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*Case Report*

# Ectopic intraconal orbital meningioma – A rare case report

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Received : 28 January 2021
Accepted : 04 June 2021
Published : 28 June 2021

**DOI**
10.25259/SNI_84_2021

**Quick Response Code:**

## ABSTRACT

**Background:** Ectopic orbital meningiomas (OM) are a rare subset of OMs which are neither attached to the optic nerve sheath nor to the surrounding bone.

**Case Description:** We report the case of a 65-year-old female who presented with a 1 year history of proptosis followed by visual loss and restricted right eye movements since 3 months. Radiology of the orbits was suggestive of intraorbital, intraconal, and heterogeneous contrast enhancing right eye lesion which was completely excised through supraorbital orbitotomy approach. Intraoperatively, the right optic nerve, though compressed, and displaced inferiorly, was free from the lesion. The final histopathological diagnosis was "Meningioma WHO Grade I." At 3 months follow-up, patient's vision in the right eye improved from perception of light positive to 6/12 and there was no evidence of recurrence.

**Conclusion:** Rarity of ectopic OM, total surgical excision with an excellent postoperative visual outcome prompted us to report this case.

**Keywords:** Ectopic orbital meningioma, Intraconal, Optic nerve, Proptosis, Visual loss

## INTRODUCTION

Orbital meningiomas (OMs) comprise 0.4–2% of all meningiomas.[1] Primary OM arises from the optic nerve sheath arachnoid cap cells and constitutes less than 1/3rd of all OMs.[3] Secondary OM is a direct extension of intracranial meningiomas into the orbit and constitutes approximately 2/3rd of all OMs.[3] A third rare subset of OM, "ectopic orbital meningioma" arises from ectopic rests of arachnoid cells and is attached neither to the optic nerve sheath nor to the surrounding bone (sphenoid wing, anterior clinoid, tuberculum sellae, etc.).[4]

## CASE REPORT

We report the case of a 65-year-old female who presented with complaints of painless proptosis of the right eye since 1 year followed by gradual progressive visual loss and restricted right eye movements since 3 months [Figure 1]. There was no history of diplopia. The patient underwent

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a complete clinical, ocular, and radiological evaluation. Visual acuity of the right eye was found to be perception of light positive. Hertel's exophthalmometer revealed a 3 mm proptosis of the right eye [Figure 1]. The pupillary reaction was ill sustained. Fundoscopy revealed right eye optic atrophy.

Computed tomography and magnetic resonance imaging (MRI) of the orbits (plain plus contrast) revealed an intra-orbital, intra-conal, and heterogeneous contrast enhancing mass in the right eye [Figure 2]. The lesion was causing displacement of the globe anteriorly and slightly inferiorly, resulting in proptosis [Figure 2]. The right optic nerve was displaced inferiorly. The left eye and orbit did not reveal any abnormality. Intracranial structures were normal and there was no intracranial extension of the lesion.

Patient underwent total excision of the mass through supraorbital orbitotomy approach, using supra-brow incision. This approach allows an excellent exposure and optimal surgical operability in the orbit with minimal manipulation of the orbital structures, is not limited by tumor size, and has an excellent cosmetic result. Lesion was approached by a simple skin incision of 4 cm along the orbital rim. After detachment of the periosteum, the supraorbital nerve was dissected free and the periorbita was separated from the inner orbital roof. A single key burr hole was placed in the temporal fossa, at the frontosphenoidal suture just behind the zygomatic process of the frontal bone. An osteotomy of the middle part of the supraorbital rim was performed using a reciprocating saw and a small frontobasal trephination was carried out respecting the lateral border of the frontal sinus. The basal dura of the frontal lobe was pushed away and the orbital roof was removed. The tumor was identified after opening the periorbita [Figure 3].

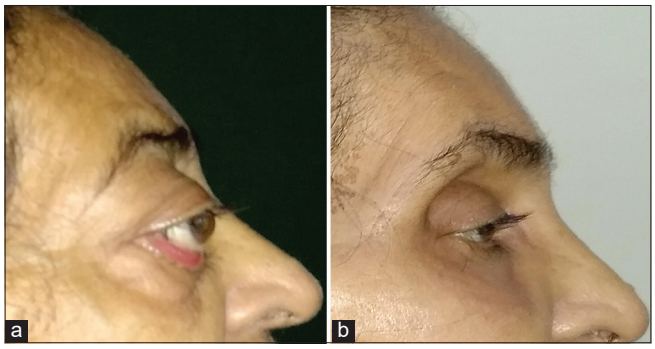


**Figure 1:** (a and b) Pre- and post-operative photographs of the patient showing clinical improvement in axial proptosis.

