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Case Report

# Optic Nerve Sheath Meningiomas : A Case Report

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**Abstract:** Optic nerve sheath meningiomas (ONSMs) represent one-third of the tumors of the optic nerve. Histologically benign, they are called primitive when they are born in the orbit and extend along the optic nerve. The decrease in visual acuity dominates the clinical findings. Diagnosis is based on the characteristic clinical and imaging findings. Biopsies and surgical resection are associated with significant visual morbaidity. we report two cases of pericallosal lipomas admitted with seizures and discuss the clinical and radiological finding in the light of the litterature.

**Keywords:** Meningiomas, optic nerv sheath, benign, imaging.

## INTRODUCTION

Optic nerve sheath meningiomas (ONSMs) are rare tumors of the orbit and account for 1% to 2%of all meningiomas (Berete, R. *et al.*, 2006). However, they represent the second most common tumor of the optic nerve after gliomas, comprising about one-third of all primary optic nerve tumors. Importantly, they can be associated with neurofibromatosis type 2. Most ONSMs are secondary tumors that extend from intracranial sites, whereas primary ONSM arise from the intraorbital, or less commonly, intracanalicular dural sheath. CT scans and MRI both allow excellent visualisation of ONSM however MRI is considered superior and the gold-standard for confirming the diagnosis. Despite their classification as histologically benign tumours they cause progressive visual loss that often leads to blindness if left untreated.

## CASE REPORTE

A 34-year-old woman presented with slowly progressive rightsided unilateral visual loss over 03 years. Her birth history was unremarkable and she had no medical problems and no history or clinical signs of neurofibromatosis. CT scan demonstrated a calcified unilateral lesion confined to the optic nerve with no intracranial extension, consistent with an ONSM. Visual potentiel evoked showed a decrease of the conduction speed on the right. She was managed conservatively with regular clinical and radiological surveillance. After 01 years of follow-up she developed discreet proptosis and no perception to light on the right side. She had restricted upgaze on the right but all other extra-ocular movements were normal and ocular motility on the left side was not affected. Fundoscopy of the right eye demonstrated marked optic disc atrophy but the left fundus was entirely normal. In view of the progression she underwent a course of focused radiation therapy. Her scan 6 months following treatment demonstrated a slight reduction in tumour volume.

## DISCUSSION

In a review of 5000 orbital meningiomas, Dutton reported that 90% were secondary tumours resulting from intracranial extension and 10% were primary orbital tumours, 96% of which originated from the optic nerve sheath and 4% from other ectopic locations within the orbit. Of the primary ONSM reported, 92% had an origin within the intraorbital nerve sheath whilst only 8% were intracanalicular in origin. Most ONSM are unilateral (95%) however 65% of bilateral lesions are intracanalicular (Dutton, J. J. 1992).

ONSM typically affect middle-aged women, however the age of presentation of ONSM is slightly younger than for other meningiomas (Shapey, J. *et al.*, 2013).

A female predomiannace in ONSM, is widley accepted (61%), and pregnancy may further accelerate the growth of these tumours (Dutton, J. J. 1992) Bilateral and multifocal ONSM are more likely to

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present in childhood, frequently within the first decade of life,9 and are most commonly found in patients with neurofibromatosis type 2 (Spencer, W. H. 1972).

The classic clinical presentation of ONSM is the triad of progressive visual loss, optic atrophy, and the presence of retinalchoroidal collaterals.3 However, the simultaneous occurrence of all 3 findings is rare (Kahraman-Koytak, P. *et al.*, 2018) Visual loss is the most frequent symptom of ONSM and patients usually complain of decreased colour vision or blurred vision in the affected eye. Visual loss is painless, gradual, and progressive, and is typically present for 1–5 years before presentation.

It commonly progresses to complete blindness if left untreated (Wilson, W. B. 1981).

Proptosis is is usually slowly progressive and generally mild (2–5 mm), present in 30% to 40% of patients and less frequently seen in patients with canalicular lesions. Strabismus is (47%), usually in attempted upgaze (Miller, N. R. 2004).

