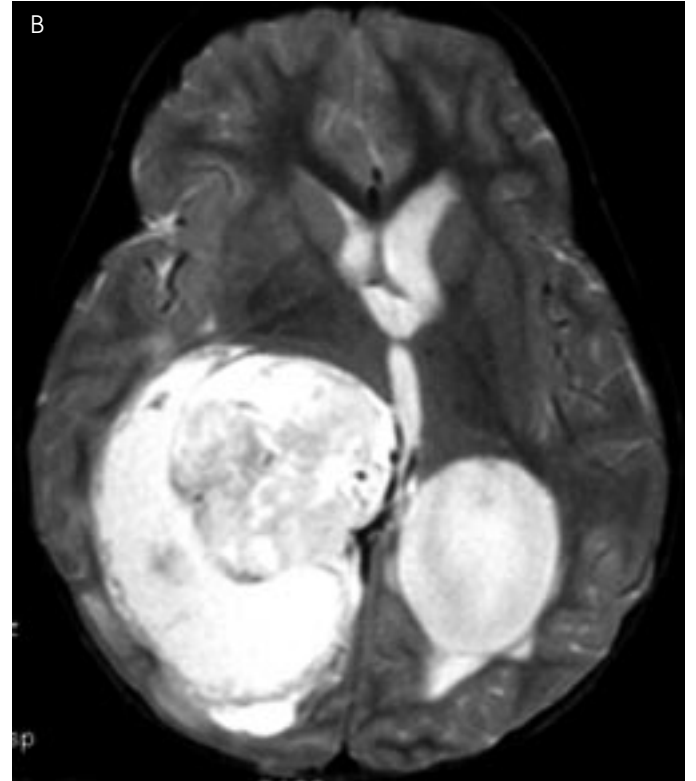
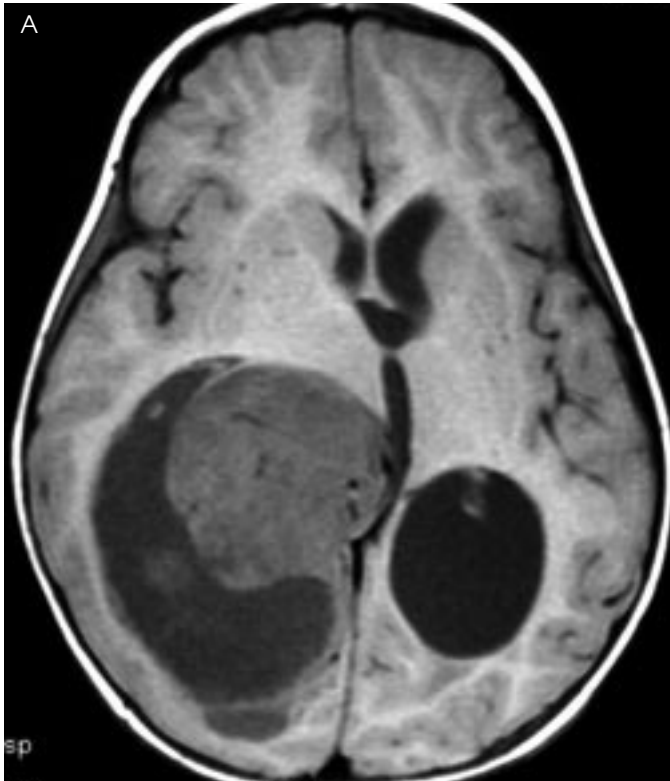


