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Extra-temporal pediatric low-grade gliomas and epilepsy

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Abstract

Low-grade gliomas, especially glioneuronal tumors, are a common cause of epilepsy in children. Seizures associated with low-grade pediatric tumors are medically refractory and present a significant burden to patients. Often, morbidity and patients´ quality of life are determined rather by the control of seizures than the oncological process itself and the resolution of epilepsy represents an important part in the treatment of LGGs. The pathogenesis of tumor-related seizures in focal LGG tumors is multifactorial, and mechanisms differ probably among patients and tumor types. Pediatric low-grade tumors associated with epilepsy include a series of neoplasms that have a pure astrocytic or glioneuronal lineage. They are usually benign tumors with a neocortical localization typically in the temporal lobes, but also in other supratentorial locations. Gangliogliomas and dysembryoplastic neuroepithelial tumors (DNET) are the most common entities together with astrocytic gliomas (pilocytic astrocytomas and pleomorphic xanthoastrocytoma) and angiocentric gliomas, and dual pathology is found in up to 40% of glioneuronal tumors. The treatment of low-grade gliomas and associated epilepsy is based mainly on resection and the extent of surgery is the main predictor of postoperative seizure control in patients with a LGG. Long-term epilepsy-associated tumors (LEATs) tend to be well-circumscribed, and therefore, the chances for a complete resection and epilepsy control with a safe approach are very high. New treatments have emerged as alternatives to open microsurgical approaches, including laser thermal ablation or the use of BRAF inhibitors. Future advances in identifying seizure-related biomarkers and molecular tumor pathways will facilitate targeted treatment strategies that will have a deep impact both in oncologic and epilepsy outcomes.

Keywords: Epilepsy; Glioneuronal tumors; Neuroepithelial; Pediatric brain tumor; Resective epilepsy surgery; Seizures.