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Case Report

# Isolated 3rd Nerve Schwannoma: A Rare Entity; Mimicking Middle Fossa Meningioma and Review of Literature

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## Abstract

Cranial Schwannomas affecting the motor cranial nerves in absence of neurofibromatosis type II are quite rare as it generally affects the sensory cranial nerves sporadically. Therefore, oculomotor schwannomas as such are rare. In this article, we present a case of middle-aged adult who presented with a diplopia and progressive proptosis and ptosis which was neglected in view of the COVID pandemic. In this article, we present a case of orbito-cavernous type of schwannoma which is the rarest entity amongst all type of oculomotor schwannomas along with review of the appropriate literature.

**Keywords:** Orbito-Cavernous; Oculomotor Schwannoma; Diplopia; Covid; Proptosis; OCM; Oculomotor

## Introduction

Intracranial schwannomas are benign tumours and majority of them arise from the sensory or mixed nerves such as vestibular and trigeminal. Schwannoma of oculomotor nerve (OMS) is unique in the sense that oculomotor is purely a motor nerve. The course of this nerve is long originating from brainstem, traversing through perimesencephalic cistern, oculomotor triangle, lateral wall of cavernous sinus, superior orbital fissure and orbit. There have been about sixty cases reported in the literature so far, and out of them only 9 cases of orbito-cavernous type have been described. We are reporting a rare case of orbito-cavernous type of oculomotor schwannoma , this is a unique case as all the cases reported have been diagnosed based on the clinical manifestation of 3rd nerve, none by intra-operative origin of the tumour. In this case, besides its rare location; we could locate the origin of the tumour from the nerve in the orbit. Of the 52 lesions reported in the literature, 22 (42%) were based in the cisternal space, 16 (30%) were entirely in the cavernous sinus, 11 in the cavernocisternal space, and 3 in the orbito-cavernous region [1].

In the present case report, we describe an oculomotor nerve schwannoma (OMS) of the rare orbito-cavernous type (Type 3) [2] which was mistaken as an atypical meningioma and patient was neglected in view of the COVID pandemic amounting to the vision loss in the patient.

## Case Presentation

A 52 years old patient from a remote area in a village presented with complaints of diplopia and progressively increasing proptosis of the right eye over last 1 year with deterioration of vision in the same eye for 8 months. The patient had undergone some ophthalmological procedure 6 months back for the same but showed no improvement. There was no vision in right eye for 6 months. Examination revealed no perception of light in right eye with ptosis in the same eye. Eye ball was rotated outwards, downwards and fixed i.e. no extra-ocular movements. She has complete paralysis of the eye movements and pupil was fixed and dilated (Figure 1,2). Gadolinium contrast enhanced MRI brain with orbital cuts was done which showed a well defined extra-axial lobulated lesion in the right middle fossa from the medial sphenoid wing extending anteriorly into the right orbit displacing the eyeball outwards and medially. Mass was extending into seller region eroding the anterior clinoid process, completely encasing the right ICA with invasion into the ethmoid sinus. The lesion was T1- hypo-intense, T2- heterogeneously hyper-intense with heterogeneous contrast enhancement. The differential diagnosis included an atypical meningioma, fungal mass or schwannoma (Figure 3,6).

