CNS TUMOR MAP, 2023 WHO-5 REVISION WITH MOLECULAR INTEGRATION

LEGEND

- E = EGBs
- R = Rosenthals
- = Seizures
- = Perivascular pattern
- = Enhancing
- = Cyst+mural nodule
- = Calcifications
- *b* = BRAF alteration
- *i* = IDH1/2 mutation
- *o* = 1p/19q co-del
- = Low grade
- = Grade III
- = Grade IV
- = Not elsewhere classifed

* Tumor summaries below may not necessarily state the formal WHO preferred terminology, in the name of brevity.

Not all CNS tumors are described here. Specifically, this chart will not address meningiomas, CNS lymphomas or mesenchymal tumors.

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Ref: WHO Tumors of the CNS (5th ed, 2023)

SUPRATENTORIAL

MIDLINE

INFRATENTORIAL

BRAINSTEM & CORD

VENTRICLES

INFANT HIGH-GRADE GLIOMA
MX: Usually embryonal "PNET-like" histology. Sometimes papillary, rosetted or spindled growth patterns. Occasionally lower-grade glial pattern.
IHC: POS (if mutated): ALK1, panNTRK, ROS1 NEG: retained INI1/BRG1
MOL: HGG pattern: ALK / ROS / NTRK / MET alterations LGG pattern: ALK alterations only
DDX: Usually don't have H3K27M or BRAF mutations

ANGIOCENTRIC GLIOMA
I
MX: Circumscribed, spindle cell tumor with elongated cells radially arranged around vessels - akin to ependymomatous rosettes
IHC: POS: GFAP, **MYB**, EMA (dot-like), D2-40 NEG:Ki67 (<5%), Synap, p53, IDH1, Olig2
MOL: MYB (6q23) rearrangement, usu MYB::QKI
DDX: Ependymoma, pilomyxoid astrocytoma, astroblastoma

ASTROBLASTOMA, MN1 ALTERED
MX: Well defined tumor arranged in radial "rosettes" with broad bases. **Stromal sclerosis**. +- papillae and necrosis
IHC: POS: GFAP (var), S100, EMA (var), Olig2 (usu) NEG: IDH1, BRAF V600E
MOL: Rearrangements involving MN1 (22q12)
DDX: ST-EPN,

PLEOMORPHIC XANTHOASTROCYTOMA
II III
MX: Cellular/pleomorpic,, sometimes lipidized with multinucleation & nuclear inclusions. Frequent perivascular lymphocytes and **reticulin**.
IHC: POS: GFAP, BRAF V600E, MAP2 (var), CD34 (var) NEG: **p16 null** (usually), IDH1
MOL: BRAF mut (80%), CDKN2A del (60%)
DDX: Grade 3 = >5 mitoses & necrosis | Epithelioid GBM

GBM, GIANT CELL
MX: Glioma with many large/giant cells. Increased reticulin w/ mesenchymal look. Zonal necrosis and +/- MV proliferation. Often in younger patients
IHC: POS: S100, GFAP (patchy), EMA/CK (focal) NEG: H3, retained INI1/BRG1, **MMR loss**
MOL: **IDH WT,H3 WT**. Pedi cases usu. BRAF V600e neg High mutation burden + MMR gene mutations
DDX: Related to anaplastic PXA or may arise from Gr2PXA

ASTROCYTOMA (IDH MUT)
MX: Infiltrative glial tumor with: hypercellularity, mitoses, and vascular proliferation. **Less** pronounced **necrosis.** Often arising from lower grade glioma
IHC: POS: GFAP, IDH1 R132H, p53 NEG: H3K27M, ATRX, p16, IDH1 (if alternative mutation)
MOL: **IDH mutated & CDKN2A/B homozy. deletion** Additionally: TP53 mut, ATRX mut, MGMT hypermeth
DDX: IDH-wt GBM, IDH-mut anaplastic astro

GLIOBLASTOMA (IDH WT)
MX: Infiltrative glial tumor with: hypercellularity, mitoses, **pseudopalisading necrosis** and vascular proliferation. Small cell var = minimal atypia
IHC: POS: GFAP, IDH1 R132H, p53, S100, **focal** INI-1 loss NEG: H3K27M, CK5/6
MOL: **IDH WT & H3 WT,** TERT promoter mut., EGFR alterations (esp in small cell var.), TP53 mut
DDX: Epithelioid GBM, Oligodendroglioma (small cell var)

