Pediatric Thalamic Tumors: The Role of Surgery In The H3 K27M-mutant Midline Glioma Era

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Disclosure

• The authors have nothing to disclose
Introduction

• The WHO 2016 classification of CNS tumors defined \textit{diffuse midline glioma (DMG), H3 K27M-mutant} as a new tumor entity

• With this new entity WHO IV and including DIPG as well as thalamic infiltrative gliomas the role of surgery in the management of thalamic pediatric non-pilocytic gliomas needs to be revisited
Methods

• A retrospective review of a consecutive series of children operated on a thalamic tumor between 1992 and May 2018 was performed.

• Neuroimaging data were reviewed for localization and extent of resection

• Pathology was re-reviewed according to the current WHO classification, including assessment of histone H3 K27 mutational status.
Results

- Forty-nine patients with a thalamic tumor aged <18 years at diagnosis were identified.

- Twenty-five patients (51%) had a non-pilocytic infiltrative glioma, of which the H3 K27M status was available in 22, of which 14 were diagnosed as diffuse midline glioma (DMG) H3 K27M-mutant.

- There was no statistically significant difference in survival between patients harboring the H3 K27M mutation and wildtype (Fig.1a).

- Resection ( “any resection >50%” vs “biopsy”) (Fig.1b) and histological tumor grade (“II” vs “III+IV”) were statistically significant predictors of survival. These results remained significant on multivariate analysis.
Results

Fig. 1a

H3K27M status
- wildtype (n=7)
- mutated (n=14)

overall survival

p=0.357

years

Fig. 1b

surgery
- biopsy (n=9)
- resection (n=15)

overall survival

p=0.044

years
Discussion

• Currently there is no consensus on the impact of resective surgery in the treatment of infiltrative non-pilocytic astrocytomas of the thalamus.

• A majority of infiltrative thalamic gliomas in children harbor a H3 K27M mutation, this could be confirmed in our series.

• Current data suggest a H3 K27M mutation as a molecular signature associated with a poor prognosis in so-called “diffuse midline gliomas” to outweigh the impact of tumor location.

• Although retrospective and with a limited number of cases our data do not corroborate the reported negative impact of the presence of the H3K27M mutation.


Summary

- Based on our institutional review and adding our experience to reported surgical series in experienced centers we would still advocate to consider an attempt at maximal safe resection in the multidisciplinary treatment also of thalamic infiltrative non-pilocytic gliomas.