

CNS TUMOR MAP, UPDATED WITH 2016 rev WHO DESIGNATIONS

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LEGEND: **E** = EGBs **⚡** = Enhancing
Sz = Seizures **⦿** = Cyst+mural nodule
R = Rosenthal's **C** = Calcifications



SUPRATENTORIAL

DIA / DIG
 Rx: Massive, cystic lesion with a solid, enhancing mural nodule
 Mx: Desmoplastic stroma w/ dense reticulin+ network & small cells
 Neoplastic large gemistocytic cells
 Ix: GFAP+, +/- synap, MAP2, - desmin
 Dx: mits in small cells, low Ki-67 <5%

AT/RT
 Rx: Enhancing, solid-necrotic tumors
 Mx: Classic: rhabdoid cells
 Typical: small embryonal cells
 Rare: Epithelial papillary structures
 Ix: Absence of INI1
 Mol: SMARCB1 mutations on Chr 22

EMBRYONAL TUMOR WITH MULTILAYERED ROSETTES
 Rx: Embryonal tumor with scattered multi-layered rosettes, can resemble medulloblastoma or generic PNET
 Ix: LIN28 is specific marker for this
 Mol: Chr19 amplification > C19MC mRNA

DNET
 Rx: Usu: temporal lobe, +/- calcs
 Variable enhancement
 Mx: Simple: neurons floating in mucin
 Complex: linear arrays of oligo-like cells and capillaries; multifocal distribution in cortex
 Ddx: cortical dysplasia, ganglioglioma

PXA
 Rx: Superficial/juxta-meningeal
 Temporal lobe, +/- cystic
 Mx: Lymphoplasmacytic infiltrate
 Reticulin network around cells
 Can have ganglion cells
 Ix: GFAP+, -CD34+, -synap+ cells
 Px: Anaplasia = >5/10 mit, necrosis
 An 7= Epithelioid GBM
 Mol: 50% BRAFV600E, - IDH1

ASTROCYTOMA
 Cx: Usually hemispheric, often associated with seizures
 Mx: Infiltrating neoplasm with many variants. Incr grade based on 4 factor-cellularity, mitoses, necrosis, vascular proliferation
 Ix: GFAP+, incr Ki67, IDH1/2+, TP53+

GLIOBLASTOMA
 Cx: Cerebral hemisphers, may cross midline, ring enhancing
 Mx: Highly cellular, mitotically active, pseudopalisading necrosis, vascular proliferation
 Ix: GFAP+, IDH1/2+, TP53+, Ikeratins +/-
 Mol: TP53, EGFR, Chr 10 common, BRAF in epithelioid GBM

OLIGODENDROGLIOMA
 Rx: Cortical white matter, frontal> other lobes
 Mx: Monotonous cells, calcs, capillary network
 Dx: Atypia+ some mites = II
 Mites >6/10, vasc prolif, nec = III
 Px: Good when <5% Ki-67
 Mol: p19q co-del is required; IDH1/2-

GANGLIOGLIOMA
 Rx: Usu: temporal lobe, cystic with mural nodule, +/- enhancement
 Mx: Neuroglial tumor, bi-nucleate neurons, perivascular lymphs
 Anaplasia = malignant glioma
 Ix: CD34 in neurons (70%)
 Mol: BRAFV600E 50%

ANGIOCENTRIC GLIOMA
 Rx: Superficial, cortical
 Non-enhancing, well delineated
 Mx: Spindle cells with perivascular condensation, no atypia
 May mimic prominent endothelium
 Ix: GFAP+, S100, "dot-like" EMA

ASTROBLASTOMA
 Rx: Hemispheric, cystic and lobular
 Contrast enhancing
 Mx: Sharp margin, rosettes with thick processes; vascular hyalinization
 Well-diff: Mites <5, Ki67<3%, -necrosis
 Poor-diff: Mites >5, Ki67>3, +necrosis

PAPILLARY GNT
 Rx: Well demarcated, +/- cystic
 Contrast enhancing
 Mx: Pseudopapillae with hyalinized vessel = inner neuronal cells and outer clear cells
 Ix: Inner = GFAP, Outer = Synap
 Ki-67 <2%
 Dx: PA, extraventricular neurocytoma



INFRATENTORIAL

PILOCYTIC ASTROCYTOMA
 Rx: Cystic, enhancing, +/- calcs
 Mx: Bi-phasic, microcystic, +/- pleo, vasc prolif, nec
 Multi-nucleated cells are typical
 Limited infiltration or leptomeningeal spread is common
 Ix: Almost always <2% Ki-67

MEDULLOBLASTOMA
 Rx: Midline cerebellar / 4th vent
 Mx: Histology loosely correlates with genetics
 SHH - Infants, Desmo/MBEN, +MYC, d9q
 WNT - Children, classic +MYC, mono 6
 Gr3 - Children, classic/LCA, +MYC, mets
 Gr4 - Child/Adol, classic/LCA, 117q

