EFFICACY OF BEVACIZUMAB IN A PRIMARY DIFFUSE LEPTOMENINGEAL GLIONEURONAL TUMOR.

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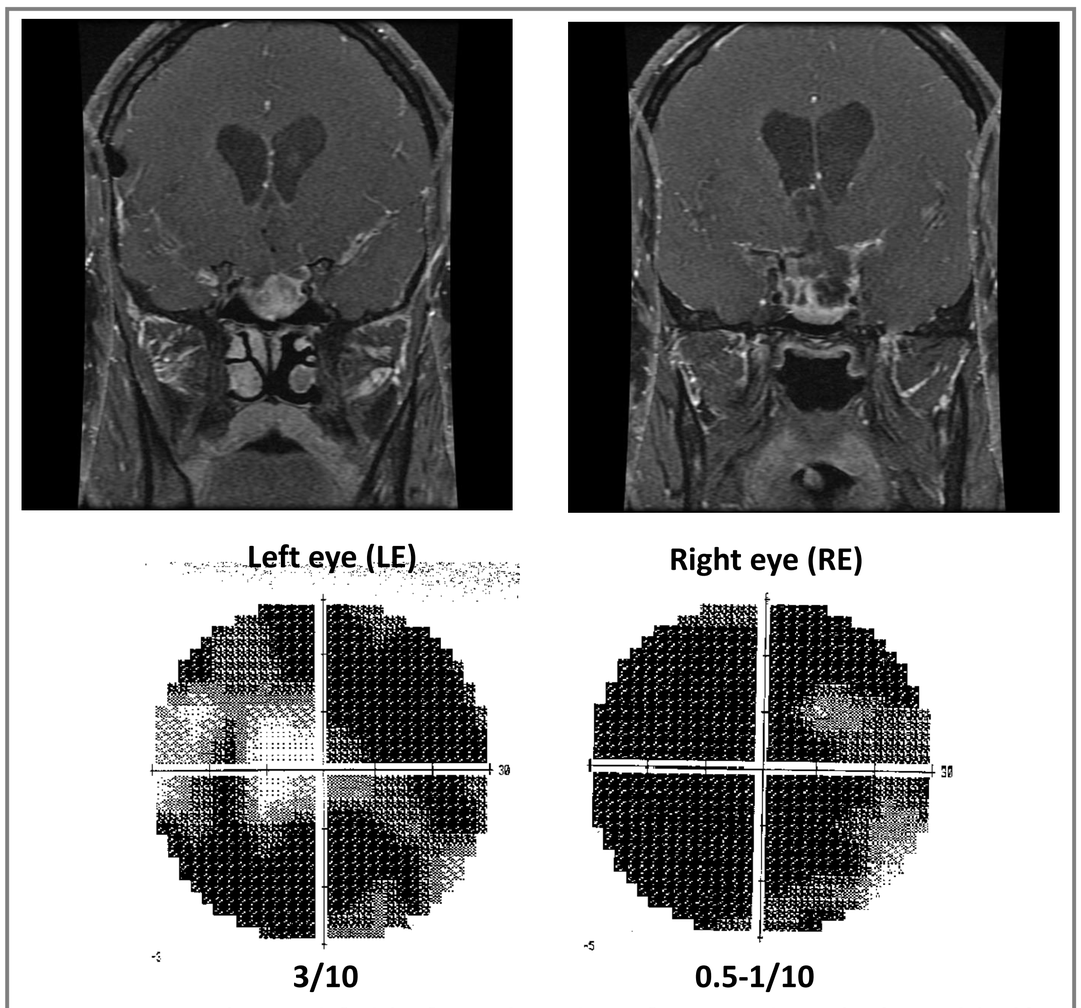
Background. Diffuse leptomeningeal glioneuronal tumor (DL-GNT) is a rare childhood brain tumor that involves the basal cisterns and the interhemispheric sulcus but lacks intraparenchymal involvement. The description in adults of DL-GNT in the literature is limited to few case reports with poor results in terms of overall survival (OS) and progression-free survival (PFS).
Currently, there are no cytotoxic drugs that has shown efficacy. Here we describe an adult DL-GNT successfully treated with bevacizumab.