Intraoperatively, the lesion was firm in consistency, encapsulated, well-defined with distinct borders and was excised completely. The right optic nerve, though compressed and displaced inferiorly, was free from the lesion [Figure 3]. The mass was completely removed using standard microsurgical techniques and sent for histopathological evaluation. Mini plates were used on both sides of the orbital rim for orbital reconstruction.

Microscopic examination of hematoxylin and eosin slides showed a tumor which was comprised of meningothelial cells arranged in microcystic pattern with cob-web like background admixed with variable sized dilated vascular channels and focal lympho-plasmacytic cell aggregates. The meningothelial cells showed slender elongated nuclei with degenerative changes. The final histopathological diagnosis was "Meningioma-mixed microcystic angiomatous pattern, WHO Grade I" [Figure 4].

In the immediate postoperative period, patient's proptosis and vision improved (finger counting at 2 feet) [Figure 1]. At 3 months follow-up, patient's vision in the right eye was 6/12 and there was no clinical or radiological evidence of recurrence [Figure 5].

## DISCUSSION

Ectopic OM is a rare entity. The etiopathogenesis of ectopic OM is not fully understood. The possible hypothesis include

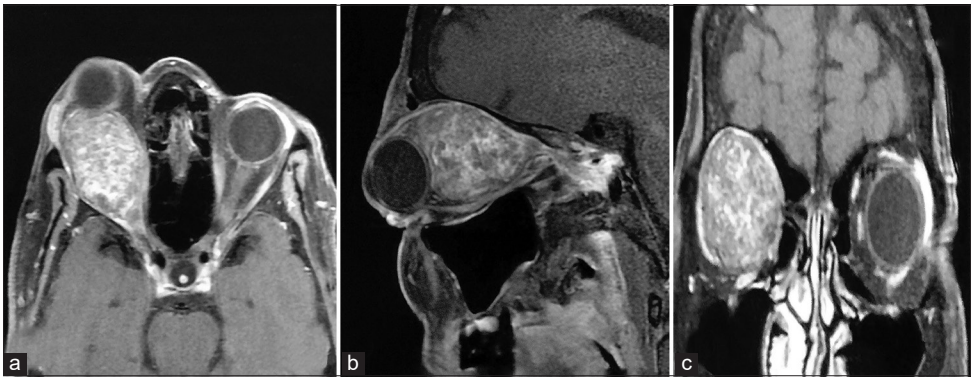


**Figure 2:** (a-c) Preoperative MRI images showing heterogeneous contrast enhancing intraorbital mass in the right eye with proptosis.

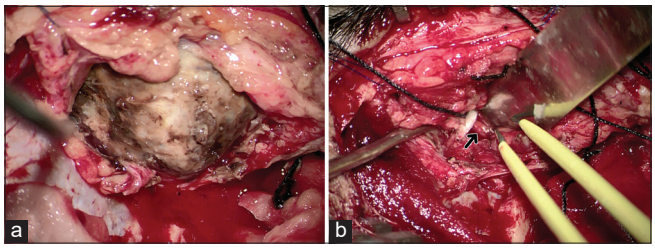

**Figure 3:** Intraoperative photographs (a) showing the tumor mass after opening the periorbita (b) The right optic nerve (arrow) which was compressed but free from the tumor.

