GLIOMATOSIS CEREBRI WITH MALIGNANT TRANSFORMATION TO A LONG-SURVIVAL GliOBLASTOMA MULTIFORME AFTER TEN YEARS OF EVOLUTION

J. Pérez1, J. Fernández1, T. Ribas2, P. Barrio1, and A. Mostaza1; 1Department of Neurosurgery, University Hospital of León, León, Spain; 2Department of Anatomic Pathology, University Hospital of León, León, Spain

OBJECTIVES: We report an unusual case of gliomatosis cerebri which transformed into a high-grade glioma, both of them with a long-survival period. We emphasize in the lack of large studies about the biological and clinical behavior of these tumors, impeding the improvement of therapeutic options and our knowledge.

METHODS: A 34-year-old male came to consulting room suffering an intense pain located around the right mastoid apophysis, with no neurological deficit associated. In the Magnetic Resonance Image ('MRI') it is shown a left frontal cortico-subcortical mass with no enhancing-image and not well-defined edges. This image was not initially related to the clinical process of the patient. The surgery was performed successfully and the pain was solved. Once the mass was diagnosed as a gliomatosis cerebri, the patient received adjuvant treatments and, after 10 years of uneventful consultations he referred headache and showed a nominal dysphasia. The ‘MRI’ and ‘SPECT’ confirmed the malignant transformation of the left frontal lesion into a high-grade glioma, without cerebrospinal fluid dissemination and/or multifocal progression. This mass was removed in 2011 guided with 5-ALA, and the post-operative period had not any complications. The patient received new cycles of chemotherapy and radiotherapy and, after 3 years of progression-free survival and 14 years of overall survival since the first diagnosis, the patient keeps a high-quality of life, with a Karnofsky Performance Status of 100.

RESULTS: The first sample showed slight architectural changes in the white matter with higher cellular density and atypical cells which followed the myelinated bands. There was no endovascular proliferation and/or necrosis. It was positive for p53. The second sample showed all the criteria for glioblastoma multiforme, with predominance of gemistocytic cells. It was also positive for p53 and its MIB-1 index was over 20%. Currently, we are concluding the tests for IDH-1 and IDH-2 mutations.

CONCLUSIONS: The gliomatosis cerebri is not a well-known entity and its biological mechanisms are unknown yet. Its survival is very changeable, from weeks to several years. Once the malignant transformation happens, the survival of these patients is meaningfully reduced. In our case, maybe because of the immunohistochemical characteristics and/or not being a diffuse lesion, the patient is having a very long-surveillance period for both lesions, showing us that there are many concepts that we have to learn about the biological behavior of the tumors.