PEDIATRIC LOW-GRADE GLIOMA, ALK FUSION
MX: Glial or glio-neuronal tumor with mod. cellularity and mild atypia. High-grade versions also exist.
IHC: POS: GFAP, Alk, NEG: Ki67 (very low)
MOL: Alk-fusions: PPP1CB-Alk and rare other partners
DDX: PLNTY, Gangliogliooma

DNET
I
MX: Glioneuronal tumor with oligo-like cells,, mucoid microcysts and "floating" neurons. +/- glioma areas & separate FCD (IIIa)
IHC: **Oligo-like**: POS: S100, OLIG2, CD34 (focal) NEG: GFAP (in oligo-like), IDH1
MOL: BRAF mut (30%), Chr 5 and 7 gains (30%)
DDX: Pilocytic astro, oligodendroglioma, gangliogliooma

PLNTY
I
MX: Glial tumor with piloid cells, set among prominent vasculature and heavy calcification
IHC: POS: GFAP, CD34 (strong), +/- BRAF NEG: Cga, IDH1, Synap (mostly) , Ki67 usu low
MOL: FGFR2-CTNNA3 fusions, BRAF V600E, FGFR1 alterations
DDX: Pilocytic astrocytoma, DNET, gangliogliooma

PAPILLARY GLIONEURONAL TUMOR
I
MX: Low-grade biphasic glioneuronal tumor arranged in **pseudopapillae** surrounding **hyalinized vessels.** Intervening ganglionic cells with neuropil.
IHC: POS: GFAP and S100 (glial); OLIG2 (var), Synap NEG: CgA
MOL: SLC44A1-PRKCA fusion in most
DDX: Ependymoma, AVM

GBM, EPITHELIOID
MX: PXA-like glioma with large/giant cells. Zonal necrosis and +/- MV proliferation. Often in younger patients
IHC: POS: S100, GFAP (patchy), EMA/CK (focal), BRAF NEG: H3, retained INI1/BRG1, **P16 null**
MOL: **IDH WT,H3 WT**, BRAF V600E (50%), **CDKN2a del** High mutation burden + MMR gene mutations
DDX: ? related to PXA or may arise from Gr2PXA

ASTROCYTOMA (IDH MUT)
II III
MX: Cellular astrocytic, fibrilar neoplasm with mild to moderate nuclear atypia, angulated nuclei + hyperchromasia. No MVP or necrosis allowed.
IHC: POS: GFAP, TP53, IDHR132H (80%) NEG: ATRX (loss)
MOL: (80%) IDH1/2 mut & TP53 mut + **CDKN2A/B retained,** MGMT promoter methylation (50%
DDX: Grade 3 = Hypercellular + increased mitoses

EXTRAVENTRICULAR NEUROCYTOMA
II
MX: Round cell neurocytic tumor with prominent intralesional vessels and pseudorosettes. Oligo-like with occasional ganglioid differentiation.
IHC: POS: Synap NEG: Cga, IDH1, GFAP (mostly) , Ki67 usu low
MOL: Rarely co-del 1p/19q, FGFR1-TACC1 fusions Never IDH1/2 mutation or BRAF V600E mutation
DDX: Oligodendroglioma, DNET, gangliogliooma, PGNT

DESMOPLASTIC INFANTILE ASTRO/GANGLIO
I
MX: Triphasic tumor: glial cyst wall, desmoplastic embryonal mural nodule +/- ganglion cell component.
IHC: POS GFAP, reticulin fibres, Synap (neurons), desmin (rare)
MOL: Rare aneuploidy. Rare BRAF V600E mutations. No BRAF-fusions or TP53 mutations
DDX: Embryonal tumors, gangliogliooma, **PXA**

DICER1-ASSOCIATED CNS SARCOMA
MX: High-grade spindle cell neoplasm with PPB-like pattern, eosinophilic globules and - rhabdo cells.
IHC: POS: Desmin (focal), nyogenin (focal) NEG: GFAP, Olig2, Synap, INI1 (retained)
MOL: Bi-allelic DICER1 mutations
DDX: Embryonal tumors, GBM, AT/RT

