Familial brain tumor: a case report of homozygous twins affected by glioma

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Introduction

Genetic syndromes such as neurofibromatosis, Turcot syndrome, Li-Fraumeni syndrome, are associated with inherited cancer, including brain tumor, although the mechanism of action as well as the involved genes are unknown. Cases of familial gliomas other than these syndrome have not yet been definitively demonstrated and represent a sporadic event. We report the case of homozygous twins treated at our Institution for glioma without an identifiable genetic cause.

Materials and methods

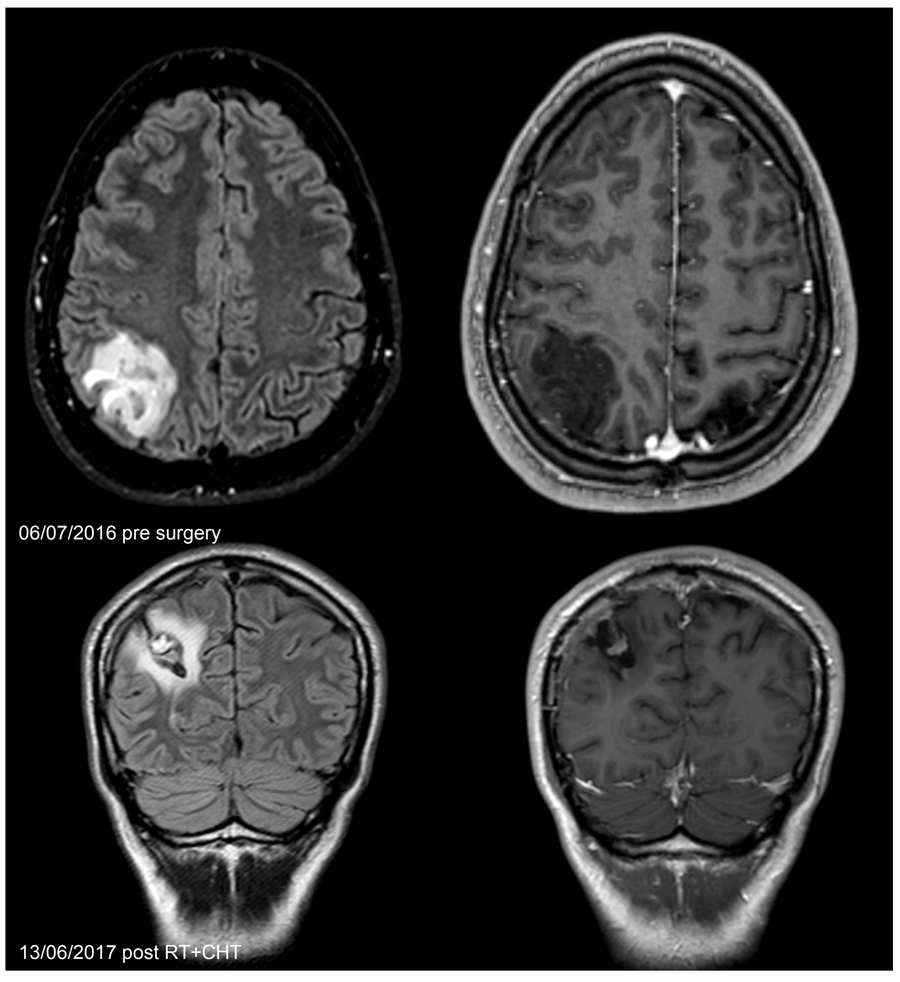
CASE I

A 32 year-old-male in a good general conditions presented with a new onset seizure. MRI showed a non-enhancing right parietal lesion and he underwent to a total surgical resection with histologic diagnosis of **anaplastic oligodendroglioma** (IDH 1 positive, negative P53, positive ATRX, negative TERT, MIB-1 8-10%, loss of heterozygosity on 1p and 19q, MGMT methylated). He was treated with conformational radiotherapy (56 Gy) and adjuvant chemotherapy with PCV. The patient is currently in good clinical condition with radiological stability.

