

Optic Nerve Sheath Meningioma: A Case report

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Abstract

We report a case of Recurrent Optic nerve sheath meningioma of Transitional variant in a 42 years old male, who presented with Proptosis and loss of vision in right eye. At immunohistochemistry the meningotheliomatous and fibroblastic component showed vimentin positive. Optic nerve sheath meningiomas are very rare comprising of only 1 to 2% of all the meningiomas. This case is presented for its rarity of occurrence in unusual site, in male gender and its recurrent nature.

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I. Introduction

Optic Nerve Sheath Meningiomas (ONSMs) are very rare tumors of anterior visual pathway and constitute 1-2 % of all meningiomas and 2% of orbit. Mean age of presentation is 40 years in women and 36 years in men (younger age). These tumors show female preponderance in the ratio of 2:1. Primary ONSMs arise from the meningothelial capsules of arachnoid villi and seen along the entire course of optic nerve sheath. 10% are of primary orbital origin rest are secondary tumors extension from intracranial origin. Common histopathological types are either meningothelial or transitional pattern (WHO grade I). Recurrence is uncommon. Over all primary ONSM has an excellent prognosis for survival but visual prognosis is poor.

II. Case Presentation

A 42 years old male presented to ophthalmology OPD with chief complaints of proptosis and loss of vision in right eye for one year. Similar complaints in the same eye 3 years back. Underwent lateral orbitotomy and resection of tumor. Previous biopsy report was optic nerve meningioma. There was no family history of Neurofibroma 2. Ophthalmic examination shows Inferotemporal displacement of eyeball in right eye and facial asymmetry present. Fundoscopy was normal. General examination was normal. High Resolution Computed Tomography (CT) orbit and paranasal sinuses showed a mass of size 4x4x3.7cms in superior and superomedial quadrants of right orbit. MRI showed tubular mass in right orbit along the length of the nerve sheath (tram track appearance) without any intracranial extension. Exenteration of right orbit with flap reconstruction was done. The specimen was received in Department of pathology, Andhra Medical College, Visakhapatnam. On gross examination the mass was single, globular and grey tan measuring 6 x3x 3 cms. Cut section shows eye ball measuring 2x2x2 cms with attached capsulated grey tan lobulated tumor measuring 4x3x3 cms. On histopathology, diagnosis of transitional meningioma was given and was confirmed by positivity for Vimentin, Ki67 1%, EMA and S100 protein were negative on immunohistochemistry.

III. Discussion

Optic nerve sheath meningiomas (ONSM) are benign neoplasms of meninges surrounding the optic nerve which arises from the meningothelial cap cells of arachnoid villi and can develop from anywhere along the course of optic nerve. ONSM refers to both primary and secondary meningiomas of optic nerve.

Patients typically present in fourth decade and show a marked female predominance¹. Proptosis is common in these patients².

ONSM's are very rare tumors of anterior visual pathway and constitutes of only 1-2% of all the meningiomas and 2% of orbital tumors². Primary ONSM's account for one third of primary optic nerve tumors³. It was reported by Dutton that 92% of primary ONSM's arise from within intra orbital nerve sheath while only 8% are intracanalicular².

The usual presenting symptoms are loss of vision, optic atrophy and optociliary shunt vessels⁴. Here in this case, our patient was a 42 years old male who presented with loss of vision and proptosis which was recurrent.

Diagnosis of ONSM's is highly dependent on imaging studies. The axial CT scan images show a mass of size 4x4x3.7cms in superior and superomedial quadrants of right orbit. MRI is the investigation of choice in ONSM's. On MRI, a tubular mass is seen in the right orbit along the length of the nerve sheath (tram track appearance) without any intracranial extension. Tram tracking with meninges hyper intense /hyperdense on either side is a very characteristic sign which is seen only in 24% of the tumors⁵ which is seen in this case.

