## A HOMOGENEOUS TREATMENT FOR NON-DIPG DIFFUSE MIDLINE GLIOMA

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## Abstract

Introduction. The H3K27M-mutant diffuse midline glioma (DMG) was first included in the WHO Classification of CNS (central nervous system) tumors in 2016, and confirmed in its fifth edition. The biological behavior and dismal prognosis of this tumor resemble DIPG (diffuse intrinsic pontine gliomas). Homogeneously-treated series are rarely reported. Methods. From 2016 onwards, we treated patients with DMG with radiotherapy and concomitant/adjuvant nimotuzumab/vinorelbine, plus re-irradiation at relapse, as already done for DIPG (DOI10.1007/s11060-014-1428-z). Results. We treated nine patients, seven females, median age at diagnosis of 13 years-old. Tumor sites were: thalamic in five cases, pontocerebellar in two, pineal in one, and paratrigonal with nodular/leptomeningeal dissemination in one. Three patients were biopsied, and six had partial tumor resections. Central review of the pathologists' diagnoses was performed. The median time to local progression was 12.7 months, and the median overall survival was 17.8 months. Six patients died of tumor progression, one of cerebral bleeding whose tumor was progressing. Two were alive, one in continuous remission, the other after a relapse, at 38.6 and 46.3 months after diagnosis, respectively. Progression-free survival was 33.3% at one year. Overall survival was 88.9%, 33.3% and 22.2% at 1, 2 and 3 years, respectively. Conclusions. This is one of only a handful of reports on homogeneously-treated series. The results obtained are comparable with those seen in patients with DIPG. Given the phenotypically- and molecularly-defined setting of DMG and severe outcome in this orphan population, they should be treated and included in registries and protocols of DIPG.

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TABLE 1 .docx available at https://authorea.com/users/356325/articles/713545-a-homogeneous-treatment-for-non-dipg-diffuse-midline-glioma

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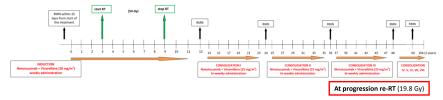
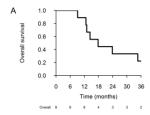


Figure 1



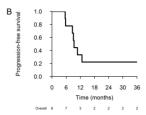


Figure 2