Case Report

# A Rare Case Report on Unilateral Optic Nerve Sheath Meningioma

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## Abstract

Optic nerve sheath meningiomas (ONSM) are rare, slow-growing benign tumors, which approximately constitute 2% of all orbital tumors of the anterior visual pathway and 1-2% of all meningiomas. The middle-aged females are primarily affected. The primary orbital meningiomas originate from the optic nerve sheath or from the extension of an intracranial meningioma into the orbit. ONSM left untreated always leads to progressive defective vision, color blindness and complete loss of vision, which is often associated with optic atrophy. Most of the time management is conservative in long-standing ONSM, as there is no much of visual improvement after the surgery. Here, we present a case of a 55-year-old female with unilateral optic nerve sheath meningioma. The diagnosis was delayed more than 6 years from initial symptoms.

**Keywords:** Contrast-enhanced computed tomography, Meningioma, Optic nerve, Optic nerve sheath meningiomas

## INTRODUCTION

Optic nerve sheath meningiomas (ONSM) are slow growing benign, and typically unilateral tumors that arise from the arachnoid cap cells around the intraorbital portion of the optic nerve and are intimately associated with the optic nerve. The natural history of primary ONSM involves indolent growth during a period of many years. Intracranial extension is rare. ONSM is confined to the dura matter and hence it often appears as a well-defined, tubular thickening of the optic nerve on computed tomography (CT) or magnetic resonance imaging. Definitive treatment of ONSMs is challenging, however, because of the lesions intimate circumferential relationship with the optic nerve and its vascular supply. Surgical excision has almost always resulted in post-operative blindness in the affected eye and is reserved for patients with intra cranial extension of tumor.[1]

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## CASE REPORT

A 55-year-old south Indian female presented to ophthalmology outpatient Department complaining of gradual decrease in vision in the left eye and drooping of upper eyelid since 5 years with a history of mild intermittent headache. During this period, she has been treated elsewhere with different topical medications, details not provided. She did not give any previous history of eye pain, eye trauma or projectile vomiting. Patient is not a known diabetic or hypertensive.

Patient underwent detailed examination, ocular, medical and neurological from which the following observations were made. The best-corrected visual acuity was a perception of light in left eye without showing any improvement. The pupillary reaction was sluggish to direct light in left eye. Extraocular eye movements were restricted in all directions (Figure 1). Anterior segment examination under slit lamp was normal. The funduscopic examination showed optic atrophy with retino-choroidal collaterals in the left eye, and the right eye optic disc showed mild temporal pallor (Figure 2). Contrast-enhanced CT (CECT) brain study revealed evidence of hyper-dense mass lesion showing foci of calcification arising from left optic nerve sheath, suggestive of optic nerve sheath meningioma (Figure 3). The tumor had an intracranial extension along the

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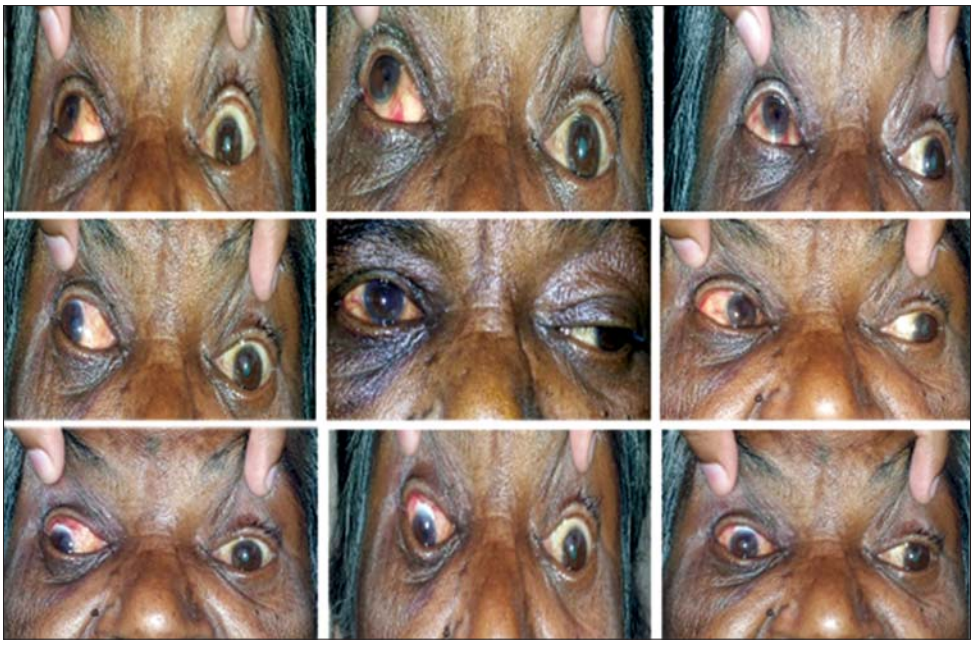

**Figure 1: Nine cardinal positions of gaze**

optic nerve, so the case was referred to neuro center for intracranial approach for excising the tumor, as the optic nerve showed signs of atrophy. Thou the tumor is benign it is necessary to approach it intracranial and excise, since it may produce complications of space occupying lesions.

**Figure 2: Primary gaze**

## DISCUSSION

ONSM are rare benign tumors of the optic nerve. 60-70% of cases occur in middle age females, and is more common in older adults (mean age 44.7 years). It is also seen in children, but this is rare. The tumors grow from cells that surround the optic nerve, and as the tumor grows, it compresses the optic nerve. This causes loss of vision in the affected eye. Rarely, it may affect both eyes at the same time.

It is typically a slow growing tumor and has never been reported to cause death. However, there is concern that the tumor can grow into the brain and cause other types of neurological damage. In some patients, the tumor grows so slowly that the treatment is not necessary.[2-5]

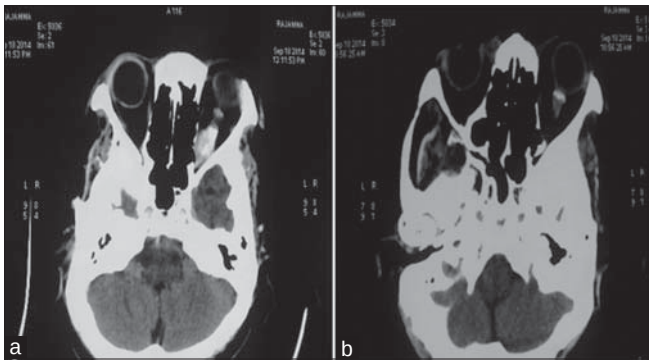


**Figure 3: (a and b) Hyperdense mass lesion showing foci of calcification arising from left optic nerve**

## CONCLUSION

Often meningiomas show homogenous and well-defined mass on CECT. CECT is the procedure of choice for diagnosis of ONSM. The findings in optic nerve sheath meningioma in this case are quite typical. The benign non-invading, well-defined growth pattern, the clinical manifestation strongly suggests the possibility of meningioma. As the patient's age is relatively advanced, aggressive procedure such as biopsy was considered unnecessary.

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**How to cite this article:** Padmini HR, Prabhu P. A rare case report on unilateral optic nerve sheath meningioma. Int J Sci Stud 2014;2(9):100-102.

**Source of Support:** Nil, **Conflict of Interest:** None declared