UNILATERAL OPTIC NEUROPATHY REVEALING AN OPTIC NERVE MENINGIOMA

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Abstract

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Optic nerve sheath meningioma (ONSM) is a rare orbital tumor that can mimic inflammatory optic neuropathies. We report the case of a 32-year-old woman presenting with rapidly progressive unilateral visual loss, pain with eye movements, and papilledema. MRI revealed an enhancing perioptic meningeal lesion extending into the optic canal, consistent with ONSM. Visual evoked potentials and electroretinography confirmed marked right-sided dysfunction, while systemic evaluations were unremarkable. Given the acute decline and suspected inflammatory component, high-dose intravenous corticosteroid therapy was initiated. The patient showed significant visual improvement and MRI demonstrated clear regression of the optic canal lesion after one month, with stabilization at 11-month follow-up. This case emphasizes the importance of considering meningioma in atypical optic neuropathies, particularly in young women with hormonal exposure, and highlights the potential benefit of corticosteroids when peritumoral edema is present.

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Introduction:-

Meningioma is a benign, slow-growing tumor that develops from arachnoid cells. It accounts for 13 to 18% of primary intracranial tumors depending on the series. (1) (2) ONM represent a small subset, 2%, of all orbital tumors. Most meningiomas that reach the orbit are an extension of intracranial meningiomas (90%). (3)

We present the case of an optic canal meningioma revealed by optic neuropathy, the evolution of which was marked by total regression under corticosteroid therapy.

Results:-

- 32-year-old woman

- Primary infertility for 10 years: salpingoplasty + HRT (IVF), phytoestrogen self-medication.

- Brain tumors in two paternal uncles

- Reason for consultation: rapidly progressive unilateral visual loss OD + pain on eye movement + paresthesia + chronic headaches