# Case W2

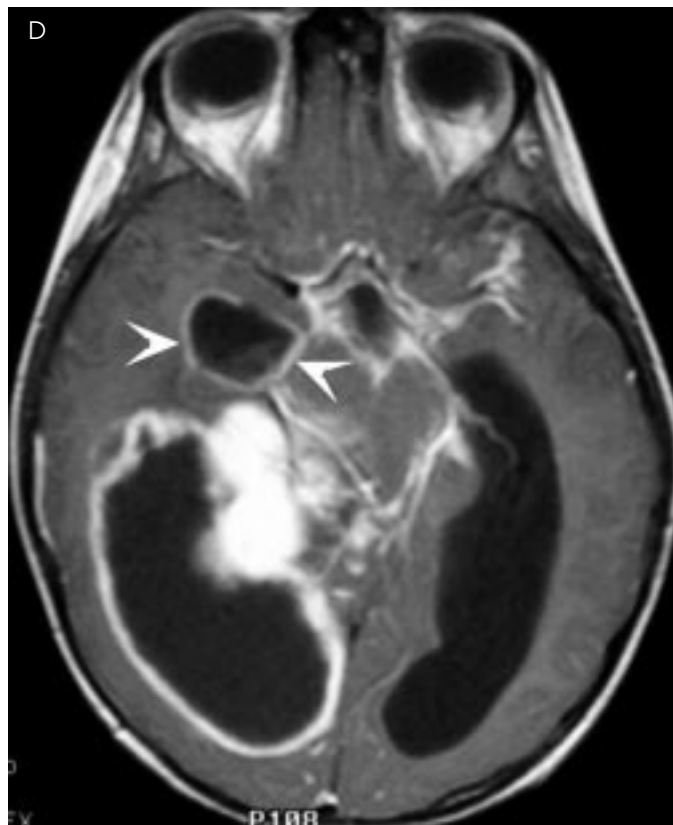
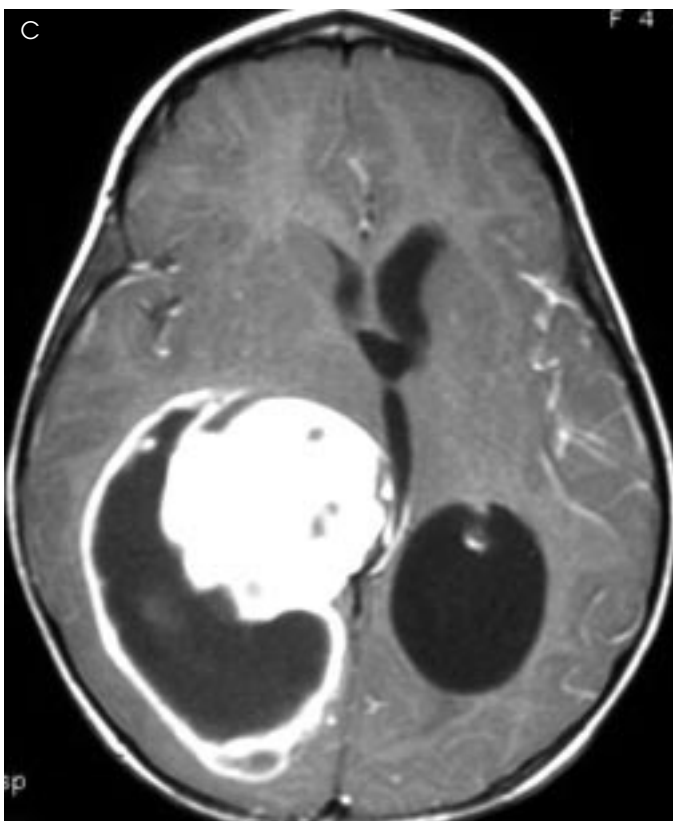
## Clinical Presentation

A 4-year-old girl presents with increasing headache and vomiting.



## Radiologic Findings

An axial T1-WI (Fig. A) demonstrates a large round mass centered near the right ventricular atrium. An adjacent cyst is present. On this image, one cannot be certain whether this represents a tumor cyst or the trapped lateral ventricle peripheral to this lesion. An axial T2-WI (Fig. B) demonstrates that the mass is heterogenous in signal intensity. In addition, transependymal flow of CSF is appreciated around the left ventricular atrium secondary to obstructive hydrocephalus.



### Pearls

- MR better delineates the extent of disease and is more sensitive to ependymal and/or subarachnoid spread of disease.
- Because of a high incidence of CSF spread of disease, the spinal axis should be screened with contrast-enhanced images at the time of initial diagnosis and in follow-up.

A post-gadolinium T1-WI (Fig. C) demonstrates that the mass enhances intensely. In addition, there is enhancement around the periphery of what appears to be a trapped ventricle, consistent with CSF spread of tumor. An axial post-gadolinium T1-WI (Fig. D) at the level of the mesencephalon shows abnormally intense linear enhancement around the mesencephalon, as well as the right temporal horn (arrowheads), also consistent with CSF spread of tumor.