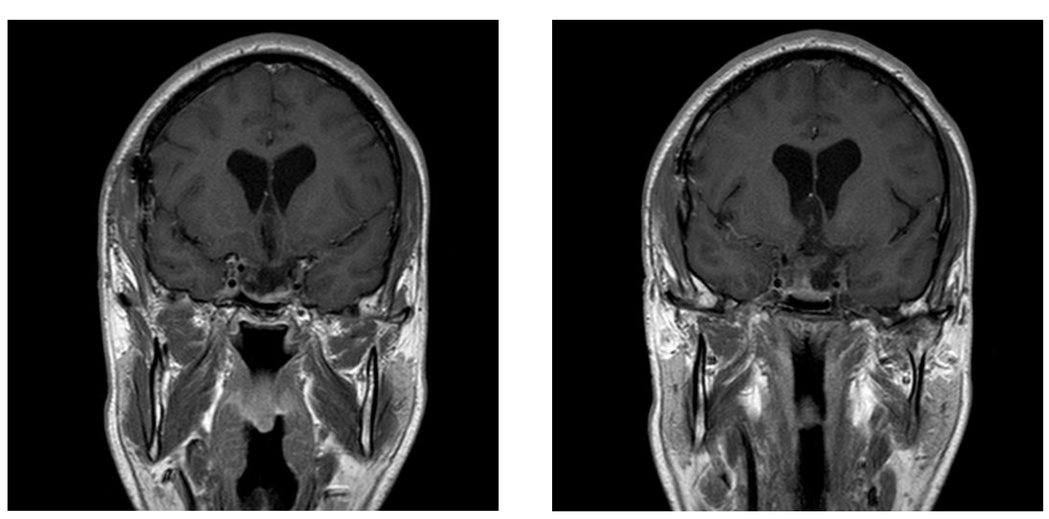
Case report. In June 2012 a 33-year old man presented with transient decrease in visual acuity on the left eye (LE). Fundus oculi revealed an asymmetric bilateral papilledema and the visual field examination showed a complete defect in the right eye (RE). A brain and spinal MRI displayed multiple enhancing cystic localizations in optic nerves and chiasma, pre-pontine, parasellar and basal cisterns with diffuse leptomeningeal spinal involvement. An endoventricular biopsy revealed a diffuse leptomeningeal spread of a BRAF negative low-grade glioneuronal tumor.
The CSF examination showed a blockage of CSF flow and presence of sporadic neoplastic cells. After 12 cycles of temozolomide standard schedule the brain and spinal cord MRI was stable but visual symptoms worsened, as confirmed by a campimetry assessment (Dec 2013). In January 2014 the patient began a treatment with bevacizumab 10 mg/kg every 2 weeks; after 3 months MRI showed a significant reduction (> 60%) of chiasmatic and parasellar enhancement with a dramatic improvement of visual acuity and visual field defects and a reduction of protein level in CSF.

Discussion. The distinctive characteristics of DL-GNT are the involvement of the basal cisterns and interhemispheric sulcus with a wide leptomeningeal spread and the absence of a well-defined intraparenchymal mass. Up to date, a selective involvement of the optic nerves and chiasmatic leptomeninges has not been described in the literature, thus far representing a new type of clinical presentation. Radiotherapy and several chemotherapies have been used to control leptomeningeal spread without efficacy. Bevacizumab has been proposed as a treatment for non-glial tumors with some results [1]. In our patient we observe a normalization of the blood brain barrier permeability resulting in a reduction of chiasmatic enhancement on MRI and protein level along CSF with a dramatic neurological improvement.

Conclusion. This is the first report in the literature of an adult DL-GNT with predominant optic leptomeningeal involvement with a clinical and neuroimaging response to bevacizumab.



Brain MRI and visual field after 12 cycles of temozolomide (Dec 2013)



Brain MRI and visual field after 12 cycles of bevacizumab (Mar 2014)

LCR	DEC 2013	APR 2014
Glucose	111 mg/dl	80 mg/dl
Total protein	1968 mg/dl	762 mg/dl
Cells	16/mmc	3/mmc
Cytology	+	+/- (infammatory)

LCR test before and after bevacizumab treatment

References.
1. Trevisan E, Bertero L, Magistrello M, Rudà R, Soffietti R, Is there a Role for Bevacizumab in Non-Glial Tumors?, Curr Drug Targets, 2015; 16(7):684-8.

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