**Figure 1**

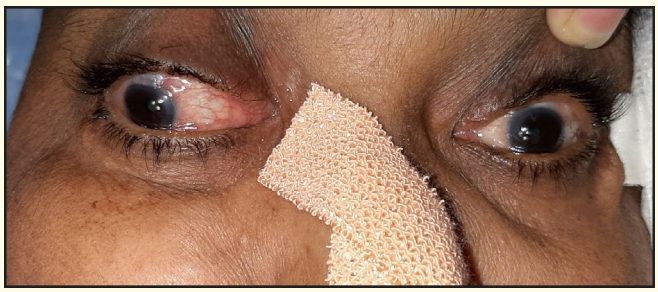

**Figure 2**

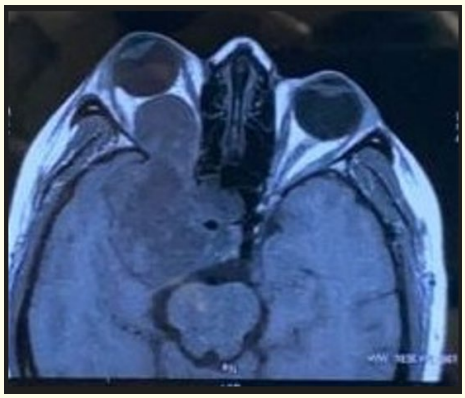

**Figure 3**


**Figure 4**

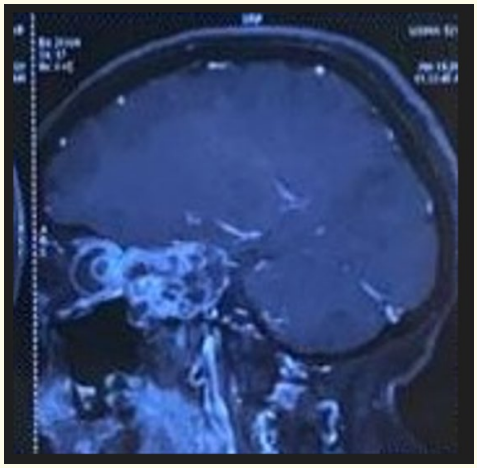

**Figure 5**

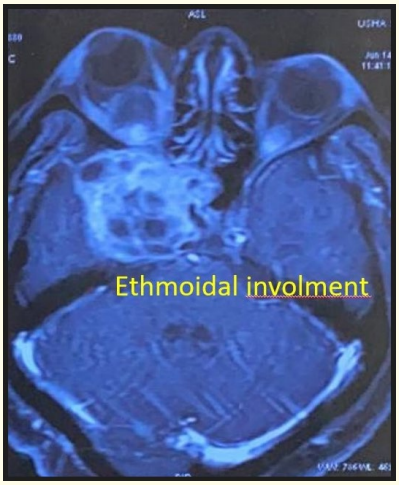


**Figure 6**

Patient was planned for right pterional orbitozygomatic osteotomy, anterior clinoidectomy with deroofing of optic canal and tumour removal through sub-temporal interdural supraorbital approach. Tumour was in interdural plane in middle fossa, cavernous sinus going to orbit through SOF and was found to be arising from the oculomotor nerve. Tumour was greyish-white, soft, suckable, mildly vascular with well defined plane of cleavage encasing the ipsilateral ICA. Tumour was dissected from the oculomotor nerve and nerve was skeletonized. Optic nerve could be seen separately (Figure 7). Postoperatively patient's 3rd nerve deficit increased and she developed complete ptosis, else postoperative period was uneventful. Histopathology showed spindle cells arranged in a palisading fashion or in an organoid arrangement (Verocay bodies) with areas of cyst formation and varying areas of localised haemorrhage (Figure 8).

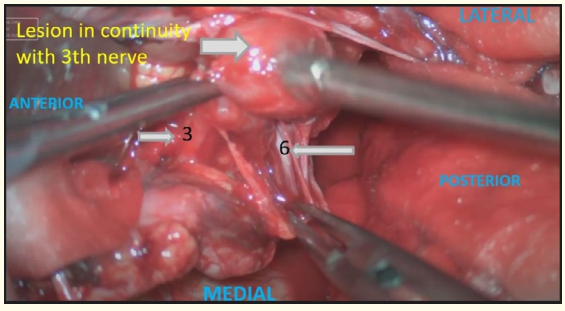


**Figure 7**

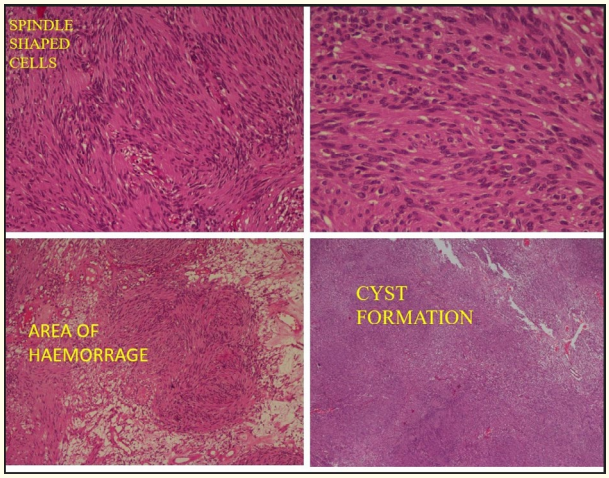


**Figure 8**

## DISCUSSION

Schwannomas are benign, encapsulated and slowly growing tumours arising from Schwann cells. Schwannomas can occur sporadically but are also associated with Recklinghausen's disease (NF-1). Intra-cranial schwannomas usually involve sensory nerves, most commonly involving the inferior division of the vestibular nerve and trigeminal nerve [3]. The highest reportage of OCM schwannomas is from the Asian region [3]. Almost all of the reported cases are histologically benign, there are only 3 cases of malignant OCM schwannoma. The oculomotor nerve exits the brainstem (mid brain) from the medial aspect of cerebral peduncle into the interpeduncular cistern, from where it travels between superior cerebellar artery (SCA) and posterior cerebral artery (PCA) and goes medial to the uncus in relation to medial border of the tentorium before piercing the roof of cavernous sinus. At the roof, it lies in oculomotor cistern. It then travels along the lateral wall of cavernous sinus in the inter-Dural plane and then enters the orbit through the superior orbital fissure.

