Neuronal and Mixed Glioneuronal Tumors

Overview

	Age range (median age in years)	Female: male ratio	Cell of origin	WHO Grade	Molecular/genetic markers	Prognosis
Dysembryoplastic neuroepithelial tumor	0-20	1:1.5	Glioneuronal cells	I	S100, NeuN, synaptophysin, GFAP	Good
Ganglioglioma	0-70	1:1.5	Ganglion cells	I	*BRAF V600E*	Favorable
Desmoplastic infantile astrocytoma (DIA) and ganglioglioma (DIG)	0-2	1:1	Astrocytes Mature neuronal cells	I	GFAP, vimentin, *MAP2* (DIG) BRAF	Favorable
Dysplastic cerebellar gangliocytoma (Lhermitte-Duclos disease)	30-50	1:1	Ganglion cells	I	PTEN germline loss; PTEN/AKT/mTOR pathway	Guarded
Rosette-forming glioneuronal tumor	6-79 (27)	1.3:1	Pluripotent cells of subependymal plate	I	Undefined	Favorable
Diffuse leptomeningeal glioneuronal tumor	0.5-46 (5)	1:1	Oligodendroglial-like cells	I	*KIAA1549-BRAF* fusion, 1p deletion	Guarded
Central neurocytoma	20-40 (29)	1:1	Mature neurocytes	II	Synaptophysin, NeuN, *MAP-2*, *MYCN*, loss of *BIN1*	Favorable
Extraventricular neurocytoma	5-76 (34)	1:1	Mislocated neural progenitor cells	II	Undefined	Good
Paraganglioma	9-75 (46)	1:1.5	Neural crest cells of paraganglia	I	*SDHD*, *SDHA*, *SDHAF2*, *SDHC*, *SDHB*	Good

AKT, Ak strain transforming; PTEN, Phosphatase and TENsin homolog; TOR, Target of rapamycin.