In 98% of patient with ONSM, optic disc abnormalities are visible at the time of presentation (Dutton, J. J. 1992). Tumours originating at the apex of the orbit or within the optic canal may not have any orbital signs, and will often present with slowly progressive visual loss and a normal-appearing optic disc (Miller, N. R. 2004).

CT scans and MRI both allow excellent visualisation of ONSM however MRI is considered superior and the gold-standard for confirming the diagnosis (Goldsmith, B., & McDermott, M. W. 2006).

CT scan demsonstrate enlargement of the optic canal and calcification of the nerve sheath which is reported to occur in 20–50% of patients (Saeed, P. *et al.*, 2003).

Contrast-enhanced CT scans can also demonstrate the classic tram track sign, in contrast to an intrinsic optic nerve tumor such as an optic nerve glioma, which will demonstrate relatively uniform enhancement of the optic nerve itself. The calcification of ONSMs can give a tram track appearance on CT in the absence of the administration of intravenous contrast. While soft tissue contrast of CT scanning is inferior to MRI, CT remains superior for the assessment of calcification and of bony anatomy (Kanamalla, U. S. 2003).

Meningiomas typically display intense homogenous enhancement with gadolinium on MRI and gadolinium- enhanced fat-suppression T1-weighted pulse sequences. On these MRI sequences, the increased intensity of the enhancing tumour is readily separated from the non-enhancing optic nerve (Mafee, M. F. *et al.*, 1999).

Khoo *et al.*, (2000) also found that MRI-defined meningioma volumes were larger but inclusive of CT scan-defined volumes, suggesting complementary spatial information and a role for treatment planning with composite CT scan/MRI volumes (Khoo, V. S. *et al.*, 2000).

Ultrasound can be used to demonstrate the tumor if it is located anteriorly, and blood flow within it can also be demonstrated with Doppler scanning. This imaging modality is noninvasive and does not require the prolonged immobilization of an MRI scan, and thus may be more appropriate for monitoring known lesions in children (Garcia, J. P. S. *et al.*, 2005).

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%) (Saeed, P. *et al.*, 2003). “Tram tracking”, with the meningioma hyperdense (or hyperintense) on either side, relative to the optic nerve in the center is a classic sign and is demonstrated in 24% of tumors. Most tumors have smooth margins (80%). Some tumors, however, demonstrate alternative growth patterns to these (Parker, R. T. *et al.*, 2018; Samarawickrama, C. *et al.*, 2016).

In patients with known ONSMs, mfVEP (Multifocal visual-evoked potential) can be used to monitor for functional compromise and progression of visual decline without the need for MRI (Jayanetti, V. *et al.*, 2018).

The slow development and growth of these tumours means that observation without treatment can be justified in some selected patients. Observation alone may be a useful approach in patients with high functional vision or in patients with negligible visual decline, as well as when the tumour is situated near the orbital apex [17].

Kennerdell *et al.*,. recommended that treatment should be initiated when visual acuity deteriorates below 20/40 or when the visual field starts to constrict and others have similarly advised to commence treatment when patients begin to experience progressive visual loss (Kennerdell, J. S. *et al.*, 1988; Turbin, R. E., & Pokorny, K. 2004).

Surgical resection is rarely indicated in the management of primary ONSM except in patients with aggressive tumours with intracranial extension, in order to prevent spread to the contralateral optic nerve.6,9 Resection may also have a role if the patient suffers from disfiguring proptosis or blindness (Eddleman, C. S., & Liu, J. K. 2007; Kim, J. W. *et al.*, 2005).

ONSM were previously considered to be unresponsive to radiation therapy but Smith *et al.*,. were the first to clearly document the effectiveness of

radiotherapy in the treatment of primary ONSM (Smith, J. L. *et al.*, 1981). stereotactic radiosurgery describes the delivery of a single high dose of radiation using a rigidly attached stereotactic guiding device and/or stereotactic image guidance system. This delivery method achieves a much higher degree of target conformity than conventional radiotherapy techniques whilst minimising radiation exposure to surrounding normal tissues such as the retina, lens, pituitary gland, and optic nerve (Shapey, J. *et al.*, 2013).

