Diffuse intrinsic pontine glioma outcome

When DIPGs are biopsied, they are usually grade III or grade IV. Occasionally, they are grade II, but because of their location in the brain they are still considered malignant. That being said, diffuse intrinsic pontine gliomas usually progress like grade IV glioblastoma multiforme tumors.

Diffuse Intrinsic Pontine Glioma is the most aggressive form of high-grade gliomas in children it is incurable, aggressive, and disabling due to localization in the midline brainstem.

They are universally fatal and associated with progression free and median overall survival rates of 5.7 and 7.9 mo, respectively $^{1)}$.

Despite advances in neurosurgery, radiotherapy, and chemotherapy trials, current treatment strategies only serve as palliation. New therapies that are specific to DMG are under active investigation. An exciting and novel approach developed by Mount et al ², may significantly advance care for these lethal tumors by using anti-GD2 chimeric antigen receptor (CAR)-modified T cells to target GD2, an antigen that is highly expressed on tumors of neuroectodermal origin such as neuroblastoma and melanoma.

CAR T cells were originally developed by Kochenderfer et al ³⁾ against leukemia and lymphoma but have recently been adapted and explored as immunotherapy against central nervous system tumors. CAR T cells are synthetically engineered from a patient's autologous T-cells to recognize cancerspecific antigens and generate a strong anti-tumor immune response.

Diffuse midline glioma, H3 K27M-mutant, represents a newly introduced, predominantly astrocytic tumor of pons, thalamus, or spinal cord with poor prognosis and a K27M mutation in one of three histone genes. While the H3 K27M mutation had been considered as being the defining mutation of this brain tumor group, it may also occur in other brain tumors, such as pilocytic astrocytoma ⁴⁾ and ependymoma ⁵⁾, without necessarily being associated with poor prognosis. Whether these tumors have to be classified as diffuse midline glioma (grade IV) with aberrant phenotype or as low-grade glioma with unusual H3 K27M mutation remains unclear to date. These cases certainly represent only a small minority of brain tumors, but their classification poses problems.

T2-FLAIR mismatch sign in DIPG may be an indicator for better response to radiotherapy and a better prognostic factor ⁶⁾.

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