DIFFUSE GNT W/ OLIGO-LIKE FEATURES AND NUCLEAR CLUSTERS
MX: **Cellular**, infiltrative oligo-like morphology with scattered "pennies-on-plate" clusters of pleomorphic nuclei
IHC: POS: OLIG2 (tumor cells), Synap (background) NEG: GFAP
MOL: Monosomy 14 is consistently present
DDX: DNET, diffuse pediatric astrocytoma, FGFR1 mutated

MULTINODULAR AND VACUOLATING GNT
MX: Temporal lobe >frontal lobes. **Nodular** arrangement of neurons with nucleoli and pericellular vacuoles. Multinucleated neurons are **absent**
IHC: POS: OLIG2, MAP2, weak Synapto NEG: Chromogranin, NeuN, GFAP
MOL: MAPK mutations, BRAF mut (**not V600E**), FGFR2 fusions
DDX: DNET, Gangliogliooma

GANGLIOGLIOMA
I III
MX: Discorganized, variably cellular lesion with glial and neuronal component. Look for binucleated and dysplastic neurons. Perivascular **lymphocytes**.
IHC: POS (Neuron): MAP2, Synap, BRAF V600E, CD34 **Rarely: H3K27M** in non-infiltrative, temporal cases
MOL: BRAF V600E (~50%), BRAF fusions (rarely) **NO IDH1/2** mutations (excludes this diagnosis)
DDX: DNET, Oligodendroglioma, co-existing FCD (type IIIb)

DIFFUSE LOW GR. GLIOMA, MAPK PATHWAY ALTERED
MX: Bland, infiltrative glial lesion, some with oligoo like morphology (FGFR1 mutated) or pilocytic morphology (BRAF mutated)
IHC: POS: GFAP, p16 (not null), OLIG2
MOL: MAPK pathway mut: BRAF, FGFR1, FGFR2, NTRK, MET **No CDKN2A/B deletion**
DDX: Pilocytic astro, PLNTY, DNET, High grade astro with piloid features if CDKN2A/B deleted

DIFFUSE ASTROCYTOMA, MYB ALTERED
MX: Bland, minimally cellular round glial cells with entrapped neurons and vague perivascular arrangement.
IHC: POS: GFAP, ATRX NEG: OLIG2, IDH1, CD34
MOL: MYB or MYBL1 fusions (but usu not MYB-QKI)
DDX: Angiocentric glioma, Diffuse astrocytomas

OLIGODENDROGLIOMA
II III
MX: Infiltrating gliomas with round, "fried-egg" cells in delicate chicken-wire vasculature background. **Grade 3** = Incr. mitoses + Microvasc prolif + necrosis
IHC: POS: IDH1 (90%), ATRX (retained), S100, OLIG2 NEG: GFAP (mostly)
MOL: IDH1/2 mut + co-del 1p/19q | Often: CIC & TERT mut. NEG: mutations in ATRX or TP53
DDX: Neurocytoma, clear cell ependymoma, DNET, Pilocytic

INFANT-TYPE HEMISPHERIC GLIOMA, GENETICALLY DEFINED
Many genetically defined embryonal tumors look similar to one another:
1. CNS NB-FOXR2: Resembles CNS neuroblastoma, ganglionic nodules
2. CNS HGNET-MN1: Solid + pseudopapillary tumor, resembles astroblastoma
3. CNS HGNET-EFT-CIC: Can resemble EWS
4. CNS HGNET-BCOR: Glial looking w/ rosettes or ependymoma-like
5. CNS HGG-ALK/ROS/NTRK
IHC: CNS NB-FOXR2: Olig2, Synaptophysin, GFAP; CNS HGNET-MN1: GFAP; CNS HGNET-EFT-CIC: NUT1; CNS HGNET-BCOR: GFAP, B-cat (nuc), BCOR; CNS HGG-ALK/ROS/NTRK: ALK1, ROS1, NTRK
MOL: CNS NB-FOXR2: Intrachromosomal rearrangement, FOXR2 upreg.; CNS HGNET-MN1: Various fusion partners; CNS HGNET-EFT-CIC: CIC-NUTM1 fusion; CNS HGNET-BCOR: BCOR ITD exon 15; CNS HGG-ALK/ROS/NTRK: Various fusion partners across all

DIFFUSE HEMISPHERIC GLIOMA, G34R MUT
MX: Embryonal or GBM-like patterns
DX: H3 G34 mut, ATRX loss