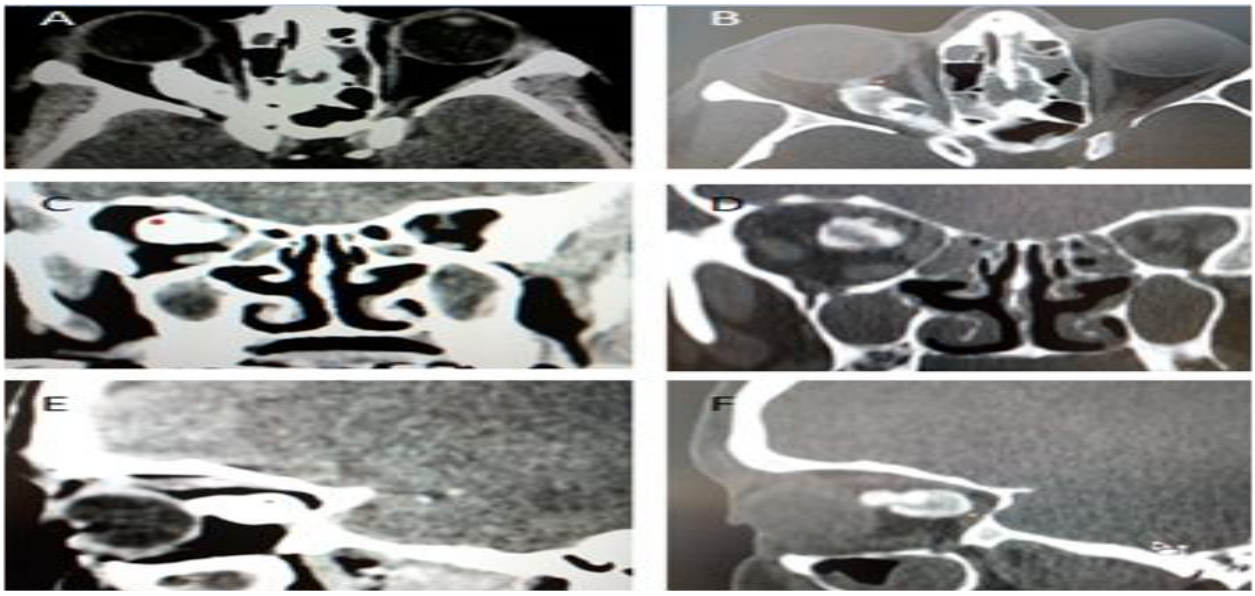
Turbin's review of longterm outcomes of patients with ONSM in 2002, proved influential. Visual outcomes for 64 patients were reported comparing surgery, observation, radiotherapy, and combination surgery and radiotherapy (Turbin, R. E., & Pokorny, K. 2004). The visual outcomes in the radiotherapy-only group were superior to all others, with thisgroup only showing no significant decline in visual acuity from diagnosis to last follow-up. Furthermore, the radiation group demonstrated a favorable complication rate of 33.3% (including radiation retinopathy, vascular occlusion, persistent iritis, or temporal lobe atrophy), compared to the complication rate of surgery of 66.7%. Similar results have been demonstrated in other series (Adeberg, S. *et al.*, 2011).

## CONCLUSION

Optic nerve sheath meningiomas are rare tumours but despite their benign histological classification, the majority of patients will eventually lose vision in the affected eye if left untreated. Furthermore, the clinical course of these tumours is highly variable and the unpredictable natural history makes patient management problematic with earlier detection with improved imaging technology, more accurate radiotherapy delivery, and improved case selection for surgical management, it is hoped that outcomes will continue to improve for these patients.

**Conflict of Interest**

The authors declare that they have no conflict of interest.




**Figures**

**Axial orbital pre-enhanced CT scan in parenchymal (A) and bony (B) windows, showing an eccentric ONSM with typical tram-track sign calcifications (B). There is no intracranial extension of the tumour nor bone involvement. Coronal orbital CT scan in parenchymal (C) and bony (D) windows, demonstrating the circumferentiel caractere of the lesion. Sagital orbital CT scan in parenchymal (E) and bony (F) windows revealing enlargement and linear calcification surrounding the optic nerve.**

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