

## *Neuronal and Mixed Tumors*

Updated: April 24, 2010

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### (CENTRAL) NEUROCYTOMA

- *benign* tumor of slowly growing **well-differentiated neurons**.

- **young adults** (15-40 yrs).

**PATHOLOGY**

**Light microscopy** - monomorphic small cells with evenly spaced, round, uniform nuclei (often mistaken for *OLIGODENDROGLIOMA* or *EPENDYMOMA*), and no anaplastic features.

Neuronal lineage must be confirmed:

1. **Immunohistochemical stains** for neurons (neuron-specific enolase, S100, synaptophysin).
2. **Electron microscopy** - true neuronal nature of neoplasm (neuritic processes, neurosecretory granules, neurofilaments, well-formed synapses).

**LOCATION**

- 3<sup>rd</sup> or lateral **ventricles** (probably commonest lateral ventricular masses in this age group).

- typical location - frontal horns and bodies of lateral ventricle, frequently attached to septum pellucidum and sometimes extending through foramen of Monro.

**CLINICAL FEATURES**

- ICP↑ caused by ventricular obstruction.

**DIAGNOSIS**

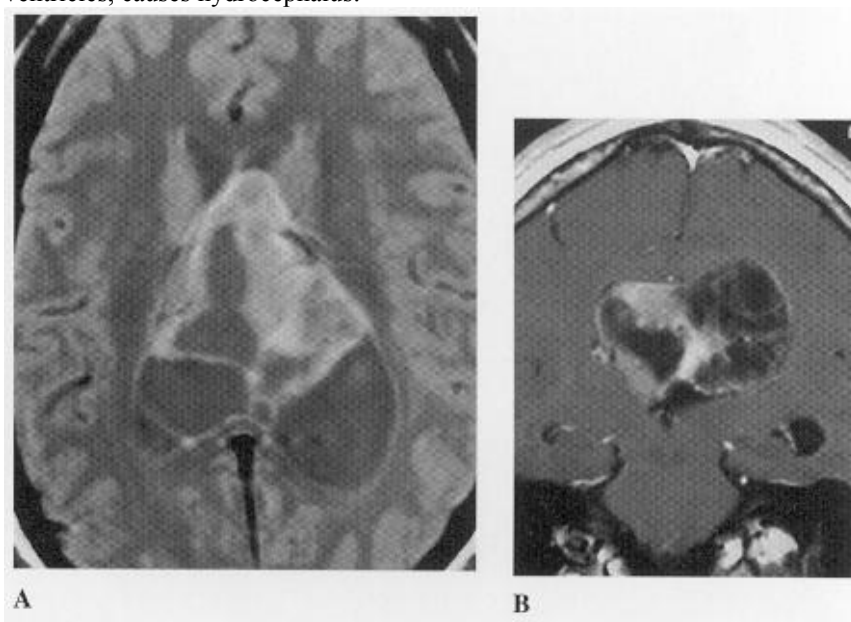
**CT** - calcification and small cysts, obstructive hydrocephalus.

**MRI** - isodense intraventricular mass with variable cyst formation and contrast enhancement.

Contrast MRI - right lateral ventricular neurocytoma producing obstruction of foramen of Monro:



Contrast MRI - partly cystic, multi-septated, enhancing mass, related to septum pellucidum, fills bodies of both lateral ventricles, causes hydrocephalus:



