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Molecular profile and clinical outcome of adult primary spinal cord glioblastoma: a systematic review

Bahie Ezzat, Tirone Young, Alexander J Schüpper, Roshini Kalagara, Jack Y Zhang, Michael Lemonick, Priya Bhanot, Addison Quinones, Tanvir Choudhri, Isabelle M Germano

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Abstract

Objective: Primary spinal cord glioblastoma (scGB) is a rare and aggressive spinal glioma, making up 7.5% of such cases. Whereas molecular profiles associated with improved overall survival (OS) are well studied for cranial glioblastoma (GB), the molecular characteristics of scGB are less documented. This review sought to document the molecular signatures of scGB, explore current treatment strategies, and evaluate clinical outcomes.

Methods: A systematic literature review following the PRISMA guidelines searched the PubMed, Embase, and CENTRAL databases (January 1, 2013, to October 14, 2023) using glioblastoma-, spine-, and genetics-related keywords. Inclusion criteria were English-language articles on humans with histologically confirmed primary scGB, excluding drop metastases. Data on demographic characteristics, treatments, molecular profile, and outcome were extracted.

Results: Over 10 years, 71 patients with adult primary scGB were reported in 31 papers. Most patients were located in Asia (53%) and the United States (23%). The median (range) age was 32 (24-47) years, with 61% of patients male. Tumors occurred primarily in the thoracic region (42%). Clinical presentation included motor deficits (92%), sensory deficits (86%), neck/back pain (68%), and bowel/bladder dysfunction (59%). Patients underwent subtotal resection (51%), gross-total resection (GTR) (23%), and biopsy (26%). Postoperative adjuvant treatment included concomitant external beam radiation therapy (XRT) and temozolomide (TMZ) in the majority of cases (66%), as well as palliative care without adjuvant treatment (17%). The molecular signature of scGB was similar to its cranial counterpart in terms of MGMT-promoter methylation (40% increased methylation) and higher for mutant TERT (50%) but decreased for wild-type tumor protein p53 (41% decreased mutation). Median (range) OS was 10 (6-18) months, and median progression-free survival (PFS) was 7 (3-10) months. PFS was significantly higher in patients treated with XRT/TMZ: median 15 months vs 4.5 months (95% CI -1.32 to 22.56, $p < 0.05$).

Conclusions: Primary scGB remains a rare disease with notable variations in treatment, potentially influenced by geographical availability. The observed molecular profile, when compared to that of cranial GB, emphasizes the need for further genomic validation and data collection. Surgical advancements to overcome the challenges of accomplishing GTR may contribute to improved OS.

Keywords: clinical outcome; molecular profile; oncology; primary spinal cord glioblastoma.