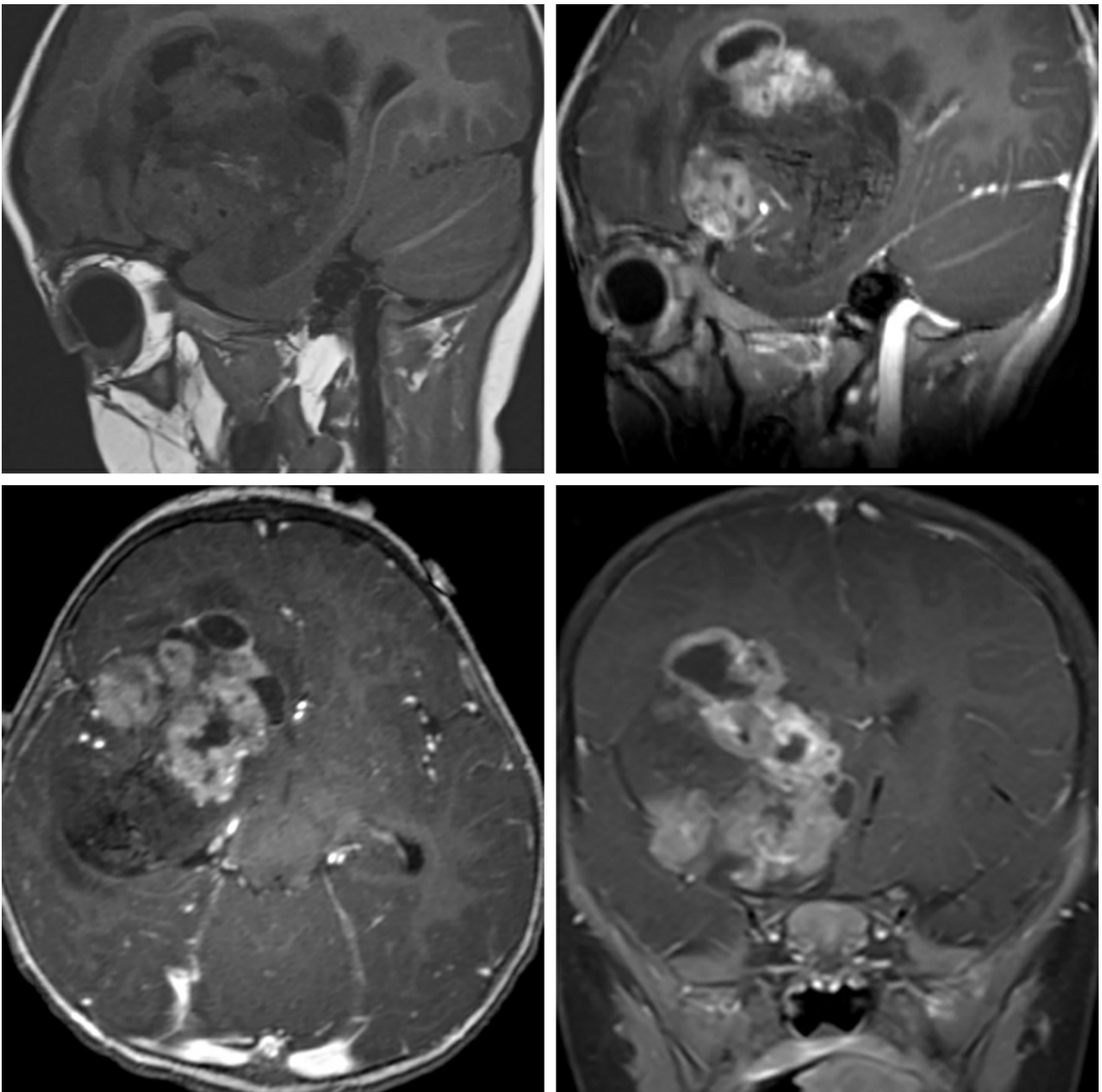


Figure 2: This AT/RT demonstrates stereotypical high complexity, with solid and cystic/necrotic components, surrounding hyperintense vasogenic edema (row 1, left, T2; row 1, right, FLAIR), restricted diffusion indicating hypercellularity (row 2, left, DWI; row 2, right, ADC) and heterogeneous contrast enhancement (row 3, right, and row 4, left and right) when compared to the precontrast T1 (row 3, left). This AT/RT is atypical in its supratentorial location, these tumors usually being infratentorial. AT/RTs are also often indistinguishable by imaging alone from other highly complex pediatric brain tumors such as primitive neuroectodermal tumors (PNETs) and [medulloblastoma](#).

## BASIC DESCRIPTION

- Aggressive, infiltrative pediatric rhabdoid tumor with highly variable appearance and histology

## PATHOLOGY

- WHO grade IV
- Involves mesenchymal, neuronal, glial, and epithelial cell lines (“teratoid”)
  - ±Rhabdoid cells, primitive neuroectodermal cells
- Mutations of SMARCB1/hSNF5 and loss of INI1 protein is diagnostic of atypical teratoid-rhabdoid tumor (AT/RT)
- Familial mutations in rhabdoid tumor predisposition syndrome (RTPS)

## CLINICAL FEATURES

- Most commonly afflicts children <3 years old and usually in the posterior fossa; tumors in adults are often in an atypical location
- No gender predilection
- Common presenting signs/symptoms related to increased intracranial pressure/hydrocephalus
  - Vomiting, altered mental status, macrocephaly
- Treatment: surgical resection ± adjuvant chemoradiation
  - Often unresectable
- Prognosis: overall 5-year survival, <30%
  - Survival with leptomeningeal spread, 16 months
  - Survival without leptomeningeal spread, 149 months

## IMAGING FEATURES

- General
  - Highly variable, nonspecific morphology and imaging characteristics
  - Supratentorial or infratentorial
    - More than half arise within the posterior fossa



- Variable size; often large at time of diagnosis
- Mixed solid-cystic mass
- ±Calcification, hemorrhage, and necrosis
- ±Obstructive hydrocephalus
- Leptomeningeal/cerebrospinal fluid (CSF) dissemination common (>30%)
- CT
  - Hyperdense mass
  - Hyperdense calcification and hemorrhage or hypodense cysts may be seen
  - Avid, heterogeneous enhancement on contrast-enhanced CT
- MRI
  - T1-weighted imaging (T1WI), heterogeneous signal due to cystic or hemorrhagic components
  - T2-weighted imaging (T2WI), heterogeneous signal due to cystic or hemorrhagic components
  - Fluid-attenuated inversion recovery (FLAIR) imaging, heterogeneous, hyperintense periventricular signal due to hydrocephalus/interstitial edema, little peritumoral edema
  - Gradient recalled echo T2\*-weighted imaging (T2\*GRE), hypointense signal due to calcification and hemorrhage
  - T1WI+C imaging, majority show heterogeneous enhancement; may see enhancing CSF dissemination along the brain, cranial nerves, spinal nerves, or spinal cord
  - Diffusion-weighted imaging (DWI), demonstrates diffusion restriction in hypercellular regions of tumor
  - MR spectroscopy, elevated Cho, decreased NAA, ±lactate/lipid peaks

## IMAGING RECOMMENDATIONS

- MRI without and with intravenous contrast including both brain and

spine due to risk of CSF dissemination

For more information, please see the corresponding chapter in [Radiopaedia](#).

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