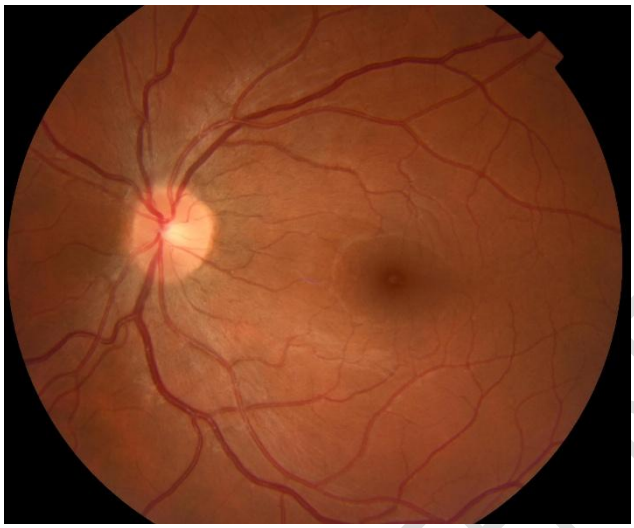
Examination:-

- VA: OD PL+, OG 10/10

- RPM + ODG

- Preserved eye motility

- SA: RAS

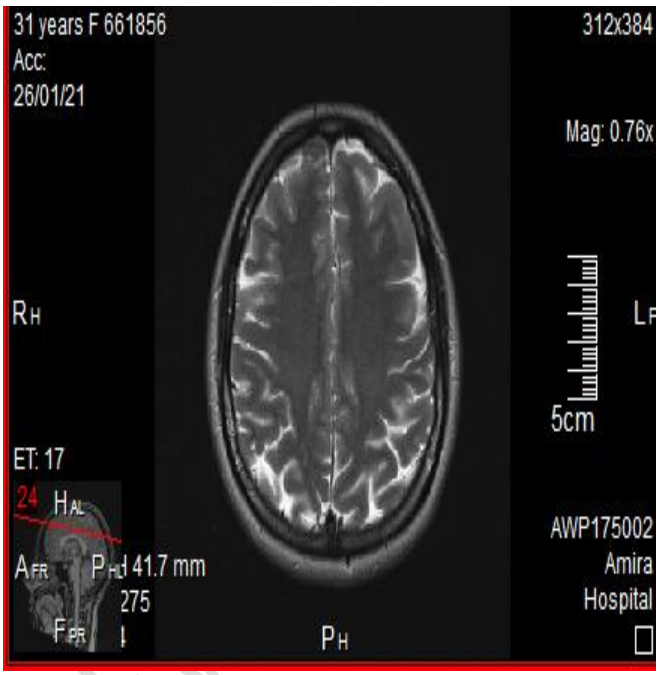


FO: OG RAS

OD: A grade III papilledema

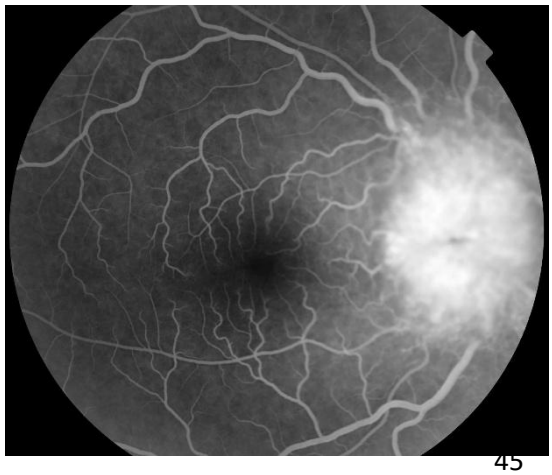
In view of this clinical picture, optic neuropathy of inflammatory etiology was considered.

- Orbital-cerebral MRI: T1 hypersignal enhanced by gadolinium at the level of the right perioptic meninges, suspecting a meningioma of the sheaths extending, invading the optic canal with the existence of an intracranial localization at the level of the cranial vault.

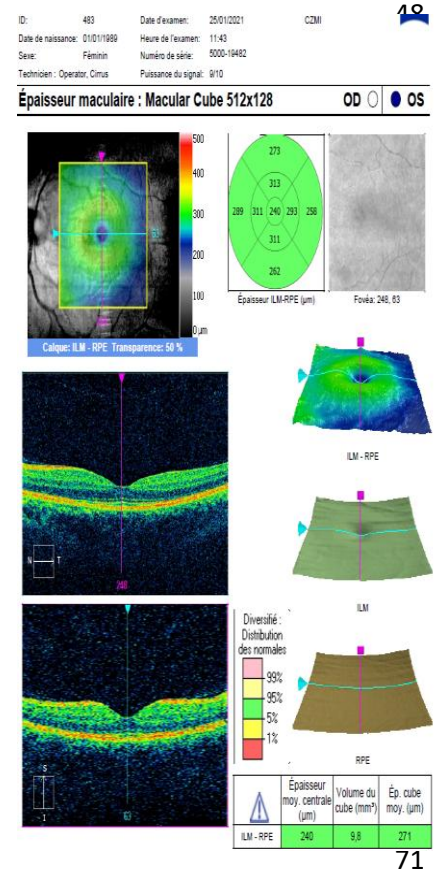


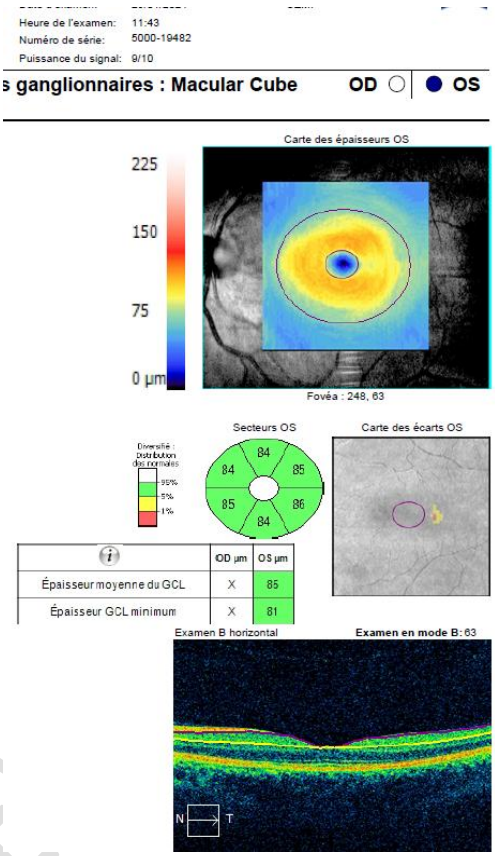
Assessments:

