ABSTRACT

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Adult diffuse midline gliomas H3 K27-altered: review of a redefined entity.

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INTRODUCTION: Diffuse midline glioma (DMG) H3 K27-altered is a type of high-grade gliomas first recognized as a new entity in the 2016 World Health Organization Classification of Central Nervous System (CNS) Tumors as DMG H3 K27M-mutant, recently renamed in the new 2021 WHO classification. The aim of this review is to describe the characteristics of diffuse midline gliomas H3 K27-altered in the adult population.

METHODS: We performed a review of the current literature regarding the genetic, clinical, imaging characteristics and management of diffuse midline gliomas H3 K27-altered in adult patients.

RESULTS: The 2021 WHO classification now designates the previously recognized DMG H3K27M-mutant as DMG H3 K27-altered, recognizing the alternative mechanisms by which the pathogenic pathway can be altered. Thus, the diagnostic criteria for this entity consist of diffuse growth pattern, midline anatomic location, and H3 K27-specific neuroglial mutations. DMGs' characteristic midline location makes them difficult to surgically resect and biopsy, carrying high mortality and morbidity rates, with median survival ranging from 9 to 12 months in adult patients.

CONCLUSION: The diagnosis of DMGs H3 K27-altered in adult patients should be considered upon neurological symptoms associated with an infiltrative midline brain tumor detected on imaging. Future studies are necessary to continue refining their characteristics in this age group.

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