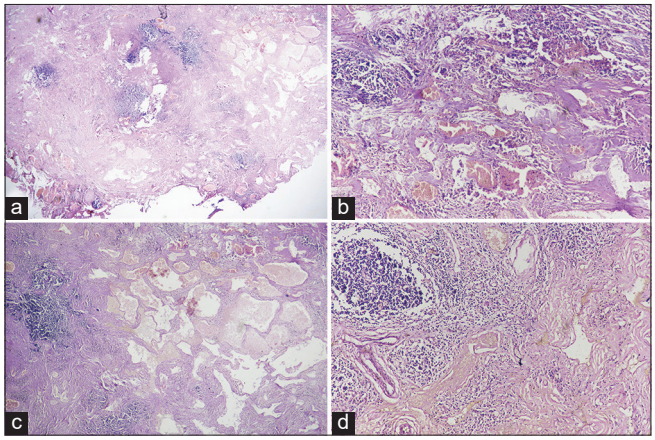

**Figure 4:** The histological images showing a) a tumor comprising of thick meningeal tissue with outlining multiple cystic spaces, a distinct area formed by conglomerate of vascular channels and focal lymphoid aggregates {hematoxylin & eosin (H and E) 20x}, b) junctional area highlighting both angiomatous and microcystic components (H and E 100x), c) Both thin and thick walled variably sized vessels in angiomatous component (H and E 40x), d) microcysts are lined by meningothelial cells with elongated slender nuclei (H and E 100x).

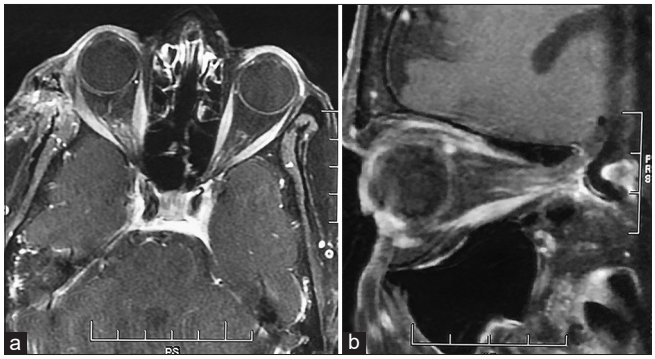

**Figure 5:** (a and b) Postoperative MRI images at 3 months follow-up showing complete excision of the mass with no radiological evidence of recurrence.

congenitally dislocated arachnoid nests within the periorbita or regressed orbital meningoceles which undergo tumor transformation or the presence of tumors originally in the optic nerve sheath that become detached and migrate to an ectopic location.[2,11] Ectopic OM may be associated with penetrating trauma to the orbit, which serves to dislodge the meningeal tissues within the orbit that later act as a nidus for the development of meningiomas.[4,8] In our case, history of orbital trauma was not present.

Ectopic OM occurs more commonly in the younger age groups and predominates in the medial orbit.[5] Ectopic OM differ from optic nerve sheath meningiomas in having late visual affection as compared to optic nerve sheath meningiomas in which visual loss is a predominant and early symptom.[7,9] On MRI, ectopic OM usually appears as a well-defined, T1 hypointense and T2 hyperintense lesion with heterogeneous post contrast enhancement with or without extraocular muscular adhesions.[5] Ectopic OM may be associated with asymmetric development of the paranasal sinuses.[12] The most common histologic type of ectopic OM is the meningotheliomatous variety.[6,7] On immunohistochemistry, these tumors are positive for epithelial membrane antigen and vimentin.[7]

Treatment of choice for ectopic OM is complete surgical excision wherever feasible. Recurrence is rare after complete excision. Residual/recurrent tumors after subtotal resection can be offered a second surgery or radiotherapy. Gündüz *et al.* in their report of two cases of ectopic OMs gave adjuvant radiotherapy in both the cases after subtotal tumor resection.[5] One, out of the two patients, developed proliferative radiation retinopathy and vision loss.[5] Pushker *et al.* in their paper reported recurrence in two out of the three cases of ectopic OMs which were treated with repeat surgical excision with no further recurrence.[10] On the contrary, the literature advocates radiotherapy as the 1st line of treatment for optic nerve sheath meningiomas.[13]

## CONCLUSION

Worldwide, only around 20 cases of ectopic OM have been reported.[7] Rarity of ectopic OM, total surgical excision with an excellent postoperative visual outcome prompted us to report this case.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

### Financial support and sponsorship

Nil.

### Conflicts of interest

There are no conflicts of interest.

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**How to cite this article:** Sharma AK, Gandhoke CS, Misra S, Ravi A, Gupta RK, Gupta SK. Ectopic intraconal orbital meningioma – A rare case report. Surg Neurol Int 2020;12:305.