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Optic Nerve Sheath Meningioma (ONSM): Case Report

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Abstract **Case Report**

Optic nerve sheath meningiomas are rare benign tumors of the central nervous system. The diagnosis of ONSM is confirmed with magnetic resonance imaging (MRI), especially with gadolinium-enhanced fat-suppression sequences. The MRI has become the gold standard for the diagnosis and has obviated the need for tissue biopsy.

Keywords: Optic nerve, meningiomas, MRI.

INTRODUCTION

Optic nerve sheath meningiomas are rare benign tumors of the central nervous system. Although they are slow-growing, their location directly affects the anterior visual pathway and can lead to severe visual loss. The diagnosis of ONSM is confirmed with magnetic resonance imaging (MRI), especially with gadolinium-enhanced fat-suppression sequences. The MRI has become the gold standard for the diagnosis and has obviated the need for tissue biopsy. Treatment of this condition is controversial, as the proximity of the tumor to the optic nerve makes it difficult to completely remove it without causing secondary complications.

CASE REPORT

Patient 34 year, without particular pathological antecedent, which has for 2 years, a non-swing right exopthlamos with a decrease in visual acuity.

MRI study of the orbital cavities was performed. In the MRI protocol T1, T2, T1 with fat saturation, T1 with fat saturation sequences, befor and after gadolinium injection on axial, sagital and coronal planes were performed. A fusiform mass was demonstated in the right orbit showing widening along the length of the nerve sheath. The mass appeared as isointense to brain and optic nerve tissue on T1 weighted images (Fig. (Fig.1) and slightly hyperintense on T2 weighted images (Fig. (Fig.2). On T1 weighted images with fat saturation after intravenous administration of paramagnetic substance (gadolinium) the mass presented a homogeneous intense enhancement suggesting in appearance a "tram track" around the hypointense optic nerve (Fig. (Fig.3). There is no intracranial extension of the lesion (Fig. (Fig.1) or any evidence of surrounding structures invasion (Fig. (Fig.3). The presence of optic nerve sheath meningioma was assumed.

DISCUSSION

ONSM is usually localized in the intraorbital or intracanalicular portions of the optic nerve sheath and originates from the cap cells of the arachnoid surrounding the nerve. The intraorbital portion is the most common site of origin (92%). About 95% of ONSM is unilateral. Rare bilateral tumors tend to occur in patients with neurofibromatosis type 2 [1, 2].

Most tumors do not have a clear etiology, but they probably have a genetic influence. Meningiomas in particular can have an association with ionizing radiation and hormonal influences, although, there is not a clear association in ONSM.

ONSM constitute about 2% of all orbital tumors and 1% of all meningiomas [3, 4]. These tumors, like their intracranial counterpart, are predominant in middle-aged females. The mean age of presentation is usually less than for intracranial meningiomas because the visual problems make patients seek examination earlier. Although rare, they may be found in children but tend to be more aggressive. Approximately 5-6% of the cases are bilateral and associated with neurofibromatosis type 2 [3]. Of those meningiomas that involve the orbit, only

10% are of primary orbital origin, while the remainders are of intracranial origin extending into the orbit.

The natural history of ONSM is characterized by slowly painless and progressive visual loss in the affected eye. If left untreated, this tumor can lead to complete blindness. Visual loss, optic atrophy, and optociliary shunt vessels are the classic triad in patients with an ONSM but most patients do not present the three components [4, 5]. Proptosis and resistance to retropulsion are also other signs seen on examination, albeit not always present. Patients can also complain of transient visual loss associated with eye movement, or what is referred to as gaze-evoked amaurosis.

In the early stages, patients can present with chronic optic nerve edema. This occurs because the tumor compresses the intraorbital portion of the optic nerve. Eventually, the edema subsides, and optic pallor develops. Optociliary shunt vessels can develop in about 30% of patients as the optic disc edema resolves. These represent venous collaterals that are connected to the choroidal circulation. They appear after chronic central retinal vein obstruction, such as in ONSM, but can also be found in other conditions such as central retinal vein occlusion, optic nerve glioma, and sphenoid wing meningioma. When vision is affected, there is a relative afferent pupillary deficit in the involved eye. Visual field defects can be present in the affected eye and are not specific; these can be seen as altitudinal defects, generalized constriction, or enlarged blind spots.

The diagnosis of ONSM is confirmed with magnetic resonance imaging (MRI), especially with gadolinium-enhanced fat-suppression sequences. The MRI has become the gold standard for the diagnosis and has obviated the need for tissue biopsy. On MRI, they appear isointense to grey matter on both T1 and T2 weighted imaging. ONSM is a sensitive lesion to gadolinium contrast, demonstrating vivid enhancement which contrasts with the non-enhancing optic nerve. On MRI axial images, it will present with the characteristic "tram-track" sign, which corresponds to the enhancing outer ONSM encircling the inner non-enhancing optic nerve. On the coronal images, this will be seen as a "doughnut" or "non-enhancing dot" sign. Typical appearances of ONSMs on imaging are tubular expansion of the meninges surrounding the optic nerve (62%), globular (23%), fusiform (11%), and focal enlargement of the optic nerve (4%) [8]. An optic nerve glioma will demonstrate uniform enhancement of the optic nerve and will not have the classic tram track sign.

A computed tomographic (CT) scan of the orbit and head can show the bony anatomy and hyperostosis produced in secondary tumors. Calcifications can also be seen in the ONSM. CT scans can also demonstrate the classic tram track sign.

The mortality risk from ONSM is practically null. If the tumor is left untreated, progressive vision loss will occur. The tumor produces direct compression of the optic nerve and compromises its vascular supply, and in some cases infiltrates the optic nerve [6]. Most patients will progress to blindness in the eye. At that point a decision regarding surgery is assessed. For those cases where expert surgery is performed, there is approximately 33% - 50% chance for worsening the vision [6].

