

MedPeer Publisher

Abbreviated Key Title: MedPeer

ISSN : 3066-2737

homepage: <https://www.medpeerpublishers.com>

Optic Nerve Meningioma Revealed by Exotropia in a Young Patient: A Case Report

DOI: 10.70780/medpeer.000QGXI

AUTHOR AND AFFILIATION

A. Hajjaji¹, S.Sadiki², A. Bouzidi¹, A. El Ouafi¹¹ Department of Ophthalmology, Moulay Ismail Military Hospital, Meknes, Morocco² Department of Ophthalmology, Military hospital Mohammed V, Rabat, Morocco

Corresponding author: A. Hajjaji

ABSTRACT

Introduction: Optic nerve sheath meningioma (ONSM) is a rare benign tumor arising from the arachnoid cap cells surrounding the optic nerve. Although its course is usually slow, it is a serious cause of progressive visual loss because of its location in the confined orbital space. It is uncommon in young adults, in whom it may follow a more aggressive course.

Clinical case: We report the case of a 25-year-old man who presented with rapidly progressive visual loss and ocular deviation of the left eye. Examination showed a best-corrected distance visual acuity of 1/10 in the left eye, exotropia, a normal anterior segment, and diffuse pallor of the optic disc. Orbito-cerebral computed tomography (CT) revealed a focal fusiform thickening of the intraorbital optic nerve, 10.7 mm in size, involving its lateral aspect, with homogeneous contrast enhancement and punctate calcifications. Magnetic resonance imaging (MRI) showed an iso-intense nerve on T1-weighted images and slight hyperintensity on T2-weighted images; after gadolinium injection, the central nerve appeared hypointense surrounded by a markedly enhancing lateral thickening, consistent with the "tram-track" sign. The diagnosis of intraorbital ONSM was made on imaging alone, without biopsy. Because of the tumor's adherence to the optic nerve and the high risk of postoperative blindness, the patient was referred to radiotherapy.

Conclusion: ONSM should be considered in any young patient with unexplained visual loss, even when the presenting sign is an ocular deviation. MRI is the key diagnostic tool, and fractionated stereotactic radiotherapy is the treatment of choice for progressive forms.

KEYWORDS

optic nerve sheath meningioma; exotropia; tram-track sign; magnetic resonance imaging; stereotactic radiotherapy; young adult

MAIN ARTICLE

INTRODUCTION

Optic nerve sheath meningioma is a rare benign tumor that develops from the arachnoid cap cells surrounding the optic nerve. It is one of the main primary tumors of the optic nerve and accounts for approximately 2% of orbital tumors [1] and about one-third of primary optic nerve tumors [2,3]. Its growth is generally slow, but its location within the narrow orbital space and its tendency to compress optic nerve fibers make it a serious cause of progressive visual loss. The diagnosis rests mainly on imaging, particularly orbital MRI.

We report a case of intraorbital ONSM in a young adult, revealed by exotropia and rapidly progressive visual loss, and discuss its diagnostic and therapeutic challenges.

CASE PRESENTATION

A 25-year-old man presented to our department with rapidly progressive visual loss associated with deviation of the left eye. His general condition was good.

Ophthalmologic examination of the left eye showed a best-corrected distance visual acuity of 1/10 and exotropia (Figure 1). The anterior segment was normal. Fundus examination revealed diffuse pallor of the optic nerve head.

Orbito-cerebral CT showed a focal fusiform thickening of the intraorbital portion of the optic nerve (10.7 mm), involving the lateral part of the nerve. The lesion enhanced homogeneously after contrast injection and contained a few punctate calcifications. The sphericity of the globe, the orbital fat, and the extraocular muscles were preserved (Figure 2). These findings pointed first to an optic nerve sheath meningioma.

Orbital MRI showed a thickened optic nerve, iso-intense on T1-weighted sequences and slightly hyperintense on T2-weighted sequences. After gadolinium injection, the central part of the nerve appeared hypointense, surrounded by a strongly enhancing lateral thickening. The diagnosis of intraorbital optic nerve sheath meningioma was thus confirmed.

Given the adherence of the tumor to the optic nerve and the high risk of blindness after surgery, no biopsy or resection was performed, and the patient was referred to a radiotherapy department for further management.



Figure 1. Clinical photograph showing exotropia of the left eye.

