



Long term survival of the patient with thalamic K27M mutant glioblastoma after stereotactic radiosurgery

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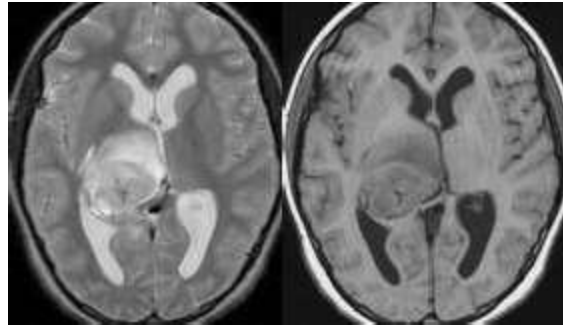
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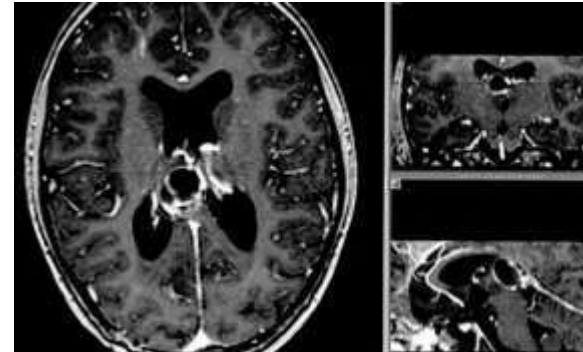


Objective: It was shown that K27M mutation is associated with especially aggressive clinical course of the pediatric midline GBM. These tumors disclosed low rate of total tumor resections, frequent postoperative metastatic dissemination and extremely short overall survival after standard treatment. In the present case, the tumor resected showed all typical histological, cytogenetic and epigenetic patterns of K27 mutant GBM grade IV, including PDGFRA oncogene amplification.

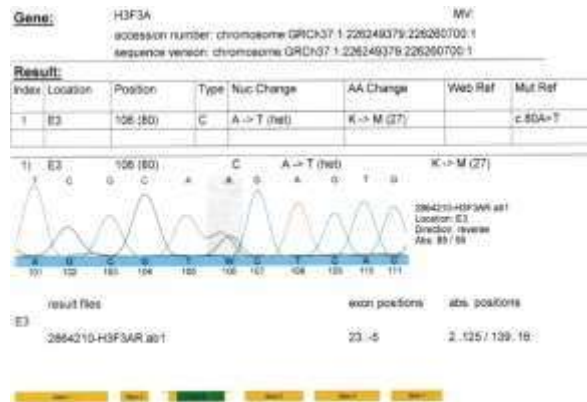
Pre-surgery MRI scan



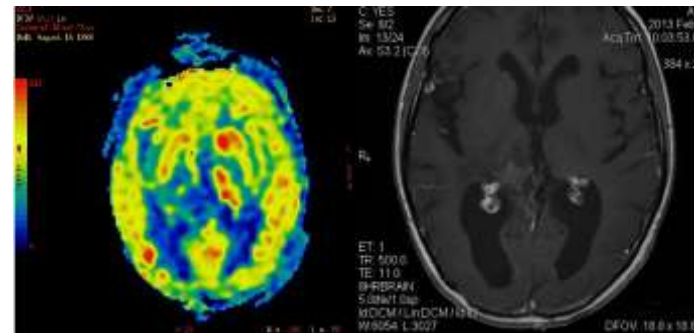
Postoperative recurrence and SRS treatment plan



DNA sequencing revealed K27M mutation of H3.3. gene in this tumor



Absence of recurrence and radiation-induced cavernoma formation 7 years after SRS



Methods: Subtotal surgical removal of the tumor was performed in 13-year-old female with deep-seated heterogeneous intracerebral tumor involving the left thalamic region at with followed conventional radiotherapy. Histopathological data were consistent with glioblastoma WHO grade IV with isomorphic, small-cell phenotype. Direct DNA sequencing disclosed the presence of a K27M (c.83A>T, p.Lys27Met) mutation of the H3F3A gene in this tumor. MGMT promoter was unmethylated. 8 months after operation patterns of clinical deterioration were noted and neuroradiological investigation identified a local relapse of the tumor. Radiosurgery using the gamma knife (total dose 14 Gr 45% isodose) was held. No additional therapy was also performed. After this the patient was followed.

Conclusion: The present unusual case could suggest that stereotactic radiosurgery could be as an optimal postsurgical treatment of K27M mutant pGBM taking into an account a low efficacy of TMZ-based therapy due to lack of MGMT promoter methylation in almost all these tumors. On the other hand, some unique and yet undiscovered molecular characteristics of this individual K27M mutant GBM case could underlie its favorable clinical course and guarantee a stable disease for a long period of time, regardless of the treatment applied.