On histopathology, ONSM's usually present in either meningotheial pattern or transitional pattern⁶. In meningotheial pattern polygonal cells are arranged in sheets separated by vascular trabeculae. In transitional pattern spindle or oval cells are typically arranged in whorling pattern with central hyalinization. In the present case, the tumor was capsulated and was composed of spindle cells arranged in short fascicles and whorling pattern. There were syncytial sheets of epithelial cells with indistinct cell borders, eosinophilic cytoplasm, round to spindle uniform nuclei. There was no nuclear atypia, necrosis or mitotic activity. On IHC it showed vimentin positivity and Ki67 1%, EMA and S100 protein were negative.

Similar to intra cranial meningiomas, according to WHO three histological grades were identified, Grade 1 is benign, grade 2 is atypical, grade 3 is malignant. However, recurrence rates may vary. They are reported as 6.9%⁷, 34.6%⁸, 72.7%⁹ according to various studies.

Stereotactic fractionated radiotherapy is current best available modality of treatment which delivers sufficient dose of radiation to the tumor, sparing surrounding tissue⁴. Severe proptosis often warrants enucleation or exenteration. Surgical treatments are often associated with worse visual outcomes. Due to technical difficulties and increased rates of postoperative blindness surgery have been replaced by radiation therapy. Radiation therapy is now emerging vision preserving therapy in ONSM.

The benign nature and rarity of the tumor is consistent with the previous reports. In contrary to previous reports the nuance in this case is its presentation in male gender, with recurrence and showing characteristic tram track appearance on CT.

IV. Conclusion

ONSM's often have an indolent course and impose a management difficulty to the clinician most of the times. Diagnosis of ONSM largely rely upon clinical picture and appropriate neuro imaging. As surgical intervention carries significant mortality and morbidity, current treatment protocols recommend stereotactic fractionated radiotherapy to patients.

Prior detection of ONSM with improved neuro imaging technology and histopathological examination with IHC, and more accurate radiotherapy delivery along with surgical management will improve the prognosis in these patients.

LEGENDS

Fig 1: Clinical picture showing proptosis of right eye

Fig 2: RADIOLOGICAL IMAGE, CT showing a mass of size 4x4x3.7cms in superior and superomedial quadrants of right orbit.

Fig 3: MRI showing tubular mass in right orbit along the length of the nerve sheath (tram track appearance) without any intracranial extension

Fig 4:GROSS FINDINGS

Received a single globular grey tan mass measuring 6x3x3 cms. Cut section shows eye ball measuring 2x2x2 cms with attached capsulated grey tan lobulated tumor measuring 4x3x3 cms

Fig 5: MICROSCOPIC FINDINGS

LOW POWER VIEW: showing retinal epithelium and capsulated tumor composed of spindle shaped cells arranged in fascicles and whorling pattern (H&E Stain 100X)

Fig 6 :HIGH POWER VIEW : Syncytial sheets of epithelial cells with indistinct cell borders, eosinophilic cytoplasm, round to spindle uniform nuclei, no nuclear atypia, necrosis and mitotic activity (H&E, 400X)