**TREATMENT**

**Surgical resection** is often curative (± radiotherapy).

### DYSEMBRYOPLASTIC NEUROEPITHELIAL TUMOR

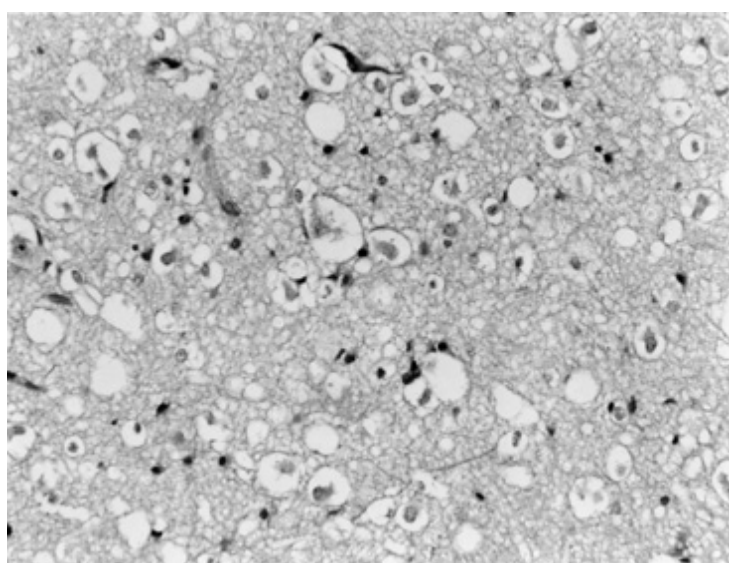
- extremely slow-growing benign **mixed glial-neuronal tumor** (neurons, astrocytes, and oligodendrocytes).

- may have germinal origin.
- patients' ages range 3-35 years (mean 21.5 yrs).

**PATHOLOGY**

- *intracortical* nodular-appearing neoplasm (features similar to *CORTICAL DYSPLASIA*) enlarging gyrus (forming *megagyrus*).
- 62% in temporal cortex, 31% in frontal cortex.
- cystic changes, frequent association with dysplastic cortex.

- well-differentiated "floating neurons" in pool of mucopolysaccharide-rich fluid and surrounding neoplastic glia without anaplastic features.



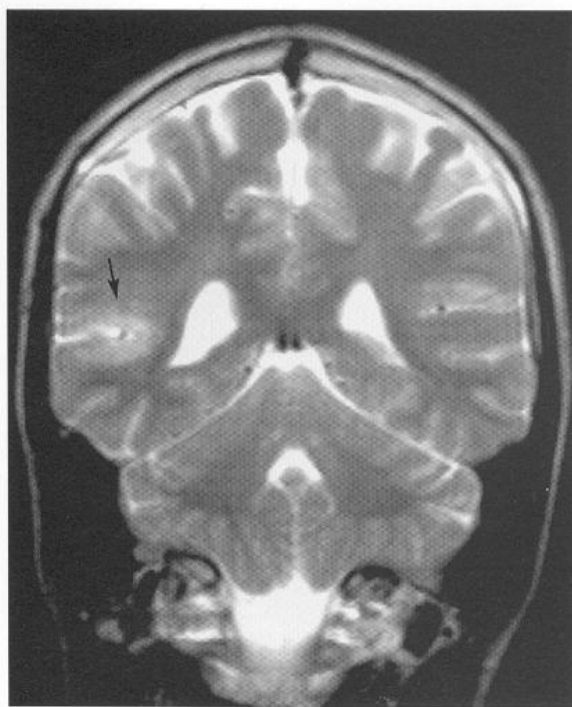
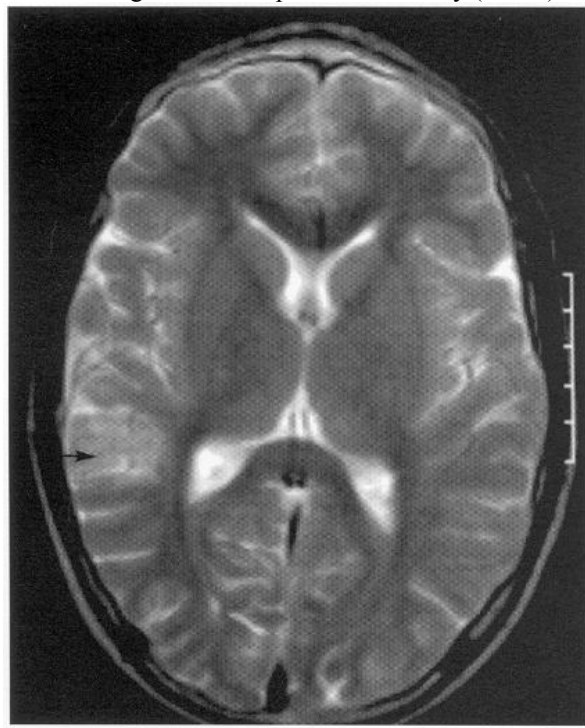
**CLINICAL FEATURES**

- often presents as **intractable partial seizures**.
- no neurological deficits (or stable congenital deficit).