EPENDYMOMA (&VARIANTS)
 Rx: 4th vent + S.C. > 3rd/lateral vent
 variable enhancement, calcs
 Mx: Gr II: Sharp circumscription, rosettes +/- non-palisading necrosis
 Anaplasia (grade III) = >5 mites + palisading nec + high cellularity
 GFAP+, S100, dot-like EMA
 Mol: Chromosome instability (CIN)
 Pediatric: CIN -, group A, worse
 Adol/Adult: CIN+, group B, better

DYSPLASTIC GANGLIOCYTOMA
 Rx: aka - Lhermitte-Duclos, usu adults
 can present congenitally
 Rx: non-enhancing, hemispheric enlargement
 Mx: Dysplastic ganglion cells in the granular layer of cerebellar foia

HEMANGIOBLASTOMA
 Rx: VHL assoc = multiple & infratentorial
 Mx: Large, vacuolated stroma + vessels
 Gliosis at periphery of lesions
 Nuclear atypia common, EMH present
 Ix: Stroma = S100, -GFAP, inhibin +
 D240+, CD10- (unlike RCC), EMA-
 Dx: RCC (esp. with VHL), CD10+

LIPONEUROCYTOMA
 Rx: Cerebellar hemispheres, heterogeneous enhancement
 Mx: Biphasic: neurocytes + fat
 Ix: Synap+, MAP2+ (both cell types)
 Px: Usu <5% Ki-67, can recur

CHOROID PLEXUS CARCINOMA
 Rx: More often seen in younger patients
 Cx: Lateral vent > 3rd vent
 Mx: Frankly malignant, invasive tumor + solid sheets, rhabdoid cells
 Ix: Kerat+, GFAP +/-, EMA, INI retained
 Ddx: Papillary ependymoma, papillary meningioma

CHOROID PLEXUS PAPILLOMAS
 Rx: Lateral vent > 3rd vent (kids)
 4th vent (adol and adults)
 Mx: Gr I: low mites and focal atypia
 Gr II: (atypical CPP) mites >2/10 + complex archt
 Ix: Ker+, TTR+, S100+, -GFAP in 50%

PAPILLARY PINEAL REGION TUMOR
 Rx: Posterior 3rd vent, hydrocephalus
 Mx: Epithelioid papillary tumor with eosinophilic clear columnar cells
 Ependymoma-like in areas
 Ix: Keratin +, focal GFAP, TTR
 Dx: Ependymoma (diffuse GFAP), TTR

CENTRAL NEUROCYTOMA
 Rx: Lateral & 3rd vent >> cortex/S.C
 Enhances, can be multifocal
 Mx: Round cells in neuropil, +/- calcs
 Nuclear free zones around vessels
 Hyalinized vessels
 Ix: Synap+, NeuN, GFAP-
 low Ki-67, <2%

SUBEPENDYMOMA
 Rx: Non-enhancing, nodules,
 4th vent > lateral vent > 3rd vent
 Mx: Lobular clusters of round nuclei + fibrillary glial matrix
 +/- vasc prolif, +/- "rosettes"
 Px: <1% Ki-67

CHOROID GLIOMA OF THE 3RD VENTRICLE
 Rx: Adult tumor, enhancing, bulky
 Mx: Cords of epithelioid glial cells
 mucoid matrix, +/- lymphocytes
 Ix: GFAP+, S100-, low Ki67 <2%
ROSETTE FORMING GNT OF 4TH VENTRICLE
 Mx: Pilocytic-like glial tumor + Synap+ neurocyte rosettes



BRAINSTEM & CORD

PILOMYXOID ASTROCYTOMA
 Rx: Circumscribed, enhancing
 Hypothal/chiasm common
 Mx: Bipolar cells + mucoid matrix
 Angiocentric arrangement
 Mitoses & vasc prolif are seen
 Ix: GFAP+, S100+, Ki67 2-20%
 Dx: More aggressive than PA

DIFFUSE MIDLINE GLIOMA WITH HISTONE K27M
 Cx: Midbrain, thalamus, pons + cerebellum
 Mx: Pilo-to-embryonal spectrum of morphology, Epithelioid, spindle etc...
 Ix: Histone K27M, GFAP+, TP53-
 Mol: No IDH1/2 or EGFR aberrancies
 Rarely can have BRAF mutations

MYXOPAPILLARY EPENDYMOMA
 Rx: Conus/Flum, multifocal
 Rarely can present in sacral soft tissues
 Mx: Radial arrangement around vasc cores; abundant myxoid matrix
 Ix: GFAP+, S100+, Ker-

TANYCYTIC EPENDYMOMA
 Cx: Often C or T spine
 Mx: spindle cells, no rosettes
 Mol: Chrom. instability often seen with Gr III tumors

0 - 12 mo

1 - 10 years

11 - 25 years

>25 years