Now often H3G34R mut

ATYPICAL TERATOID/RHABDOID TUMOR
MX: Supratentorial > infratentorial & midline Polyphenotypic, hypercellular embryonal tumor with rhabdoid and ocasional anaplastic cells.
IHC: POS: INI1 or BRG1 (aberrant loss), GFAP (focal)
MOL: SMARCB1 > SMARCA4 alterations. Mut >> deletions, 33% germline
DDX: All embryonal tumors in children

EMBRYONAL TUMOR WITH MULTILAYERED ROSETTES
MX: Cerebellar > Midline/Posterior fossa. Embryonal tumor with layered rosettes and islands of nucleus free neuropil
IHC: POS: LIN28 (strong & diffuse), Synap (neuropil) NEG: INI1 (retained), GFAP
MOL: C19MC-altered
DDX: Embryonal tumors, anaplastic ependymoma, CPC

pHGG, H3 and IDH WT
MX: Supratentorial >> Brainstem. 3 types: RTK1, RTK2, MYCN type (often biphasic: infiltrative + embryonal nodules)
IHC: POS: GFAP+/-, Neuronal markers+/-, Olig2 NEG: H3 K27m, IDH1/2
MOL: **IDH & H3 wt**. Intact CKDN2A/B. PDGFRA(RTK1 type), EGFR (RTK2 type), MYCN (MYCN type)
DDX: All embryonal tumors in children

EPENDYMOMA (ST)
II III
MX: Can be papillary or clear cell morphology. Clear cell = arranged in cellular groups with perinuclear halos, focal perivascular rosettes.
IHC: SP-YAP1: GFAP, S100, EMA (dot-like), **no LICAM** SP-ZFTA: Cyclin-D1, **LICAM, p16 null**
MOL: ST-EPN-ZFTA: ZFTA-RELA fusion, Chromothripsis ST-EPN-YAP1: YAP1 fusions
DDX: Oligodendroglioma, neurocytoma, hemangioblastoma

PINEOCYTOMA
I
MX: Moderately cellular round cell tumor growing in sheets and multilayered (pineocytomatous) neuropil rosettes, set among rich vasculature.
IHC: POS: Synap, NF, MAP2 (var), CgA (var) NEG: GFAP, S100
MOL: No relevant diagnostic molecular genetic profile
DDX: PPTID, EV neurocytoma, Germinoma

PINEAL PARENCHYMAL TUMOR OF INTERMEDIATE DIFFERENTIATION
? II ? III
MX: Two patterns: Diffuse neurocytoma-like and lobulated/nested pattern w/ distinct vessels. More cellular and occasional dysplastic ganglioid cells.
IHC: POS: Synap, NF, CgA (var) NEG: GFAP, S100, NeuN
MOL: aCGH: gains of 4q, 12q and loss of 22
DDX: Pineoblastoma, Germinoma,

CRIBRIFORM NEUROEPITHELIAL TUMOR
MX: **Strands and ribbons**, pseudo-rosettes. **No rhabdoid cells**
IHC: POS:EMA, **Tyrosinase**, GFAP (var) NEG: **INI1 loss**
MOL: SMARCB1 loss (22q)
DDX: ATRT, malignant chordoma?

DIFFUSE MIDLINE HIGH-GRADE GLIOMA, H3K27M
MX: Infiltrative tumor involving midline nuclei or brainstem. Monomorphic tumor cells with variable morphology resembling pilocytic astro to GBM.
IHC: POS: GFAP (var), H3K27M, OLIG2, MAP2 NEG: retained INI1/BRG1, CGA, ATRX, OLIG2
MOL: H3K27M mutation (midline), H3G34R (hemispheric) TP53 (50%), PDGFRA amp.
DDX: Related to anaplastic PXA or may arise from Gr2PXA

PINEOBLASTOMA
MX: Embryonal, hypercellular tumor of pineal region with focal rosette formation. Often invasive and disseminated.
IHC: POS: Synap (var), NF (focal), CgA (focal) NEG: INI1 & BRG1 (retained)
MOL: RB1 deletion, DICER1 mutation
DDX: All embryonal tumors of childhood, AT/RT

PAPILLARY TUMOR OF PINEAL REGION
? II ? III
MX: Biphasic solid and papillary tumor with ependymoma-like pattern, hyalinized vessels and occasional PAS+ cytoplasmic globules
IHC: POS: Keratins (AE1/3, CAM5/2, CK18), GFAP (var) NEG: NF, Synap (focal), CgA (focal)
MOL: aCGH: del 10, + 4. PTEN alteration occasional
DDX: Ependymoma, Germ cell tumor, Metastasis