Tumor resection is almost impossible without incurring a severe visual loss, due to the intimal relationship of the ONSM to the optic nerve. However, surgical resection can be justified in cases of disfiguring proptosis where the visual function has significantly decreased or in cases of intracranial extension. If the eye with the ONSM is blind and the tumor is confined to the orbit the patient should be observed as they grow very slowly, although, some surgeons advocate surgically resecting the tumor to avoid extension to other areas. If the eye is blind and intracranial extension is present, the tumor and the nerve should be removed. Some authors favor surgical intervention as a primary treatment not only stop disease progression and reduce the risk of subsequent vision loss, but also, to some extent, reverse visual deficits that have already occurred [7].

Transnasal endoscopic optic nerve decompression has been recently proposed and has showed stabilization of the disease and in some cases improvement from the baseline visual deficit [9, 10].

Conventional radiotherapy has been used both pre-operatively and postoperatively for many years [3, 11]. More recently stereotactic radiotherapy (STR) has been employed as an alternative to surgery, and may well be superior. In cases where visual function decreases, SRT has become the modality of choice since it delivers the appropriate amount of radiation to the tumor in a localized fashion. Risks of radiation-induced retinopathy or optic neuropathy are prevalent [12, 13].

CONCLUSION

Optic nerve sheath meningiomas are best managed by an interprofessional team of neurosurgeons, neuro-ophthalmologists, radiation oncologists, and neurologists. The treatment depends on size, mass effect, and symptoms. Unfortunately despite optimal treatment, vision loss may be unavoidable in most patients. To improve outcomes, prompt consultation with an interprofessional group of specialists is recommended.

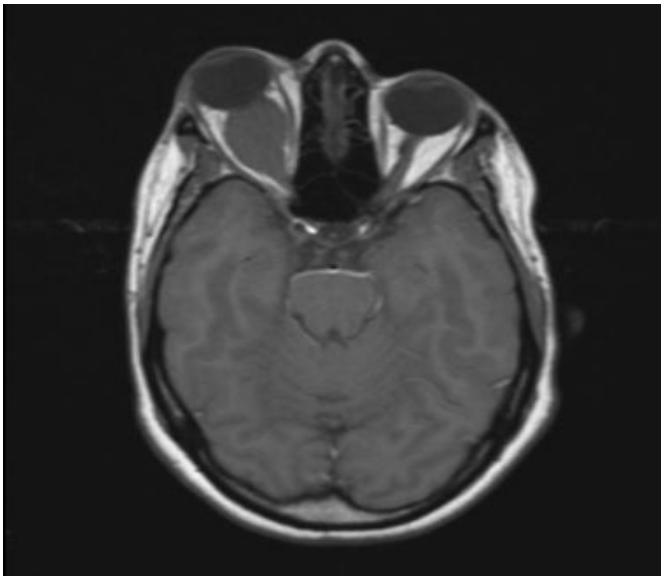

Fig-1: Axial MR image (T1 without fat saturation). An isointense to brain and optic nerve (arrow) lesion which produces exopthalmos. The lesion appears as marked widening along the path of the optic nerve but there is no intracranial extension

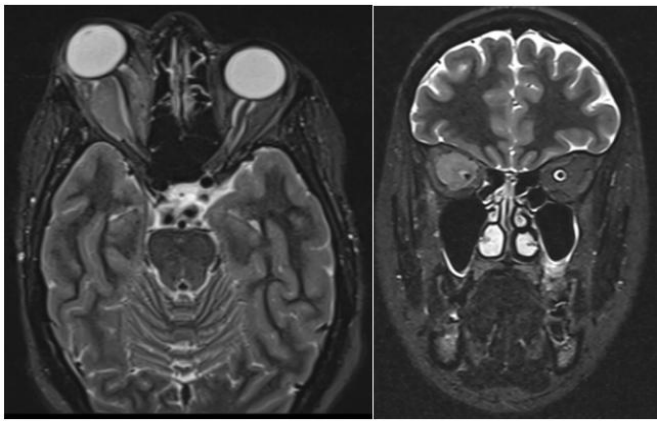

Fig-2: Axial (a) and coronal (b) MR image (T2 with fat saturation). The lesion is slightly hyperintense to the optic nerve (arrow)

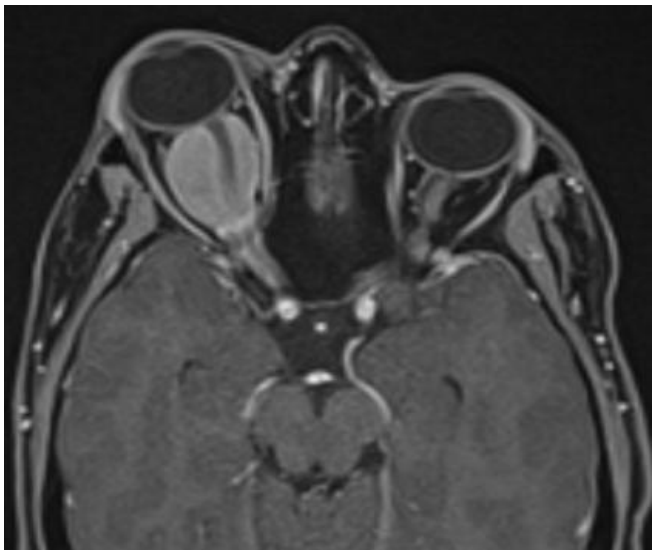

Fig-3: Axial MR image (T1 with fat saturation) after intravenous administration of paramagnetic media. There is homogeneous intense enhancement producing a "tram track" appearance around the hypointense optic nerve. Surrounding structures remain intact

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