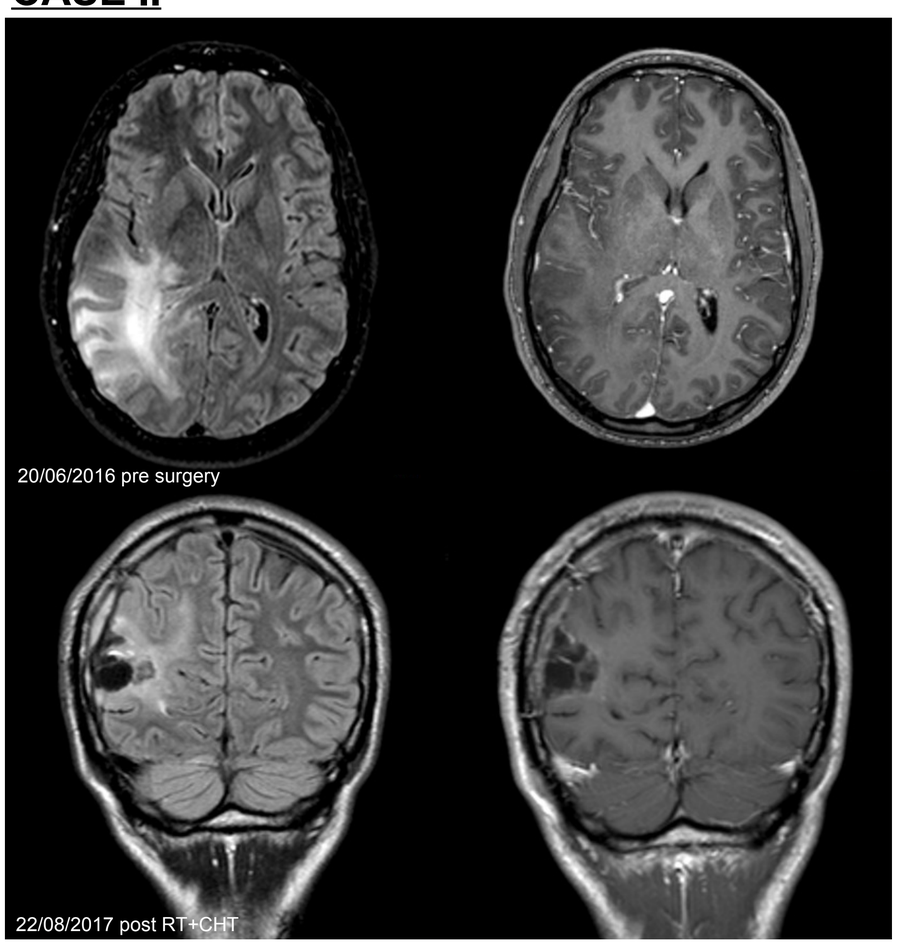
CASE II

A 32 year-old-male, totally asymptomatic, becoming aware of the brother's diagnoses, underwent radiological screening with the detection of non-enhancing right parietal lesion. He underwent a partial resection of the tumor, with diagnosis of **gemistocytic astrocytoma** (IDH1 mutated, MIB 1-2%, Immunohistochemistry: p53 and IDH1 positive, ATRX negative, molecular genetics analysis not performed due to material shortage). Then he was treated with radiotherapy (56 Gy, 2 Gy/fraction). Due to tumor residue, Physician suggested to start adjuvant temozolomide according to 5/28 schedule.

CASE I



CASE II



Discussion and conclusion

Although familial gliomas represent a rare event with a limited epidemiological relevance, there are some cases described in literature. The current knowledge does not allow to provide consistent evidence of hereditary factors in the development of some gliomas. Large prospective studies and cancer registries are needed to determine the role of inheritance in gliomas. Particular attention should be paid to the role of environment as well lifestyle in the onset of familial brain tumors. This could allow to implement corrective and preventive strategies, to date inexistent.

References

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