PILOMYXOID ASTROCYTOMA VARIANT OF PILOCYTIC ASTROCYTOMA
MX: Piloid astrocytic tumor with subtle angiocentric growth, myxoid background & microcysts. Variable mitotic activity. +/- Pilocytic-like areas
IHC: POS: GFAP, S100, ~CD34, Ki67 (up to 20% labeling) NEG: BRAF V600E, H3K27M
MOL: Rarely can have BRAF rearrangement
DDX: Pilocytic astrocytoma, Angiocentric glioma

MEDULLOBLASTOMA
MX: Embryonal tumor with variable nodules of neuronal differentiation. Cerebellum and 4th ventricle. Anaplasia usu = TP53 mutation
IHC: POS: Synap, MAP2, p53, B-Cat (WNT),, GAB1 (SHH) NEG: GFAP, INI1 (retained)
MOL: 4 molecular groups: WNT - Older children, some adults. SHH-TP53 wt - Often hemispheric & Desmoplastic/EN. SHH-TP53 mut - TP53 mut confers worse prognosis G3/4 - Infants/children. Large/anaplasia & MYC amp

PILOCYTIC ASTROCYTOMA
I
MX: Astrocytic tumor with elongated processes, biphasic density, microcysts and occasional multinucleation. Low mitoses. +/- Vasc prolif, Leptomening. spread
IHC: POS: GFAP, ~OLIG2, BRAF (hemispheric) NEG: p53, IDH1/2
MOL: Posterior fossa: KIAA1549-BRAF fusion Cortex/Midline: BRAF mut, FGFR1 (5%), NTRK (~5%)
DDX: Pilomyxoid (no EGBs/ Rs), Oligo., DNET

EPENDYMOMA (PF)
II III
MX: Monomorphic glioma arranged in rosettes with perivascular anuclear zones. Can have dense cellularity, focal necrosis and hemorrhage
IHC: POS: GFAP, S100, EMA (dot-like) ***H3K27me3:*** Lost in EPN-A, Retained in EPN-B
MOL: PF-EPN-A: Few copy # changes, Cpg-me + PF-EPN-B:Chromosomal instability, Cpg-me -
DDX: Choroid plexus tumor, Medulloblastoma, Metastasis

DYSPLASTIC CEREBELLAR GANGLIOCYTOMA
?I
MX: Expansion of molecular and internal granule layers with variably sized ganglionic cells that preserves overall architecture.
IHC: POS: Syna NEG: PTEN (loss in adult cases)
MOL: Adult: PTEN mutations (Cowden syndrome) Children: no PTEN mutations
DDX: Gangliogliooma, Glioneuronal tumors

HIGH GRADE ASTROCYTOMA WITH PILOID FEATURES
MX: Cellular, moderately pleomorphic infiltrative tumor with pilocytic morphology, vascular proliferation and focal areas of necrosis.
IHC: POS: GFAP NEG: ATRX (loss), IDH1, H3K27M
MOL: Defined by methylation studies. Characteristic: NF1/BRAF alterations + del ATRX + del CDKN2A
DDX: Pilocytic astrocytoma, IDH-wt GBM

CEREBELLAR LIPONEUROCYTOMA
II
MX: Cerebellar version of neurocytoma with prominent neoplastic adipocyte-like component.
IHC: POS: Synap, NeuN, MAP2, GFAP (focal) NEG: Ki67 (usu <10%)
MOL: TP53 mutation (20%), Chr 14 and 2p deletions No BRAF or IDH mutations
DDX: Astrocytomas, lipid-rich SFT?

MYXOPAPILLARY EPENDYMOMA
?II
MX: Typically found in distal spinal cord. Radially arranged tumor cells in papillary / balloon arrangement around myxoid substance.
IHC: POS: GFAP, S100, CD99, Ker AE1/3 NEG: CK5/6, CK7, CK20, EMA, Ki67 (<1%)
MOL: Whole chromosome aneuploidy **MYCN amplification** (more aggressive variant)
DDX: Metastatic papillary tumors (adult), Chordoma

EPENDYMOMA (SPINAL CORD)
II III
MX: Usually typical perivascular pattern. Rarely can by Tanycytic resembling spindle cells where rosettes typically subtle.
IHC: POS: FOXJ1, GFAP, S100, EMA (dot-like) MYCN+ in rare subtype **(no H3K27m)**
MOL: Frequent NF2 mutations, del Chr 22
DDX: Pilocytic astrocytoma, Schwannoma, Met.