## Diagnosis

Malignant rhabdoid tumor

## Differential Diagnosis

- Parenchymal malignant rhabdoid tumor: ependymoma (may be indistinguishable), supratentorial primitive neuroectodermal tumor (may be indistinguishable), teratoma/teratocarcinoma (often contains fat, typically midline)
- Intraventricular malignant rhabdoid tumor: choroid plexus papilloma/carcinoma (often more lobulated, “cauliflowerlike”)

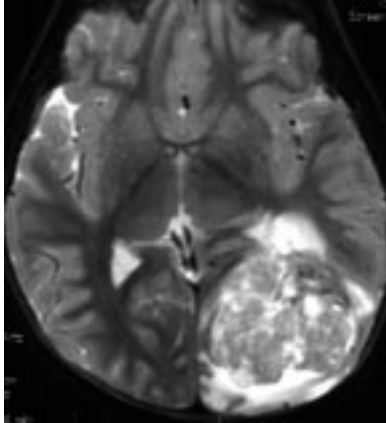
## Discussion

### Background

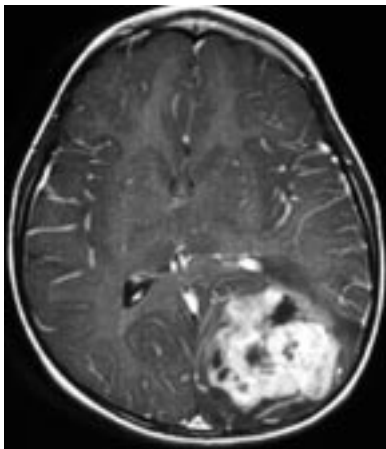
Malignant rhabdoid tumor is a rare primary brain neoplasm that affects infants and young children, with a mean age of occurrence of 3 years. These tumors may

### Pitfall

- May present as a parenchymal (Figs. E and F) or an intraventricular mass, so include in both differentials in a child.



**Fig. E.** Axial T2-WI in a 3-year-old demonstrates a heterogeneous mass centered in the left occipital lobe. A moderate amount of surrounding vasogenic edema is present.



**Fig. F.** A post-gadolinium T1-WI of the patient in Figure D shows heterogeneous enhancement of the partially cystic or necrotic mass. At surgery, this was found to represent a malignant rhabdoid tumor.

be parenchymal or intraventricular. They are typically large at the time of presentation and have often already disseminated through the CSF at the time of diagnosis.

## Clinical Findings

Patients present with signs of elevated intracranial pressure (lethargy, vomiting), seizures, and/or focal neurologic deficits.

## Etiology

The histogenesis of this tumor is controversial, and these tumors were named for their light microscopic resemblance to rhabdomyosarcoma.

## Pathology

### Gross

- Mainly solid with areas of necrosis

### Microscopic

- Pathologically identical to malignant rhabdoid tumor of the kidney
- Uniformly round tumor cells; vacuolated cytoplasm with occasional PAS-positive inclusions; positive immunoreactivity for vimentin

## Imaging Findings

### CT

- Very heterogeneous
- Patchy enhancement
- May contain flecks of calcification

### MR

- Usually isointense to gray matter on T1- and T2-WIs
- Enhance heterogeneously
- Edema varies from minimal (intraventricular lesion) to severe

## Treatment

- Surgery
- Chemotherapy
- Craniospinal irradiation

## Prognosis

- Generally very poor, with survival usually less than 12 months
- Combination of surgery, radiation therapy, systemic chemotherapy, and triple intrathecal chemotherapy has been reported to lead to cure

## Suggested Readings

Hanna SL, Langston JW, Parham DM, Douglass EC. Primary malignant rhabdoid tumor of the brain: clinical, imaging, and pathologic findings. *AJNR* 14:107–115, 1993.

Satoh H, Goishi J, Sogabe T, et al. Primary malignant rhabdoid tumor of the central nervous system: case report and review of the literature. *Surg Neurol* 40:429–434, 1993.