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HGG-13. INFANT-TYPE HEMISPHERIC GLIOMA (IHG): AN INDIVIDUAL PATIENT DATA META-ANALYSIS
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BACKGROUND: Infant-type hemispheric gliomas (IHG) are epigenetically distinct pediatric high-grade gliomas characterized by fusions in receptor tyrosine kinase (RTK) genes. METHODS: We performed a methodical literature search, including 30 publications (22 case reports), to identify patients who met the diagnostic criteria of IHG based on the 2021 WHO Classification of CNS Tumors. Individual patient data were obtained from published literature and/or via the authors of the publications. Survival analysis was conducted using the Kaplan-Meier method, and multivariate analysis was performed to investigate the effect of clinical and molecular variables on outcomes. RESULTS: Hundred-fifty-five previously reported and one unpublished IHG were identified: 131 (84%) had fusions in RTK genes, of which *ALK* was most prevalent (62/131), followed by *NTRK1/2/3* (30/131), *ROS1* (30/131), and *MET* (9/131). Twenty-five patients, with either no identified RTK fusion (6/156) or not assessable fusion (19/156), had methylation scores ≥ 0.9 for IHG (using the Molecular Neuropathology brain tumor classifier versions ≥ 11b4). Surgery followed by adjuvant chemotherapy in 69% (67/97) was the most common primary treatment used in our cohort. The 3-year EFS and OS were 55% (95%CI: 45-67) and 81 % (95%CI: 73-89). 41 patients with relapsed or progressive tumors received various second-line treatments including surgery, chemotherapy, radiotherapy, and targeted therapy. Based on multivariate analysis, complete resection resulted in better EFS ($p = 0.05$) and OS ($p = 0.009$), and the presence and type of RTK gene fusion were not associated with clinical outcomes. CONCLUSION: Our results show that despite favorable OS, patients with IHG often show early progression, indicating that the primary optimal treatment for IHG is yet to be established. Our analysis further indicates that achieving a safe, complete resection may play an important role in treating these patients. A comprehensive analysis of the salvage regimen is required to understand their role in OS.