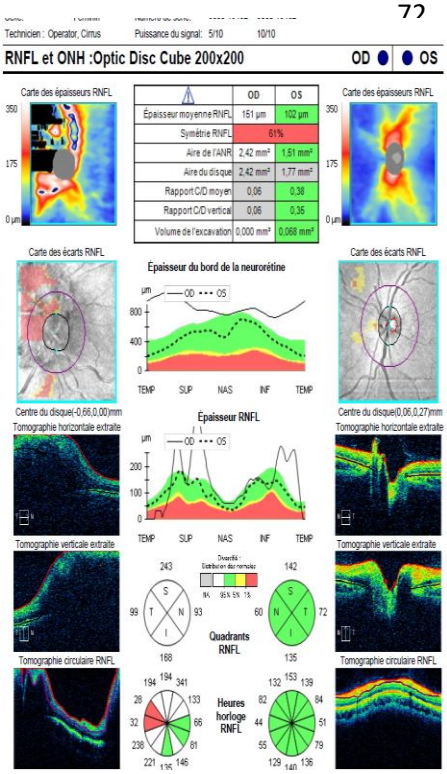
- Syphilitic serology, Lyme, normal ECA,
- Old immunity to toxoplasmosis
- IDR to tuberculin at 12 mm.
- The VEP was abolished on the right
- The ERG was altered on the right.



Fluorescein angiography and macular-papillary OCT confirmed the existence of papilledema in OD without other associated lesions.







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The general and in particular neurological examination was normal.

Therapeutic management

Given the low visual acuity, the young age of the patient and the pain on eye movement suggesting an inflammatory component:

We had indicated in consultation with the neurosurgery department.

A bolus of solumedrol 1mg / kg / day for three days with relay orally.

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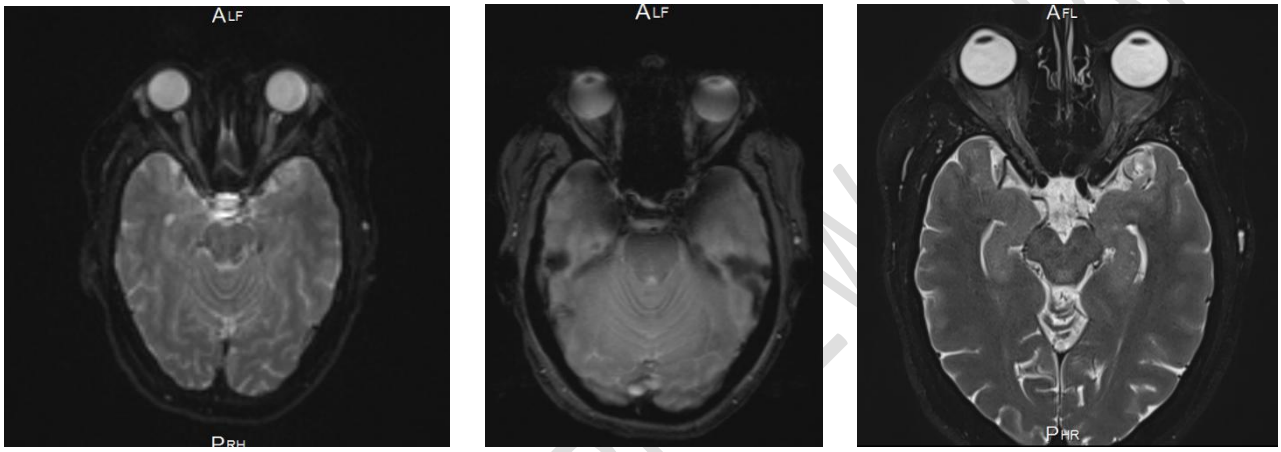
Treatment results:

The immediate post-bolus visual acuity rose to 5/10

(initially PL+)

On the Snellen scale without regression of the papilledema on fundus examination

An orbital-cerebral MRI control with sections centered on the optic nerve performed after 1 month revealed:



A clear regression of the optic canal meningioma and stabilization of the parietal

Patient under corticosteroid tapering currently. For a follow-up of 11 months

Discussion:-

The subacute onset of visual loss + pain on eye movement rhyme think: diagnosis of optic neuropathy.

these signs are explained by the involvement of the microvascularization of the optic nerve.

Symptomatology of meningioma

• A pathognomonic triad of optic nerve meningiomas:

Visual loss + optic atrophy + optociliary shunts

• Vision loss is extremely frequent (97%) including 24% CLD according to Wright

Meningioma and fertility treatment:-

- Hormone dependence of meningiomas: female predominance.

- Bickerstaff 1958, Mason in 1969 described a relationship between clinical manifestations and pregnancy.

- Carrol in 1993 demonstrated the presence of progesterone receptors in the membrane of tumor cells. favored by oestroprogestins.

- Therapeutic role of anti-progesterones (Sharif, 1998) in particular mifepristone (Grunberg, 1991) and anti-oestrogens (Oura, 2000), have not been confirmed by larger trials.

Discontinuation of oral contraception and hormone replacement therapy is indicated in all cases suspected of ONM.

MRI with Gado injection!