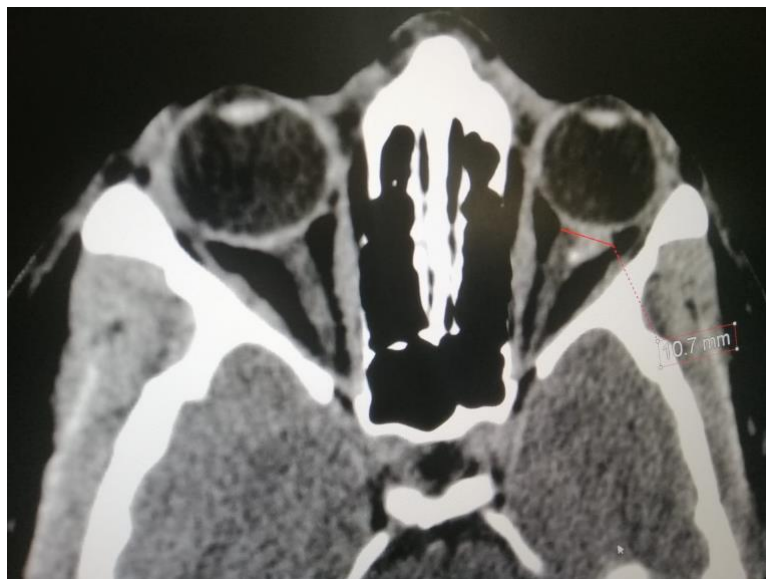


Figure 2. Orbito-cerebral CT showing fusiform thickening of the intraorbital optic nerve with homogeneous enhancement and punctate calcifications.

DISCUSSION

Optic nerve sheath meningioma is a benign tumor of arachnoid origin whose location in an anatomically confined space explains the potential severity of its visual impact. It is classically seen in middle-aged women [1,3]; its occurrence in a young subject, as in our patient, is less common and sometimes raises the question of a genetic predisposition such as neurofibromatosis type 2 [3,4]. In younger patients the tumor may behave more aggressively, making diagnosis and management more urgent [5,6].

Clinically, ONSM typically causes progressive and often insidious visual loss, sometimes accompanied by compressive signs such as proptosis or dystopia, or impaired ocular motility [1,3]. In our patient, the ocular deviation was the sign that drew attention, and visual loss was reported as rapidly progressive. The optic disc pallor observed in our case reflects advanced axonal damage and is a sign of poor functional prognosis. The slow progression of symptoms often delays diagnosis, so that many patients already have irreversible visual sequelae at the time of investigation.

Imaging plays a central role in positive diagnosis. CT shows a fusiform thickening of the optic nerve, sometimes with punctate calcifications, a frequent feature of meningiomas [3]. MRI is the reference examination, allowing precise assessment of the tumor, its extension, and its relationship with intraorbital structures. The "tram-track" sign, which reflects peripheral tumoral enhancement around a non-infiltrated central optic nerve, is considered almost pathognomonic [7–9]. The moderate T2 hyperintensity and homogeneous gadolinium enhancement in our patient match the classic description. Together, these findings allowed the diagnosis without biopsy, which is generally discouraged because of the high risk of iatrogenic injury to the optic nerve [3,10].

The management of ONSM remains controversial. Surgery was historically the main approach, but it has proved poorly effective in restoring vision and often harmful because of the intimate relationship between the tumor and the optic nerve fibers [1,3]. Resection is now reserved for very specific situations: advanced tumors with major orbital extension, intracanalicular forms threatening the chiasm, or an uncertain diagnosis. For most ONSM, radiotherapy, particularly conformal or fractionated stereotactic radiotherapy, has become the treatment of choice. It stabilizes tumor growth in more than 90% of cases and stabilizes or

even improves visual function in nearly 60% of cases when treatment is started early [11–14]. In our patient, radiotherapy was chosen for these reasons.

CONCLUSION

Intraorbital optic nerve sheath meningioma is a benign but potentially functionally devastating tumor because of its confined location and insidious course. Diagnosis relies mainly on imaging, especially MRI, which identifies typical features such as the tram-track sign and assesses tumor extension. Management is delicate: surgery, which carries a high risk to visual function, is now limited to few indications, whereas stereotactic radiotherapy is the reference treatment for progressive forms. This case underlines the importance of early diagnosis, multidisciplinary evaluation, and regular follow-up to preserve residual vision and prevent tumor progression. Visual prognosis depends largely on the stage at diagnosis, which calls for heightened clinical vigilance in any unexplained visual loss.

DECLARATIONS

Informed consent: Written informed consent for publication of this case and accompanying images was obtained from the patient.

Conflicts of interest: The authors declare no conflict of interest.

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