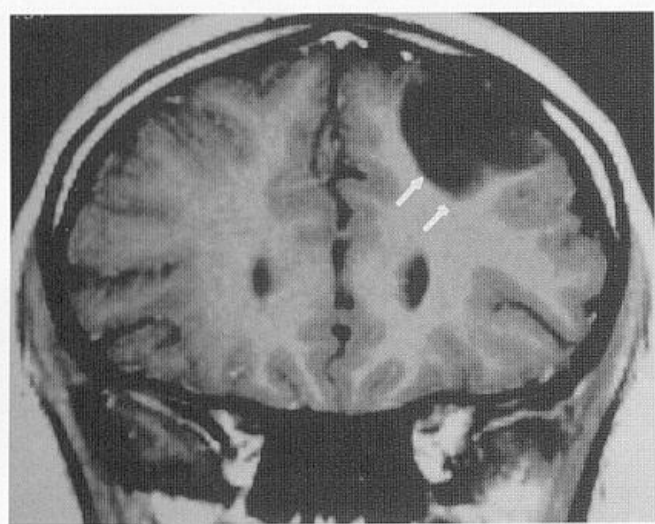
**DIAGNOSIS**

**MRI** - variable signal and enhancement characteristics ( $\approx$  *LOW-GRADE ASTROCYTOMA*).

T2-MRI - right-sided temporal abnormality (*arrow*) with thickened cortex, poorly demarcated from white matter:



T1-MRI - well-circumscribed neoplasm originating in cortical region (*arrows*); inner table of skull has been remodeled (suggesting slow-growing neoplasm):



**TREATMENT**

- good prognosis after **surgical extirpation**.
- rare postoperative complication - schizophreniform psychosis, paranoia, and depression.
- *radiation* and *chemotherapy* have no clear benefit.

**GANGLIOGLIOMA, GANGLIOCYTOMA**

- rare benign slowly growing CNS tumors:

**GANGLIOGLIOMA** (95%) - contains both **astrocytic and neuronal components**; glial component is most commonly *astrocytic*, but it may be *oligodendroglial*.

**GANGLIOCYTOMA** (5%) - **only neuronal component** without glial component.  
(its counterpart in **PNS** is **GANGLIONEUROMA**).

- 1.3% brain tumors; 1% intramedullary spinal neoplasms.

- 10% primary brain tumors in children.
- age: 2 months ÷ 70 years (most < 30 yrs).

**PATHOLOGY**

- 1) neoplastic large dysplastic/dysmorphic mature-appearing **neurons** (GANGLION CELLS), often **binucleated** (important diagnostic feature!!!); irregularly clustered; apparently random orientation of neurites.
- 2) neoplastic **astrocytes** (in **GANGLIOGLIOMA**)
- 3) relatively acellular **fibrovascular stroma**.

**DESMOPLASTIC INFANTILE GANGLIOGLIOMA** and closely related **DESMOPLASTIC INFANTILE ASTROCYTOMA**, have abundant mesenchymal component; predilection for infants and young children; good prognosis.

- **anywhere** in CNS (esp. superficial **temporal cortex**; rarely, in spinal cord).
- firm grayish tumor that may have **cystic components** and **calcification**.
- mild-to-moderately cellular; slightly pleomorphic with rare mitotic figures.
- **biologic behavior is not predicted by histology** (many anaplastic **GANGLIOGLIOMAS** do not demonstrate clinically aggressive behavior).
- metastatic spread is extremely rare (isolated report of leptomeningeal spread).
- **glial component** occasionally becomes frankly anaplastic → rapid progression (**MALIGNANT GANGLIOGLIOMA**).