CHOROID PLEXUS CARCINOMA
MX: Malignant intraventricular tumor w/ sheet-like growth, focal papillary formation, necrosis and brain invasion. Usu freq. Mitoses.
IHC: POS: CK7, p53 (50%) NEG: S100, TTR, EMA, INI1 (Retained)
MOL: Germline TP53 mut (40%)
DDX: Anaplastic ependymoma, Embryonal tumors

CHOROID PLEXUS TUMOR
I II
MX: Papillary tumor with delicate fronds with crowded cuboidal cells. **Atypical** = >2/10 mits + incr. cellularity, pleomorph., solid growth and/or necrosis.
IHC: POS: CK7, TTR, S100 (var) NEG: CK20, EMA (weak)
MOL: aCGH: hyperdiploidy. MGMT promoter methylation
DDX: CP carcinoma, Endolymphatic sac tumor, Metastasis

SUBEPENDYMAL GIANT CELL TUMOR
I
MX: Circumscribed glio-neuronal tumor with large gemistocyic or ganglion-type cells. Mitoses, tumor lymphocytes and hyalinized vessels present.
IHC: POS: GFAP, S100, Synap (var), NeuN (var) NEG: CD34
MOL: TSC1 and TSC2 mutations common 60% sporadic, 40% TS syndromic
DDX: Gangliogliooma, PXA (if not obviously near ventricle)

MYXOID GLIONEURONAL TUMOR
I
MX: DNET-like tumor arising in septum pellucidum > lateral ventricles. Myxoid stroma, microcysts and rosette-formations.
IHC: POS: OLIG2, MAP2, S100, GFAP (focal) NEG: CD34, NeuN, IDH1
MOL: Defined by PDGFRA K385I/L mutation

DIFFUSE LEPTOMENINGEAL GNT
I ?II
MX: Oligodendroglial-like tumor with predominant leptomeningeal growth pattern and lesser ganglion cell / neuropil component
IHC: POS: OLIG2, MAP2, S100, GFAP (focally) NEG: EMA, NeuN, IDH1
MOL: KIAA-BRAF fusion (75%), del 1p (50%), rare 1p/19q del. No BRAF V600E or IDH1/2.
DDX: Oligodendroglioma, Pilocytic astro, PXA

ROSETTE FORMING GNT
I
MX: Biphasic solid-cystic, tumor: neurocytic rosettes and piloid astrocytic components. Neurocytic rosettes surround neuropil core.
IHC: POS: Neurocytic = Synap, MAP2: Glial = GFAP NEG: BRAF V600E, IDH1/2
MOL: FGFR1 mut, PIK3CA mut in some No BRAF alterations or IDH1/2 mutations
DDX: Pilocytic astro, neurocytoma, Oligodendroglioma

CENTRAL NEUROCYTOMA
II
MX: Intraventricular round cell neurocytic tumor with prominent intralesional vessels and pseudorosettes. Anaplastic cytology = "atypical central neurocytoma".
IHC: POS: Synap, NeuN, MAP2 NEG: Cga, GFAP, Ki67 usu <2%. If >2% = "atypical"
MOL: aCGH: copy # alterations, MYCN amplification
DDX: Ependymoma, Pineocytoma

SUBEPENDYMOMA
I
MX: Ventricular tumor. Clusters of small nuclei arranged in fibrillar matrix with occasional microcysts. Rarely forming rosettes.
IHC: POS: GFAP NEG: EMA, Ki67 (<1%)
MOL: Not really relevant
DDX: Ependymoma variants

CHORDOID GLIOMA OF 3RD VENTRICLE
II
MX: Solid neoplasm w/ cords and nests of epithelioid tumor cells. Lymphoplasmacytic infiltrates. Mucinous stroma common. Rarely fibrotic.
IHC: POS: GFAP, TTF-1, CD34, Ker (var), S100 (var) NEG: PS3 (weak), Synapto, IDH1
MOL: PRKCA pD463H mutation IDH, H3, wild-type
DDX: Metastasis, Chordoma.

0 - 24 months

2- 10 years

10-50 years

> 50 years