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Brainstem Gliomas With Isocitrate Dehydrogenase Mutation: Natural History, Clinical-Radiological Features, Management Strategy, and Long-Term Outcome

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Abstract

Background and objectives: This study aimed to investigate the clinical, radiological, pathological features, treatment options, and outcomes of isocitrate dehydrogenase (IDH)-mutant brainstem gliomas (BSG-IDHmut).

Methods: A retrospective analysis of 22 patients diagnosed with BSG-IDHmut and treated at our institution from January 2011 to January 2017 was performed. Their clinical, radiological data, and long-term outcomes were collected and analyzed.

Results: The median age of patients was 38.5 years, with a male predominance (63.6%). All patients had IDH1 and TP53 mutations, with noncanonical IDH mutations in 59.1% of cases, 06-methylguanine-DNA methyltransferase promoter methylation in 55.6%, and alpha-thalassemia mental retardation X-linked loss in 63.2%, respectively. Tumors were primarily located in the pontine-medullary oblongata (54.5%) and frequently involved the pontine brachium (50%). Most tumors exhibited ill-defined boundaries (68.2%), no T2-FLAIR mismatch (100%), and no contrast enhancement (86.3%). Two radiological growth patterns were also identified: focal and extensively infiltrative, which were associated with the treatment strategy when tumor recurred. Seven patients (31.8%) received surgery only and 15 (68.2%) surgery plus other therapy. The median overall survival was 124.8 months, with 1-year, 2-year, 5-year, and 10-year survival rates of 81.8%, 68.2%, 54.5%, and 13.6%, respectively. Six patients experienced tumor recurrence, and all retained their radiological growth patterns, with 2 transformed into central nervous system World Health Organization grade 4.

Conclusion: BSG-IDHmut represents a unique subgroup of brainstem gliomas with distinctive features and more favorable prognosis compared with other brainstem gliomas. Further research is required to better understand the molecular mechanisms and optimize treatment strategies for this rare and complex disease.

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