**CLINICAL FEATURES**

- often presents as **intractable partial seizures**.
- most **GANGLIOGLIOMAS** are nonaggressive.
- no neurological deficits (or stable congenital deficit).

**DIAGNOSIS**

- CT** – nonspecific: hypo- or iso-dense, well circumscribed mass located superficially.
- ≈ 50% show **cystic areas** (esp. in cerebellum; single large cyst ÷ cyst with mural nodule ÷ multicystic mass)
  - ≈ 50% show **contrast enhancement** (solid tumors have more contrast enhancement).
  - punctate or fleck-like **calcification** is seen in ≈ 33-50% tumors.
  - surrounding **edema** is unusual.
  - no mass effect.

**MRI** – nonspecific.

**MR spectroscopy** – **choline-to-creatine ratio** is lower and **N-acetylaspartate-to-creatine ratio\*** is higher than in gliomas.

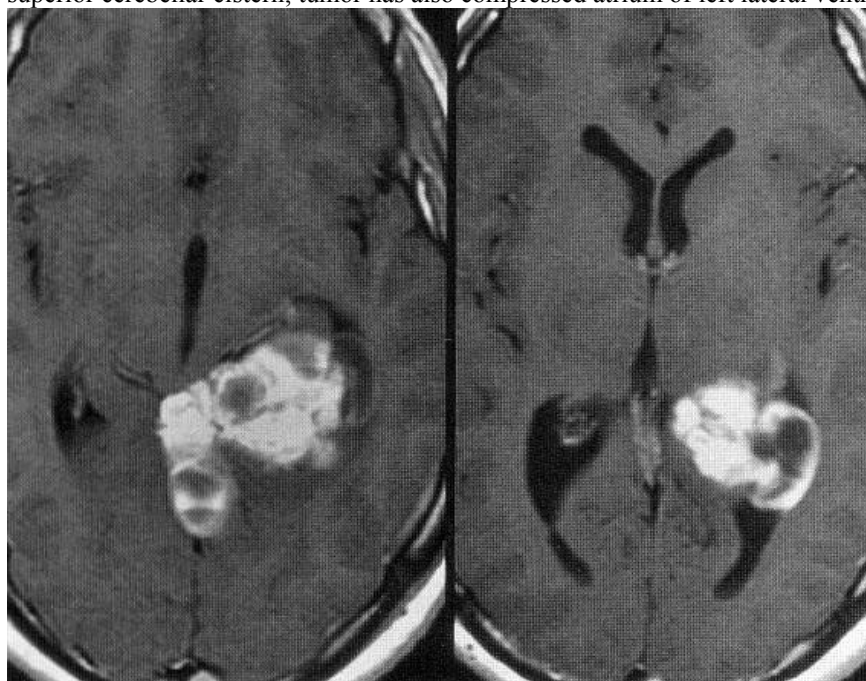
\*N-acetylaspartate↑ is due to neuronal component

Solid enhancing tumor in temporal lobe with no surrounding edema in younger patient with intractable seizures

Gadolinium-enhanced T1-MRI - enhancing tumor involving hippocampus, uncus, and amygdala:

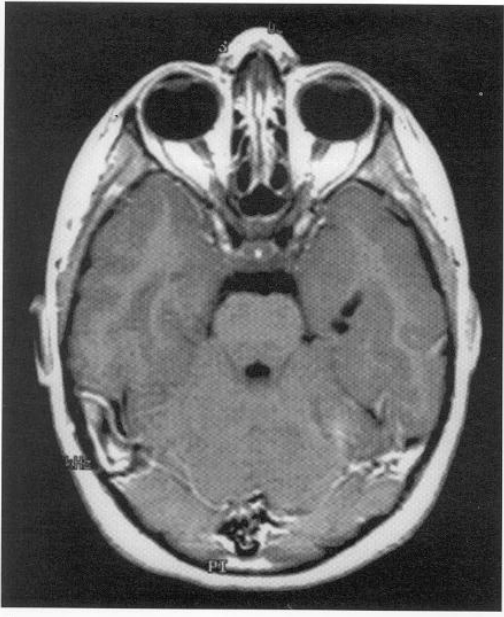


Exophytic temporal lobe ganglioglioma (T1-MRI with contrast) - large mass originating from medial aspect of left temporal lobe; both solid and cystic components; large exophytic component extends through tentorial incisura into superior cerebellar cistern; tumor has also compressed atrium of left lateral ventricle:

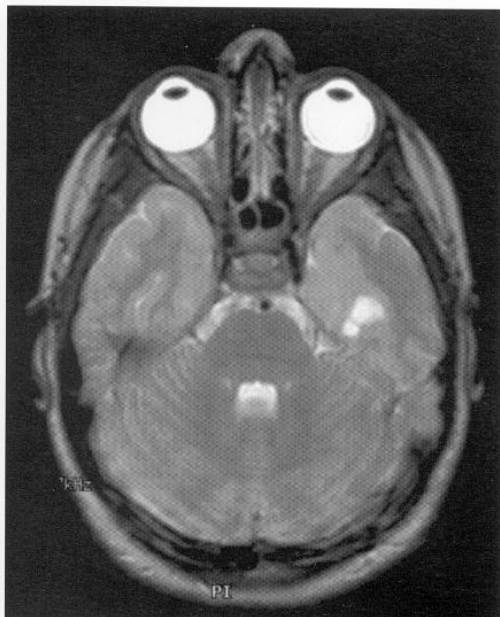


Partly cystic ganglioglioma in left temporal lobe with abnormal signal (*arrow*), without contrast enhancement:

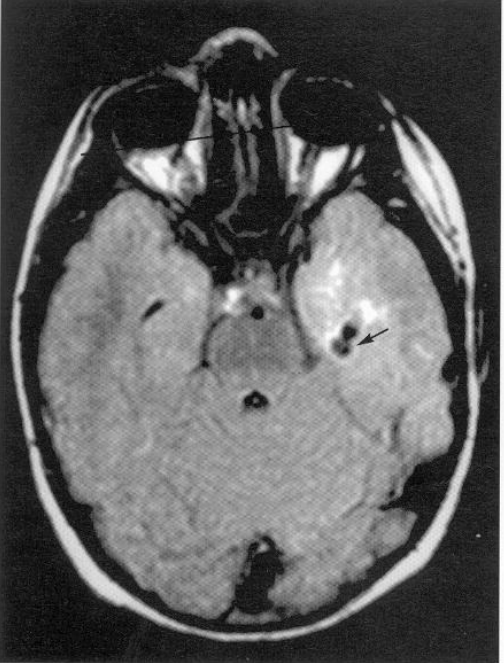
- A) Axial T1 with gadolinium.
- B) Axial FLAIR.
- C) Axial T2.
- D) Coronal T2.



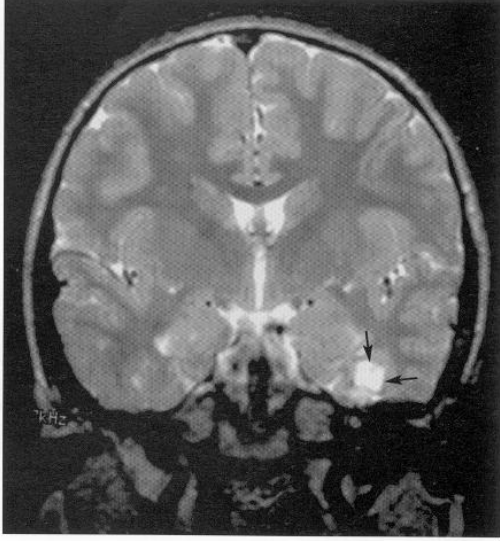
A



C



B



D

#### TREATMENT

- complete **resection** is generally curative (**radiation** is not indicated); may have good prognosis even when untreated (but incomplete removals are associated with local recurrence).
- use of **chemotherapy** has not been reported.

BIBLIOGRAPHY for ch. "Neuro-Oncology" → follow this [LINK >>](#)