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Pediatric-type high-grade glioma of the spinal cord in the molecular era: institutional case series and updated systematic review of the literature

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Abstract

Pediatric high-grade glioma of the spinal cord is an exceptionally rare entity with limited understanding of its natural history and biological behaviors, particularly in comparison to similar tumors in adults or within the intracranial compartment. This updated systematic review comprehensively investigates all cases in the English literature through March 2024 following PRISMA guidelines with an emphasis on histopathologic diagnosis, treatment paradigms, outcome, and molecular characterization where reported. Eighty-three articles met inclusion with 132 spinal tumors diagnosed as "glioblastoma" based on relevant histopathologic criteria. Fourteen studies included molecular genetic testing, with IDH status reported in only two patients and seven patients incurring a histone mutation. While a more recent year of treatment, greater extent of resection, and utilization of adjuvant chemoradiation were associated with increased overall survival (OS) to suggest that modern treatment regimens are improving clinical outcomes, insufficient data exists to determine differences in OS based upon molecular profiling thereby limiting therapeutic progress. The authors further include two new cases to juxtapose molecular tumor profiles with potential differences in outcome. This review ultimately aims to emphasize the importance of reporting and utilizing integrated diagnoses to better understand the natural history and prognosis of high-grade glial tumors in the pediatric spine. Challenges in this field continue to include tumor rarity, an inability to reliably update the molecular profiles of previously obtained tissue, and reliance on literature published prior to the "molecular era" of CNS neoplasms, which likely represents a broader spectrum of high-grade genetic variants.

Keywords: Children; Diffuse midline glioma; Glioblastoma; High-grade glioma; Molecular genetics; Pediatric spine tumor; Spinal cord.