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Atypical Teratoid/Rhabdoid Tumor (AT/RT)

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Definition

Atypical teratoid/rhabdoid tumor (AT/RT) is a rare, highly malignant tumor of early childhood, most commonly diagnosed in infants who are less than 3 years. First described by Rorke and colleagues in 1987, the AT/RT received its designation because of its complex histological components. Prognosis is extremely poor with a median survival of 6–11 months. Factors associated with improved prognosis include supratentorial location, localized disease at the time of presentation, and complete resection (Torchia et al. 2015). Over half of AT/RTs identified are located within the posterior fossa (brainstem, cerebellum, and predominantly the cerebellopontine angle) (Rorke et al. 1996). Roughly one-fourth are supratentorial and 8% may be multifocal. Clinical presentation varies largely by tumor location and size. Infants, in particular, may present with nonspecific symptoms, including lethargy, vomiting, and failure to thrive. Older children (>3 years of age) may demonstrate more specific problems, including head tilt, diplopia, cranial nerve palsy, headache, and hemiplegia (Rorke and Biegel 2000). Often histologically confused with PNET/medulloblastoma.

Cross-References

- ▶ Medulloblastoma
- ▶ Primitive Neuroectodermal Tumor

References and Readings

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