Oligodendrogliomas

Last updated: August 8, 2020

Epidemiology

Most "benign" of gliomas! - never grade IV

Classification

GENETICS

1. Oligodendroglioma (WHO grade II) = 80%; median survival 6-10 yrs.
2. Anaplastic (malignant) oligodendroglioma (WHO grade III) = 15-20%; median survival 2-2.4 yrs.

Pathology

Oligodendrogliomas (grade 2)
- grossly well demarcated (but generally infiltrative); 20% are cystic.
- very cellular - monotonous side-by-side collection of homogenous, compact, rounded cells with distinct borders and clear cytoplasm surrounding dark uniform central nucleus ("chicken wire").
- microvascularity may be extensive.
- may infiltrate diffusely into cortex around normal neuronal elements (without causing loss of function) — may extend to leptomeninges.
- histology looks like oligo, but IDH-wild type - call astrocytoma!
- N.B. deletions of both 1p36 and 19q13 = greater response to chemotherapy.

Locations

- single lesion in cerebral hemispheres (white matter): frontal > parietal, temporal > occipital lobe (3:2:2:1 ratio).
- rarely, in cerebellum, brain stem, spinal cord.
- 10% tumors disseminate throughout CNS.

Genetics

- definitive diagnosis - IDH1/2 mutation + 1p19q co-deletion (assay by FISH)
  - N.B. most of low grade oligodendrogliomas are positive for IDH1 R132H mutation with intact ATRX nuclear staining.
  - if histology looks like oligo, but IDH-wild type – call astrocytoma!
  - N.B. deletions of both 1p36 and 19q13 = greater response to chemotherapy.

100% response to chemotherapy with 1p 19q LOH.

Prognosis

- 4-19% of all intracranial tumors.
- 2-25% of all gliomas (only 6% in children).
- most commonly - young and middle-aged adults (median age 25-50 yrs).

Pathogenesis

- may extend to leptomeninges.
- No conspicuous fibrillary background.
- highly characteristic - divide cells into discrete clusters - "chicken wire" capillary pattern.
- many oligodendroglialomas have some component of astrocytoma within them.
  - it is difficult to distinguish neoplastic astrocytes from reactive astrocytes.
  - some tumors are truly mixed OligoAstrocytomas (both cell types arise from common precursor - oligodendrogliocyte type-2 astrocyte, x O2A cell); minimum proportion of astrocyte is 10-25%.

Last updated: August 8, 2020

Pathology

Low-grade oligodendrogliomas (grade 2)
- grossly well demarcated (but generally infiltrative); 20% are cystic.
- very cellular - monotonous side-by-side collection of homogenous, compact, rounded cells with distinct borders and clear cytoplasm surrounding dark uniform central nucleus ("chicken wire").
- may infiltrate diffusely into cortex around normal neuronal elements (without causing loss of function) — may extend to leptomeninges.
- neoplastic cells may tightly surround neurons (perineuronal satellitosis).
- within tumor, branching blood vessels (delicate network of anastomosing capillaries) are highly characteristic - divide cells into discrete clusters - "chicken wire" capillary pattern.
- microvascularity may be extensive.
- many oligodendroglialomas have some component of astrocytoma within them.
Oligodendrogliomas

Classic "fried egg" appearance with perinuclear halos and "chicken-wire" capillary pattern:

Figure 19-38. Oligodendroglioma. Cells are round and small and have perinuclear halos. (X 100 and 200.)
Oligodendrogliomas

Atypical (malignant) oligodendroglioma (grade 3) - increased cellularity, nuclear pleomorphism, endothelial proliferation, mitotic activity, and necrosis.

- May progress to glioblastoma multiforme.

**CLINICAL FEATURES**

- Duration of symptoms before diagnosis averages 7-11 years.
- Most common (50%) presenting symptom is seizure; 80% patients have seizures at some time.
- Seizures are more common with oligodendrogliomas than other gliomas.
- Focal cerebral dysfunction; rarely ICP↑.

**DIAGNOSIS**

- CT - invisible (unless calcified*).
- MRI: Low-grade tumors - generally do not enhance (FLAIR is positive), while anaplastic oligodendroglioma does enhance; intratumoral calcification is common (∼90%).
- Definite diagnosis - biopsy (almost always possible).

Differentiate intraventricular oligodendroglioma from central neurocytoma and dysembryoplastic neuroepithelial tumor - do not need chemotherapy and radiotherapy!

A. Noncontrast CT - calcified mass in left temporal lobe (arrows), mild mass effect but little edema.
B. MR T2 - heterogeneous mass with hypointense signal (black arrows) surrounded by higher signal zone (white arrows), consistent with calcified temporal lobe mass.

T2-MRI - highly demarcated white signal; does not enhance.
Oligodendrogliaomas

Anaplastic oligodendrogloma:
A. T1-MRI - minimal heterogeneous contrast enhancement; central area of low signal intensity indicates necrosis (arrow).
B. Spin density - better delineates extent of vasogenic edema and vascular structures within and adjacent to neoplasm (arrows).
C. T2-MRI - large portions of left temporal lobe are involved by neoplastic process.

Spontaneous hemorrhage into mixed oligodendrogloma:
A. Noncontrast CT - spheroid hematoma in mass with calcification located in left parietal lobe surrounded by zone of decreased attenuation.
B. Contrast CT - enhancing tumor and relationship of hematoma.
C. Noncontrast T1-MRI - hemorrhage in tumor and surrounding edema.
D. Noncontrast T2-MRI - edema and reaction to oligodendroglioma with acute hemorrhage.

TREATMENT

No intervention = aggressive multimodal treatment for grade II guidelines see p. Onc10 >>

- anticonvulsant therapy is recommended once oligodendroglioma is diagnosed.
- some small asymptomatic (except for controlled seizures) tumors can be observed.
- surgery - mainstay of treatment (resection is usually subtotal because of infiltrative nature of tumor - surgical cure remains unlikely!);
  - total gross resection → observation for recurrence; recurrence → radiotherapy.
  - incomplete removal → radiotherapy.

ANAPLASTIC OLIGODENDROGLIOMAS (regardless of resection extent) → radiotherapy.

- use 2-3 cm margin for 54-60 Gy radiotherapy (children – 50 Gy).
- **chemotherapy** - favorable response (most chemosensitive of gliomas)!! (esp. in combined loss of 1p/19q)
  a) for recurrences  
  b) adjuvant for **anaplastic oligodendroglioma**
  standard - PCV = **PCARM + LOMUSTINE (CCNU) + VINCRI**
  for recurrences also may be tried - **TEMNOZOLOMIDE**

**100% response to chemotherapy with 1p 19q LOH.**

**LITT**

**Tumor oligo**

LITT Laser Interstitial Thermal Therapy (LITT) Treatments to Left Insular Low-Grade Glioma.

- left-sided insular oligodendroglioma treated in two stages (3 months apart - due to large 5 cm size – to prevent severe edema and seizures) with no permanent clinical deficit (temporary mild difficulty with word repetition) → chemoradiation → near resolution of the tumor at 2 yrs.

**PROGNOSIS**

- prognosis is much better than for ** Astrocytoma**!
  N.B. late progression of disease is common (5-year survival time used to indicate "cure" in other cancers is not relevant for oligodendroglionomas)
- indolent course - patients may survive for many years.
- **combined loss (co-deletion) of 1p/19q** is significant predictor of longer survival in anaplastic oligodendroglioma.
- prognosis is worse for **mixed tumors** (**OLIGOASTROCYTOMA**).

**BIBLIOGRAPHY** for ch. “Neuro-Oncology” – follow this [LINK](#)