

# Unilateral Optic Nerve Sheath Meningioma: A Rare Case Report

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**Abstract:** *Optic nerve sheath meningiomas (ONSM) is a rare benign tumor of eye. On having a clinical suspicion, diagnosis is usually established by contrast MRI/CT scan. Here we present a case of 35-year-old female who presented with unilateral total ophthalmoplegia, mild proptosis and progressive decrease of vision. It was later diagnosed as a case of optic nerve sheath meningioma on MR imaging.*

**Keywords:** optic nerve sheath, meningioma, case report

## 1. Introduction

Optic nerve sheath meningioma (ONSM) is a rare benign neoplasm of central nervous system and arise from cap cells of the arachnoid mater around the nerve.[1]They most commonly present in patients between third and fifth decade with a female predominance in the ratio of 3:1.[2] ONSM's are usually unilateral and account for about 95% of cases. Bilateral tumors are usually associated with neurofibromatosis type II.[3]These lesions can arise primarily from the optic nerve sheath or may involve the optic nerve secondarily after arising from the cavernous sinus, falciform ligament, sphenoid wing, pituitary fossa, planum sphenoidale, front parietal area, or the olfactory groove.[4]

## 2. Case Report

A 35-year female presented with gradual progressive painless diminution of vision, progressive drooping of right upper eyelid and mild protrusion of right eye since last 2 years. On clinical examination her visual acuity was perception of light in right eye and 20/20 in left eye. Right eye had moderate ptosis with restriction of movements in all directions of gaze.(Figure 1) Movement of left eye were full and free in all directions of gaze and position of lid was also normal in left eye. The right eye pupil had RAPD, fundus showed total optic atrophy in right eye. In left eye, anterior segment examination and fundus examination were within normal limits. Systemic examination revealed uncontrolled Diabetes Mellitus and no evidence suggestive of neurofibromatosis type II.



**Figure 1:** Extraocular movements in all gazes

MRI orbit (Figure 2) showed large (57 x 42 x 42 mm) plaque like soft tissue mass lesion seen in right orbit appearing hypointense on T1 and hypo to iso intense on T2 weighted imaging and showed avid post contrast enhancement. The lesion was predominantly situated in posterior part of extraconal space and inseparable from extraocular muscle with encasement of right optic nerve. The lesion was extending into ethmoid air cells and posterior nasal cavity medially. Inferiorly the lesion was extending into retroantral space and maxillary sinus. The lesion also involved right cavernous sinus. Laterally the lesion was causing mass effect on medial temporal lobe on right side. Posterolaterally, the lesion was causing widening of foramen ovale and foramen lacerum.

On CT cuts of orbit, lesion appeared hyperdense on NCCT and showed thinning and focal erosion of bones at places with hyperostosis of involved bones.

The patient was referred for radiation therapy. The surgical intervention was not advised to our patient because of extensive involvement. Since total optic atrophy had already developed in right eye, preservation of vision in left eye was of prime importance. The patient however did not show up and was lost to follow up after referral for radiation therapy.

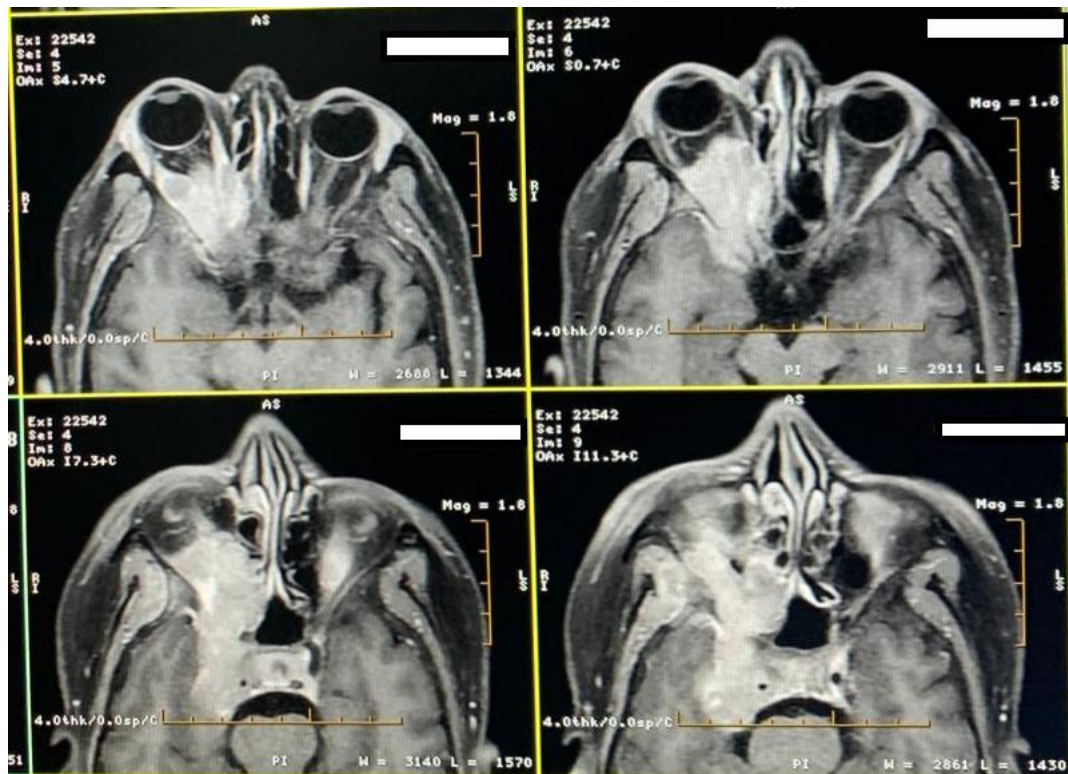


Figure 2: MR images of orbit

### 3. Discussion

Once an ONSM is suspected, it can usually be diagnosed using MRI or high-resolution CT scan. The definitive treatment of ONSM's is challenging because lesion is closely related to the optic nerve and its vascular supply. Surgical excision mostly results in blinding of affected eye. This could be due to excision of the tumour along with the affected optic nerve intraoperatively or damage to the pial vasculature.[5] Conservative management is therefore indicated if there is no significant progressive visual dysfunction or intracranial extension of the tumour. Recently it has been highlighted by Miller that surgery to remove an ONSM is rare if ever indicated and radiation is the optimum therapy. [6]

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