The transition zone/root entry zone of the 3rd nerve is only 0.6 cms from the brainstem, hence OMS can occur over multiple locations, both intradural and extradural. The first case of third nerve tumour (ONS) was reported in 1927 by Kovacs [4] based on an autopsy finding and Celli., *et al.* [2]. classified them into 4 types based on the course of the oculomotor nerves as 1. Predominantly cisternal, 2. Predominantly cavernous, 3. orbito-cavernous, 4. Cisterno-cavernous.

Cisternal type being the most common and orbito-cavernous is the least common among these. This is perhaps due to the transition zone or root entry zone of the 3rd nerve is 0.6cms distal to its origin from the brain stem, hence ONS can occur at multiple locations, both intradural and extradural [1].

The presenting symptoms of oculomotor schwannoma include diplopia or ptosis (35%), headache or periorbital pain (29%), visual disturbance (13%), hemiparesis and gait disturbance (10%). Oculomotor nerve palsy was the most common preoperative neurological sign (87%). Other cranial nerves involved are trochlear nerve paresis (23%), trigeminal nerve (26%), and abducens nerve paresis (13%) [5-8].

Only 9 cases of orbitocavernous type of ONS have been reported in the literature (Table 1) and this being the 10th case to add to the literature.

| Author | Age/sex | Presenting complaints | Approach | Post-op outcome |
|---|---|---|---|---|
| Okamoto., *et al*.[9] 1985 | 52/f | Exophthalmos, convulsions, 3rd nerve palsy, diplopia | Fronto-orbital craniotomy- sub-total resection | 3rd nerve palsy present |
| Barat., *et al*. [10] 1992 | 27/F | Exophthalmos, visual impairment cranial nerves IV, V, VI paresis | Fronto-orbital craniotomy- sub-total resection | 3rd nerve palsy present |
| Ohata., *et al*. [11] 2006 | 63/F | Diplopia, ptosis, chemosis | Fronto-temporal- OZ craniotomy Sub-total resection of cystic tumor | 3rd nerve paresis occured |
| Hironaka., *et al*. [12] 2010 | 58/M | exophthalmos, blindness, pain | Frontotemporal-OZ craniotomy Gross-total resection | 3rd nerve paresis occurred which recovered |
| Futardo., *et al*. [5] 2011 | 25/M | exophthalmos, diplopia, complete 3rd nerve palsy | FT-OZ craniotomy Near-total resection | 3rd nerve palsy |
| Kauser., *et al*. [13] 2014 | 32/M | Exophthalmos | Frontal-orbital craniotomy and gross total excison | 3rd nerve palsy |
| P singh., *et al*. [14] 2016 | 28/M | exophthalmos, blindness, 3rd nerve palsy | Near total excision | Complete 3rd nerve palsy |
| Muhammad., *et al*. [15] 2019 | 64/M | exophthalmos, blindness, partial 3rd nerve palsy | Lateral supra-orbital approach Sub-total resection | Complete 3rd nerve palsy which partially recovered |
| Present case | 52/F | Proptosis, blindness, 3rd nerve palsy | Pterional orbitozygomatic craniotomy and gross total tumor excision | Complete 3rd nerve palsy |

**Table 1:** Reported cases of orbito-cavernous oculomotor schwannomas (ONS) in literature.

The choice of the surgical route is determined by the location, size, and the tumour extent. Various approaches can be used, including cranio-orbital approach with or without skull base exposure. Also, transconjunctival, supra-orbital and lateral orbital approaches can be used. Orbito-cavernous tumours are better approached through the roof of the orbit following an orbito-zygomatic approach, with a frontotemporal craniotomy, optic canal unroofing, and varying degrees of clinoidectomy [16,17]. The wide surgical field improves the outcome by limiting the damage to the surrounding structures and preventing too much traction of the fronto-temporal lobe over the dura. The risk of damage to nerve rootlets enmeshed within the lesion is high with attempts at near-total excision, rendering the anatomical preservation of the nerve an arduous exercise. Surgical intervention was conducted in all these cases and most of them underwent total or subtotal resection of the tumour. However, oculomotor function was not improved or recovered only partially postoperatively in the majority of those cases. In our case also, the patient had gross total tumour excision, however, the oculomotor palsy was persistent post-operatively.

Therefore, surgical excision of the OCM is only recommended only for large tumours with intractable symptoms [18,19]. It is also possible to observe the natural course of the tumours in the cavernous sinus using MR image until the tumour becomes large or stereotactic radio-surgery may be an alternative to radical resection [20]. There have been only 2 reported cases in which total resection of oculomotor schwannoma was done without permanent 3rd nerve palsy [18].

## Conclusion

Due to the rarity of the ONS, these tumours are misdiagnosed and sometimes even wrongly treated leading on to catastrophic results to the patients. Clinic-radiological correlation with adequate anatomical knowledge of the orbito-cavernous region is utmost important to provide best possible surgical outcome to the patient of extra large orbitocavernous lesion.

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