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Optic Pathway Glioma: Optic Nerve Glioma Revealed by Exophthalmos: A Case Report

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Abstract | **Case Report**

Optic nerve gliomas are rare tumors that are mainly seen in children. Exophthalmos and strabismus are the main telltale signs of the disease. Optic nerve metastases are very rare and often associated with significant metastatic spread. Imaging, particularly magnetic resonance imaging, is of great benefit in the diagnosis and monitoring of these tumors. Therapeutic management of these gliomas involves different means: surgical excision, chemotherapy, radiotherapy or abstention under supervision. Indications must be discussed on a case-by-case basis.

Keywords: Optic nerve glioma (ONG), Exophthalmos, Magnetic resonance imaging (MRI), Pilocytic astrocytoma, Neurofibromatosis type 1 (NF1).

INTRODUCTION

Optic nerve glioma (or ONG for Optic Nerve Glioma) is the most common tumor of the optic nerve and most often affects children between 2 and 7 years of age. It is associated in 30 to 50% of cases with neurofibromatosis type 1 (NF1).

The most common histological type corresponds to a low-grade, slow-growing pilocytic astrocytoma, frequently associated with chiasmatic involvement or other tumors such as meningiomas or hamartomas in NF1.

Decreased vision and exophthalmos are the most common clinical signs.

CASE REPORT

A 12-year-old child patient with no particular pathological history who presenting with exophthalmos, ocular redness associated with decreased visual acuity in the left eye.

A cerebral CT scan revealed a lesion of the left optic nerve measuring 30.8x20x17 mm, it is oblong with regular contours hypodense in spontaneous contrast enhanced heterogeneously after injection of contrast product.

It represses the eyeball responsible for grade 1 exophthalmos.

A brain and orbital MRI revealed grade 2 exophthalmos in the left orbit, a lesional process centered on the optic nerve which is swollen at the level of its intra-orbital portion, well limited fusiform, measuring 21x28 mm in width. heterogeneous signal, with cystic portions, it is in heterogeneous isosignal T1, heterogeneous hypersignal T2, heterogeneous intermediate signal on the diffusion with restriction of the ADC in places, enhanced by the PDC.

This process fills the intraconical fat and comes forward in contact with the posterior surface of the eyeball without loss of its sphericity, and remains posteriorly 4 mm from the optic canal.

It represses the oculomotor muscles with loss of the fatty interface of separation in places with the internal rectus muscle.

Right Orbit: No Anomaly
At The Cerebral Level:

Some nodular signal anomalies at the level of the bilateral right frontal parieto-occipital subcortical white matter, in T1 isosignal, T2 FLAIR hypersignal, without translation on diffusion, of non-specific nature.

Minimal Maxillary Sinusitis

Overall process centered on the left optic nerve, responsible for ipsilateral grade 2 exophthalmos, first suggesting a glioma of the optic nerve.

Nodular signal abnormality at the level of the subcortical supratentorial white matter which may be related to OBNI.

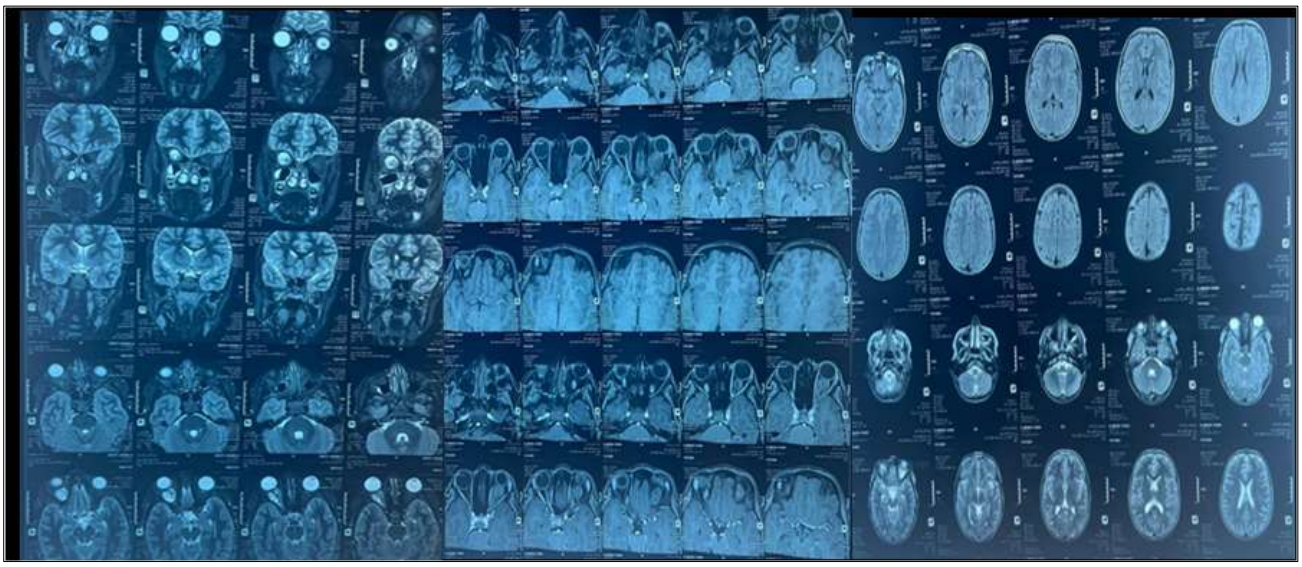

Figure 1: Brain and orbital MRI

A biopsy was done with anatomopathological study, spindle cell tumor process, immunohistochemistry, appearance compatible with a low-grade glial tumor.

The patient received primary chemotherapy according to the LGG protocol with radiological stability, then radiotherapy at a dose of 50.4 Gy with good tolerance.

DISCUSSION

Gliomas of the optic nerve are rare tumors which are mainly observed in children [1]. Bilateral optic nerve involvement is considered a characteristic of these gliomas in the context of neurofibromatosis type I (NF1) [2], as is the case in our patient. Exophthalmos and strabismus are often indicative of the disease. Magnetic resonance imaging is a key examination in the exploration of these gliomas. It not only allows us to study the optic nerves but also to look for possible extension of these gliomas to the orbit, chiasm and intracranial structures, particularly in the case of association with NF1 [3]. It also has the advantage of being less irradiating compared to CT scanning. Therapeutic management ranges from simple clinical and radiological monitoring, to surgical treatment or chemotherapy or radiotherapy. Therapeutic indications are discussed on a case-by-case basis, depending on visual acuity, the degree of exophthalmos and the extension of the tumor [4]. In the case of NF1, radiotherapy is strictly contraindicated due to the complications it can cause [5].

CONCLUSION

Optic nerve tumors are a diagnosis that must be considered when faced with symptoms of progressive optic neuropathy. Management must balance effective treatment and visual preservation. Decisions are made in multidisciplinary consultation meetings where the role of the ophthalmologist is crucial in terms of diagnosis and the effectiveness of treatment.

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