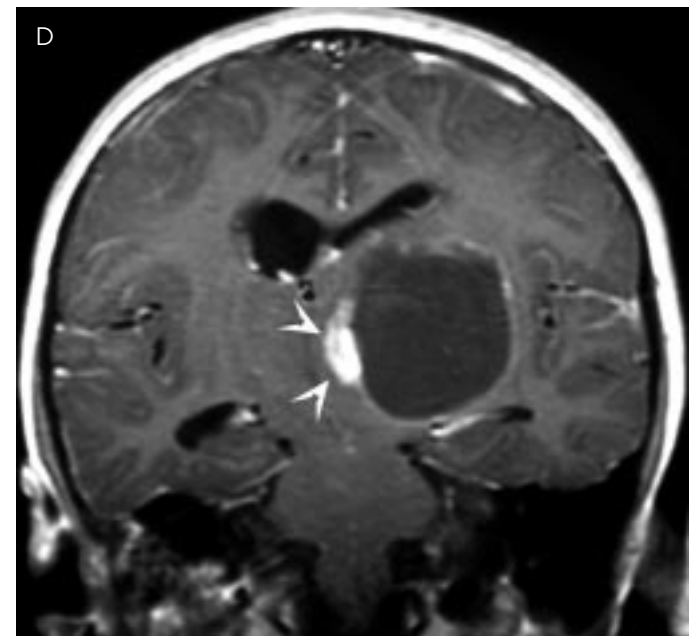
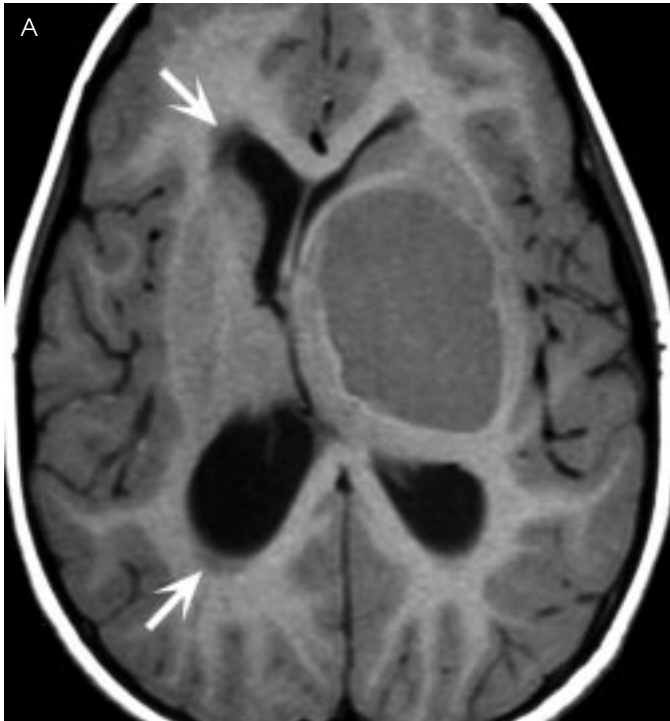


Case W1

Clinical Presentation

An 8-year-old boy with facial asymmetry noted on his school picture presents for evaluation.



Radiologic Findings

An axial T1-WI (Fig. A) demonstrates an ovoid lesion centered in the left basal ganglia, with mass effect on the lateral ventricle and third ventricle. The right lateral ventricle is mildly enlarged due to compression at the level of the foramen of Monro. There is minimal transependymal flow of CSF capping the ventricle (*arrows*). An axial T2-WI (Fig. B) demonstrates that the lesion is homogeneously hyperintense, but with a thin peripheral hypointense rim. The lesion is hyperintense compared with CSF. An axial post-gadolinium T1-WI (Fig. C) demonstrates thin irregular enhancement around the margin of the lesion. The center of the mass is nonenhancing. The signal characteristics suggest a cystic mass containing mildly hemorrhagic or proteinaceous fluid. A coronal T1-WI (Fig. D) shows a more focal intensely enhancing nodule along the medial aspect of the lesion (*arrowheads*).

Diagnosis

Juvenile pilocytic astrocytoma (JPA)

Differential Diagnosis

- Ependymoma (usually more heterogeneous, larger solid component)
- Germinoma (usually more solid, homogeneous; typically midline in location)
- High grade astrocytoma (infiltrative and heterogeneous, surrounding edema)
- Infectious cyst (e.g., echinococcus) (often contains fluid-debris level)
- Neuroepithelial cyst (does not enhance)

Discussion

Background

Astrocytomas account for 30% of supratentorial neoplasms in children. They are usually located deep within the basal ganglia, hypothalamus, or thalamus, or involve the optic chiasm and optic pathways. Most of these are low-grade tumors. The juvenile pilocytic type of astrocytoma is less common supratentorially than in the cerebellum. These tumors are often large at the time of presentation. Unlike cerebellar JPAs, which have a peak incidence at ~10 years of age, hemispheric JPAs are more commonly identified in the teens and early twenties while suprasellar JPAs are most commonly identified in infancy.

Clinical Findings

Clinical findings vary with the location of the mass: headache, seizures, focal neurological deficits, visual changes.

Pathology

Similar to cerebellar astrocytomas

Gross

- May be solid, mixed cystic and solid, or cystic with a mural nodule

Microscopic

- Low or absent mitotic activity, absence of necrosis

Pearl

- Rarely, JPAs may disseminate throughout CSF spaces even while remaining histologically “benign.”

Pitfall

- If the cyst wall enhances, it may be neoplastic rather than representing compressed brain tissue and will likely need to be removed at the time of surgery.

- Often two distinct patterns: densely compact cells with pilocytic processes and more spongiform, loosely organized foci

Imaging Findings**CT**

- Usually well circumscribed
- Little or no surrounding edema
- Cystic with an enhancing mural nodule or a solidly enhancing mass
- Calcification in ~10%

MR

- Nodule usually iso- or hypointense on T1-WI, hyperintense on T2-WI
- Solid portions of tumor and the mural nodule enhance intensely
- Cyst wall usually nonenhancing

Treatment

- Surgical resection
- Radiation therapy and/or chemotherapy depending on the extent of resection and any evidence of lesion progression

Prognosis

Supratentorial JPAs have a somewhat poorer prognosis than infratentorial JPAs, as they are often more difficult to completely resect

Suggested Readings

Finizio FS. CT and MR aspects of supratentorial hemispheric tumors of childhood and adolescence. *Child's Nerv Syst* 11:559–567, 1995.

Gilles FH, Sobel EL, Tavaré CJ, et al. Age-related changes in diagnoses, histological features, and survival in children with brain tumors: 1930–1979. *Neurosurgery* 37:1056–1068, 1995.

Palma L, Guidetti B. Cystic pilocytic astrocytomas of the cerebral hemispheres: surgical experience with 51 cases and long-term results. *J Neurosurg* 62:811–815, 1985.