ABSTRACT

World Neurosurg. 2022 Jun 10:S1878-8750(22)00803-8. doi: 10.1016/j.wneu.2022.06.020. Online ahead of print.

H3K27M-Altered Diffuse Midline Gliomas Among Adult Patients: A Systematic Review of Clinical Features and Survival Analysis.

Alamer OB(1), Jimenez AE(2), Azad TD(2), Bettegowda C(2), Mukherjee D(3).

Author information:
(1)College of Medicine, King Saud bin Abdulaziz University for Health Sciences, Riyadh, Saudi Arabia.
(2)Department of Neurosurgery, Johns Hopkins Hospital, Baltimore, MD 21224.
(3)Department of Neurosurgery, Johns Hopkins Hospital, Baltimore, MD 21224. Electronic address: dmukher1@jhmi.edu.

OBJECTIVE: To summarize the clinical characteristics, histo-genomic profiles, management strategies, and survival outcomes of H3K27M-altered adult diffuse midline gliomas (aDMGs).

METHODS: PubMed, Scopus, and Cochrane databases were used to identify relevant articles. Papers including H3K27M-altered aDMGs with sufficient clinical outcome data were included. Descriptive clinical characteristics and survival analysis were also conducted.

RESULTS: Twenty studies describing 135 patients were included. The median age at diagnosis was 42 years and there was a slight male predominance (N=60, 54%). In our cohort, fifteen (11%) patients experienced headache, 10 had nausea and vomiting (7%), and 10 had ataxia (7%). Within this cohort, histopathologic diagnoses included glioblastoma (N=22, 40%) and anaplastic astrocytoma (N=21, 38%), while genetic alterations included ATRX mutation (N=22, 16%), PTPN11 mutation (N=9, 7%), and MGMT promoter methylation (N=9, 7%). Among histo-genetic alterations, only ATRX mutation was associated with survival and correlated with worse prognosis (Log-risk, P=0.04). Neither surgical resection versus biopsy nor greater extent of resection demonstrated survival benefit in our cohort. Chemotherapy was administered in 98 (73%) cases with radiotherapy administered in 71 (53%) cases. Unlike chemotherapy, radiotherapy demonstrated a significant survival benefit (log-risk, P=0.019). Median overall survival (OS) and progression-free survival within our patient cohort were 10 and 7 months, respectively.

CONCLUSION: H3K27M-altered aDMGs were associated with relatively poor survival. ATRX gene mutation was significantly associated with survival disadvantage, while radiotherapy was associated with survival benefit. Large, prospective studies are needed to establish a standard management strategy and provide reliable prognostic conclusions.

DOI: 10.1016/j.wneu.2022.06.020
PMID: 35697228