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Insights from a Multicenter Study on Adult H3 K27M-Mutated Glioma: Surgical Resection's Limited Influence on Overall Survival, ATRX as Molecular Prognosticator

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Abstract

Background: H3 K27M-mutated gliomas were first described as a new grade 4 entity in the 2016 WHO classification. Current studies have focused on its typical appearance in children and young adults, increasing the need to better understand the prognostic factors and impact of surgery on adults. Here, we report a multicentric study of this entity in adults.

Methods: We included molecularly confirmed H3 K27M-mutated glioma cases in patients >18 years diagnosed between 2016 and 2022. Clinical, radiological, and surgical features were analyzed. Univariate and multivariate analyses were performed to identify prognostic factors.

Results: Among 70 patients with a mean age of 36.1 years, the median overall survival (OS) was 13.6 + 14 months. Gross-total resection was achieved in 14.3% of patients, whereas 30% had a subtotal resection and 54.3% a biopsy. Tumors located in telencephalon/diencephalon/myelencephalon were associated with a poorer OS, while a location in the mesencephalon/metencephalon showed a significantly longer OS (8.7 vs. 25.0 months, $p=0.007$). Preoperative Karnofsky Performance Score (KPS) < 80 showed a reduced OS (4.2 vs. 18 months, $p=0.02$). Furthermore, ATRX loss, found in 25.7%, was independently associated with an increased OS (31 vs. 8.3 months, $p=0.0029$). Notably, patients undergoing resection showed no survival benefit over biopsy (12 vs. 11 months, $p=0.4006$).

Conclusion: The present study describes surgical features of H3 K27M-mutated glioma in adulthood in a large multicentric study. Our data reveal that ATRX status, location and KPS significantly impact OS in H3 K27M-mutated glioma. Importantly, our dataset indicates that resection does not offer a survival advantage over biopsy.

Keywords: H3 K27M; H3F3A; HIST1H3B; High-grade glioma; Histone H3.

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