MRI is more effective than CT (especially for posterior extension)

fusiform lesion, with regular contours and sharp limits enhanced by the contrast product more importantly than the rest of the lesion, in the form of two parallel lines on the transverse sections, producing a railway track appearance.

Imaging should include thin axial and coronal T1 sequences (+/- sagittal), fat-suppressed T2 and post-contrast T1 with fat suppression.

T1: iso - intense to somewhat hypo-intense compared to the optic nerveT1 C + (Gd): homogeneous enhancementT2: iso - intense to somewhat hyperintense compared to the optic nerve.

ONM and Corticosteroids? Some trials

• The treatment of meningioma is not the subject of a well-established consensus. It depends on several factors: the location of the meningioma, its evolution, the patient's visual acuity, his age, the risks associated with surgery and the relationship with the cavernous sinus.

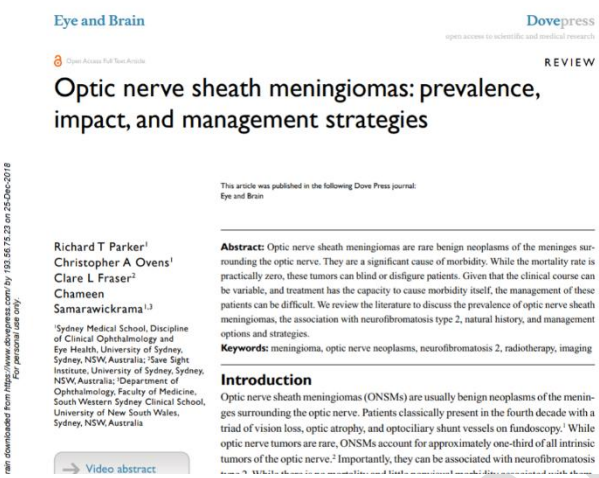
• According to Bouyana Oral corticosteroid therapy is sometimes prescribed in cases of acute visual acuity loss in order to act on the associated inflammatory component. Meningiomas do not respond to corticosteroid therapy except in cases of associated inflammatory component (peritumoral edema)

Meningioma and optic neuropathy: a diagnostic trap Optic neuropathy and meningioma: A diagnostic trap M. Bouyona,∗ , F. Blanc b, L. Ballonzoli a , M. Fleury b, C. Zaenker b, C. Speeg-Schatz a , J. de Seze b

Corticosteroids or carbonic anhydrase inhibitor?

In a study conducted by S. Mouton et al in the Neurology Department on six patients who received intravenous corticosteroid therapy, with methylprednisolone, one gram per day for three days. Complete recovery was observed in one case with an eight-year follow-up, partial and transient improvement was noted in two cases and worsening continued in four cases.

In the same series, some patients received symptomatic anti-edematous treatment such as acetazolamide. The treatment appears to be beneficial in cases of papilledema, two of the cases having been stabilized with a five-year follow-up.



Eye and Brain

Dovepress

REVIEW

Optic nerve sheath meningiomas: prevalence, impact, and management strategies

This article was published in the following Dove Press journal:
Eye and Brain

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Video abstract

Abstract: Optic nerve sheath meningiomas are rare benign neoplasms of the meninges surrounding the optic nerve. They are a significant cause of morbidity. While the mortality rate is practically zero, these tumors can blind or disfigure patients. Given that the clinical course can be variable, and treatment has the capacity to cause morbidity itself, the management of these patients can be difficult. We review the literature to discuss the prevalence of optic nerve sheath meningiomas, the association with neurofibromatosis type 2, natural history, and management options and strategies.

Keywords: meningioma, optic nerve neoplasms, neurofibromatosis 2, radiotherapy, imaging

Introduction

Optic nerve sheath meningiomas (ONSMs) are usually benign neoplasms of the meninges surrounding the optic nerve. Patients classically present in the fourth decade with a triad of vision loss, optic atrophy, and optociliary shunt vessels on fundoscopy.[1] While optic nerve tumors are rare, ONSMs account for approximately one-third of all intrinsic tumors of the optic nerve.[2] Importantly, they can be associated with neurofibromatosis

Orbital tumors are rare and ONM represent a small subset 2%

of all orbital tumors.

ONSM show a female predominance 61%.

and the average age of patients at diagnosis is around 40 years.

95% unilateral

90% are of intracranial origin.

Conclusion:-

Always consider meningioma in case of NORB (rare)

The background: young woman, hormonal disorders

Corticosteroid therapy is useful for the edematous component of the ophthalmic canal localization.

Interest of orbital-cerebral MRI to make the diagnosis

Interest of a larger and multicenter prospective study

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