Fig 7: IHC showing vimentin positive

Fig 8: Ki67 index

FIGURES

FIG 1



FIG 2



FIG 3

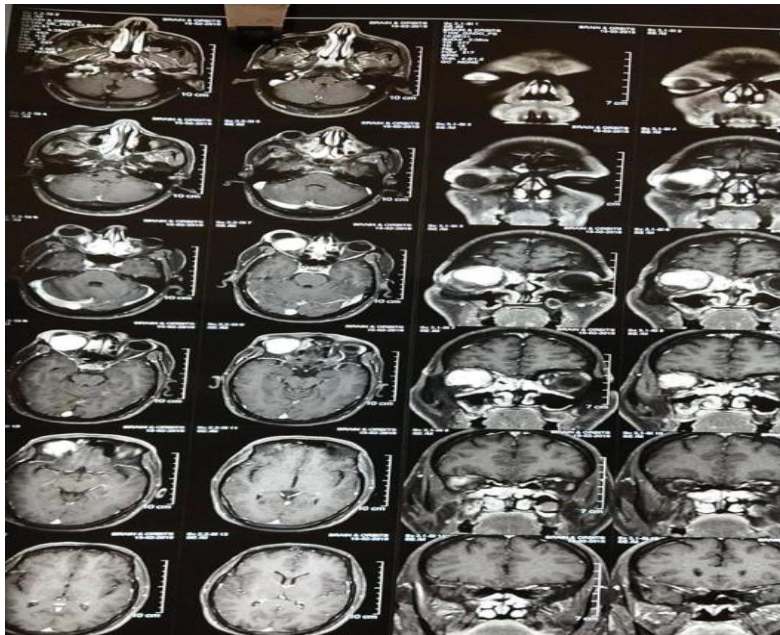


FIG 4

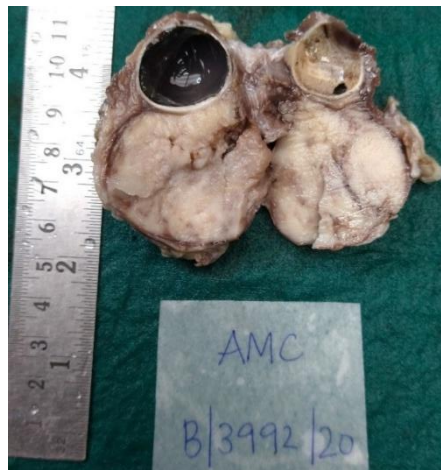


FIG 5.1

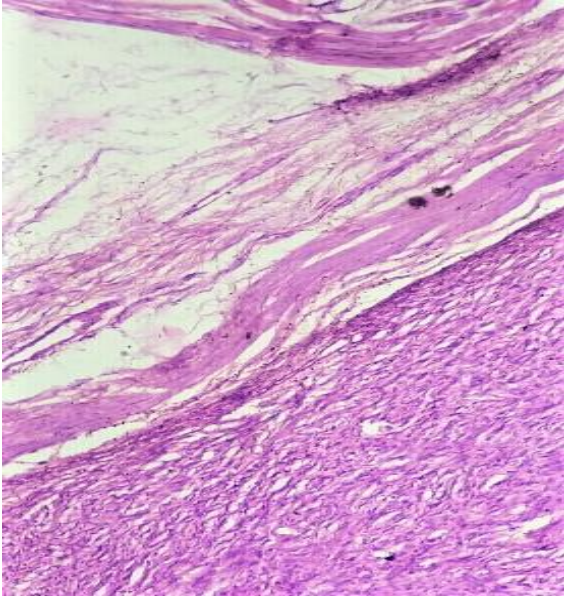


FIG 5.2

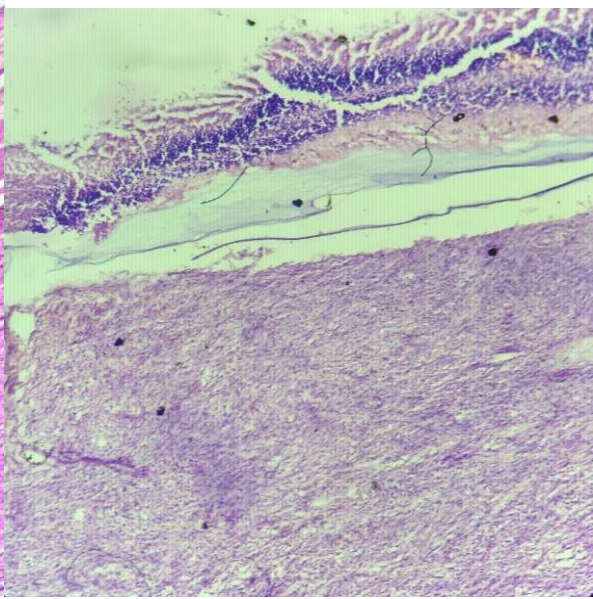


FIG 6

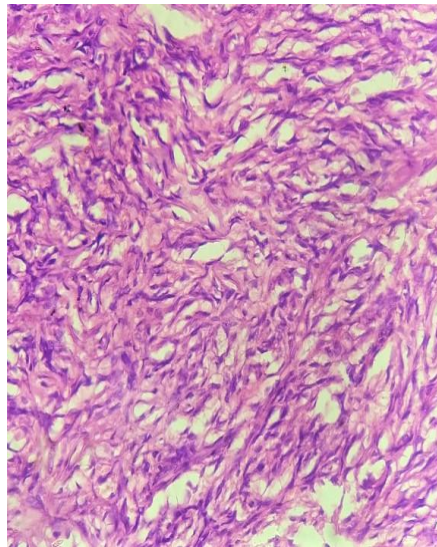


FIG 7

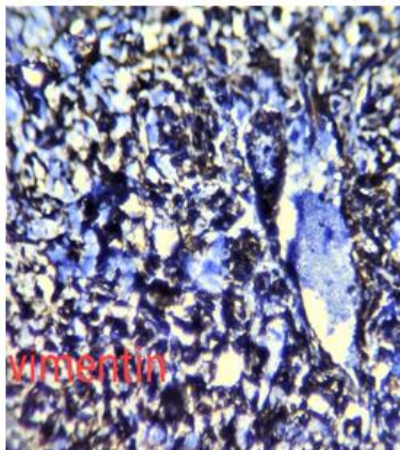
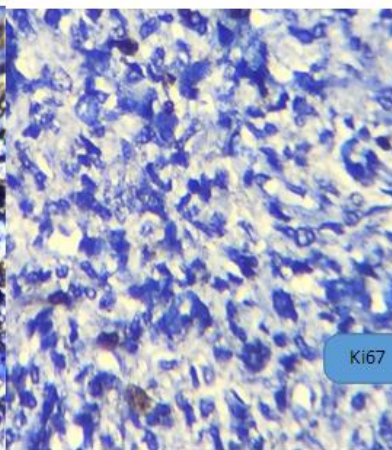


FIG 8



References

- [1]. Richard T Parker, Christopher A ovens. Optic nerve sheath meningiomas; Prevalence, impact and management strategies. *Eye Brain*. 2018;10:85-99.
- [2]. Dutton JJ. Optic nerve sheath meningiomas. *Survophthalmol*. 1992;37(3):167-183.
- [3]. Savas P Deftereos, Georgios K Karagiannakis. Optic nerve sheath meningioma : a case report . *Cases journal* 2008,1:423.
- [4]. Frisen L, Royt WF, Tengroth BM. Optociliary veins, disc pallor and visual loss. A triad of signs indicating sphenoidal meningioma. *Acta Ophthalmol*. 1973;51(2):241-249. 5.
- [5]. Saeed P, Rootman J, Nugent RA, White VA, Mackenzie IR, Koornneef L. Optic nerve sheath meningiomas . *Ophthalmology*. 2003;110(10):2019-2030.
- [6]. Eddleman CS, Liu JK. Optic nerve sheath meningioma : Current diagnosis and treatment . *NeurosurgFocus* . 2007;23(5):E4
- [7]. Turbin RE, Pokorny K. Diagnosis and treatment of orbital optic nerve sheath meningioma. *Cancer Control*. 2004;11(5):334-341.
- [8]. Carrasco JR, Penne RB. Optic nerve sheath meningiomas and advanced treatment options: *Opin Ophthalmol*. 2004;15(5):406-410.
- [9]. Berman D, Miller NR. New concepts in the management of Optic nerve sheath meningiomas. *Ann Acad Med Singapore*. 2006;35(3):168-174.

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