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IDH-Mutant Low-grade Glioma: Advances in Molecular Diagnosis, Management, and Future Directions

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Abstract

Purpose of review: IDH-mutant low-grade gliomas (LGG) have emerged as a distinct clinical and molecular entity with unique treatment considerations. Here, we review updates in IDH-mutant LGG diagnosis and classification, imaging biomarkers, therapies, and neurocognitive and patient-reported outcomes.

Recent findings: CDKN2A/B homozygous deletion in IDH-mutant astrocytoma is associated with shorter survival, similar to WHO grade 4. The T2-FLAIR mismatch, a highly specific but insensitive sign, is diagnostic of IDH-mutant astrocytoma. Maximal safe resection is currently indicated in all LGG cases. Radiotherapy with subsequent PCV (procarbazine, lomustine, vincristine) provides longer overall survival compared to radiotherapy alone. Temozolomide in place of PCV is reasonable, but high-level evidence is still lacking. LGG adjuvant treatment has important quality of life and neurocognitive side effects that should be considered. Although incurable, IDH-mutant LGG have a favorable survival compared to IDH-WT glioma. Recent advances in molecular-based classification, imaging, and targeted therapies will hopefully improve survival and quality of life.

Keywords: Astrocytoma; CDKN2A/B; Glioma; IDH mutant; Imaging biomarker; Neurocognitive; Oligodendroglioma; PCV; Quality of life.