Central Nervous System embryonal tumor

- Applications of Advanced Imaging for Radiotherapy Planning and Response Assessment in the Central Nervous System
- Growth Dynamics of Brain Tumor Through the Lens of MT-Weighted MRI
- Anatomo-functional approach to multimodal motor mapping in diffuse glioma surgery: hierarchical networks
- An Osmosis-driven 3D-printed brain implant for drug delivery
- Years of life lost due to central nervous system tumor subtypes in the United States
- Tumor Microenvironment in Melanoma Brain Metastasis: A New Potential Target?
- PIK3CA Mutations: Are They a Relevant Target in Adult Diffuse Gliomas?
- Patterns, clinical presentations, and time to diagnosis in pediatric central nervous system tumors: insights from a pediatric neuro-oncology tumor board team at a tertiary referral hospital in Ethiopia

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Central Nervous System (CNS) embryonal tumors represent a heterogeneous group of highly aggressive tumors occurring preferentially in children but also described in adolescents and adults. The World Health Organization Classification of Tumors of the Central Nervous System 2021, drastically changed the diagnosis of other CNS embryonal tumors including new histo-molecular tumor types.

Medulloblastoma

Medulloblastomas, molecularly defined

Medulloblastoma, WNT-activated

Medulloblastoma, SHH-activated and TP53-wildtype

Medulloblastoma, SHH-activated and TP53-mutant

Medulloblastoma non-WNT/non-SSH

Medulloblastomas, histologically defined

Other CNS embryonal tumors

Atypical teratoid/rhabdoid tumor

Cribriform neuroepithelial tumor

Embryonal tumor with multilayered rosettes

CNS neuroblastoma, FOXR2-activated

CNS tumor with BCOR internal tandem duplication

CNS embryonal tumor

Initially, the term primitive neuroectodermal tumor (PNET) encompassed a wide variety of previously individually named tumors which all seemed to share certain pathologic features suggesting origin from a common progenitor cell in the subependymal matrix (primitive neuroectodermal cells) (although the actual cell of origin is unknown). They are histologically indistinguishable but genetically distinct [1)].

Now, the recommendation is to call these “embryonal tumors”, [2)] but the term PNET is entrenched. These tumors include: retinoblastoma, pineoblastoma, neuroblastoma, esthesioneuroblastoma.

Medulloblastoma (MB) is more than just a PNET of the posterior fossa, as alterations involved in evolution of MBs such as beta-catenin and APC mutations are absent in pineoblastomas and supratentorial PNETs (sPNETs). At least some MBs originate from the external granular layer (EGL) of the cerebellum.

Embryonal tumors most commonly arise in the cerebellar vermis (medulloblastoma), but also occur in cerebrum, pineal, brainstem or spinal cord. Primary spinal cord PNETs are extremely rare (approximately 30 cases reported by 2007 [3)]).

Dissemination: Embryonal tumors (ETs) may disseminate via the CSF spontaneously, [4)] or iatrogenically (following surgery or shunting, the latter is a rare cause of tumor dissemination [5)]). Thus, all patients with ETs require spinal axis evaluation (gadolinium enhanced MRI is about as sensitive as water-soluble myelography) and cytologic examination of CSF. Prophylactic craniospinal XRT is indicated following surgical removal, but cranial XRT is avoided if at all possible before 3 years of age to avoid intellectual impairment and growth retardation. Extraneural metastases can also occur.

Collin ’s law: AKA period of risk of recurren ce (PRR) is often applied to children who have been treated for embryonal tumors (especially medulloblastoma) but may also be used with any tumor thought to arise from a gestational event. It states that PRR is equal to the age at diagnosis plus 9 months [6)]. Patients that remain free of recurrence beyond the PRR have a much lower risk of recurrence, however recurrence beyond this time has been reported in a small number (≈ 1.4%) of cases, [7)] and other tumors may occur e.g. as a result of induction by XRTused to treat the initial tumor.

A mass of rapidly growing cells that begins in embryonic (fetal) tissue. Embryonal tumors may be benign or malignant and include neuroblastomas and Wilms tumors. Also called embryoma.

CNS Embryonal Tumor with BRD4-LEUTX Fusion

CNS Embryonal Tumor with BRD4-LEUTX Fusion.

1)

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