

INTRAORBITAL ABDUCENS NERVE SCHWANNOMA- A CASE REPORT

Dayanand Kumar R¹, Ramya Devaraj²

¹Professor and HOD, Department of Radiodiagnosis and Imaging, MVJ Medical College and Research Hospital, Hoskote, Bangalore.

²Postgraduate Student, Department of Radiodiagnosis and Imaging, MVJ Medical College and Research Hospital, Hoskote, Bangalore.

HOW TO CITE THIS ARTICLE: Kumar DR, Devaraj R. Intraorbital abducens nerve schwannoma- A case report. J. Evid. Based Med. Healthc. 2017; 4(74), 4401-4403. DOI: 10.18410/jebmh/2017/876

PRESENTATION OF CASE

We are reporting a case of left intraorbital abducens nerve schwannoma in a 42-year-old male patient who presented with discomfort and proptosis of the left eye of 5 months duration. He underwent imaging in the Radiology Department of MVJ Medical College and Research Hospital, Hoskote, Bangalore, and was found to have a mass lesion in the intraconal compartment of the left orbit, which involved the abducens nerve. The tumour was excised by lateral orbitotomy and histopathological examination of the operated specimen was suggestive of schwannoma. Intraorbital schwannoma is a rare tumour and accounts for about 1-2% of all neoplasms of the orbit. Orbital schwannomas most common origin is from the sensory branches of the trigeminal nerve. But, intraorbital abducens nerve schwannomas are extremely rare.

Schwannomas are tumours arising from Schwann cells of the neural sheaths of motor and sensory nerves. They arise focally from the sheath of the fascicle and present as well-defined, eccentrically-placed masses.¹ Three clinical forms may present- localised schwannoma in association with neurofibromatosis (as part of von Recklinghausen syndrome) or as schwannomatosis.² Schwannoma is seen predominantly in cranial nerve VIII. Intraorbital schwannoma accounts for about 1-2% of all neoplasms of the orbit and intraconal abducens nerve schwannoma are extremely rare.³ There are less than 10 reported cases in literature. We report a case of intraorbital schwannoma arising from the terminal branch of the abducens nerve.

History and Physical Examination

A 42-year-old male patient came with history of discomfort and proptosis of the left eye. Eye examination was done and he was found to have proptosis of the left eye. The distance measuring 34 mm from the interzygomatic line to the anterior surface of the left globe. His visual acuity was normal and was 6/6 as per Snellen chart and visual fields

were symmetrical in both eyes on clinical examination. Fundoscopy was normal.



Figure 1(a)



Figure 1(b)

Figure 1(a) and (b). Proptosis of the Left Eye of the Patient was Noted

Radiological Findings

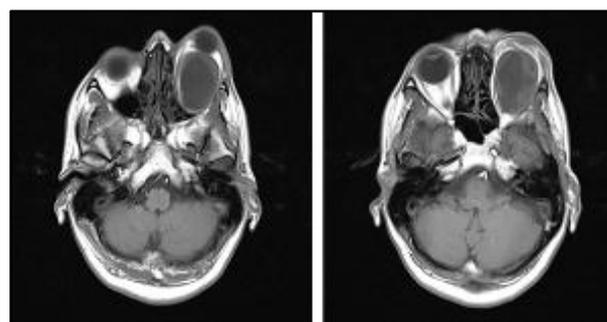


Figure 2(a)

Figure 2(b)

Figure 2(a) and (b). T1W MRI Axial Sequence Shows the Solid Component of the Lesion Appearing Isointense and the Cystic/ Necrotic Component Appearing Hypointense

Financial or Other, Competing Interest: None.

Submission 28-08-2017, Peer Review 04-09-2017,

Acceptance 12-09-2017, Published 14-09-2017.

Corresponding Author:

Dr. Ramya Devaraj,

No. 1648, Jeevan Bhima Nagar,

Main Road, HAL 3rd Stage,

Bangalore-560008.

E-mail: ramya.d685@gmail.com

DOI: 10.18410/jebmh/2017/876



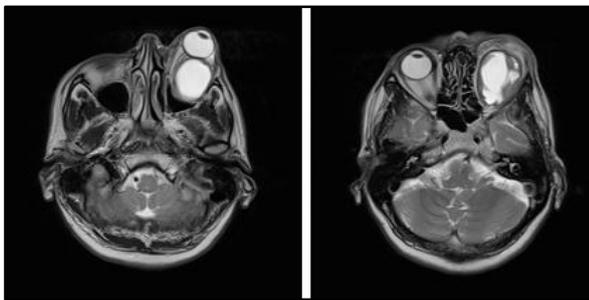


Figure 3(a) **Figure 3(b)**

Figure 3(a) and (b). T2W MRI Axial Sequence Shows the Solid Component of the Lesion Appearing Isointense and the Cystic/Necrotic Component Appearing Hyperintense

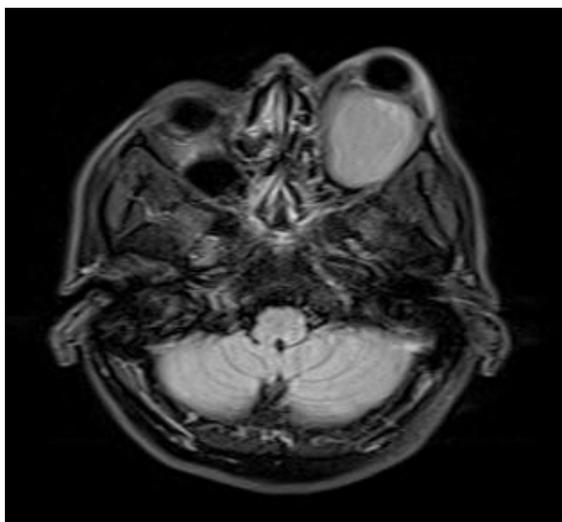


Figure 4. Flair Sequence Shows the Lesion-Appearing Heterogeneously Hyperintense

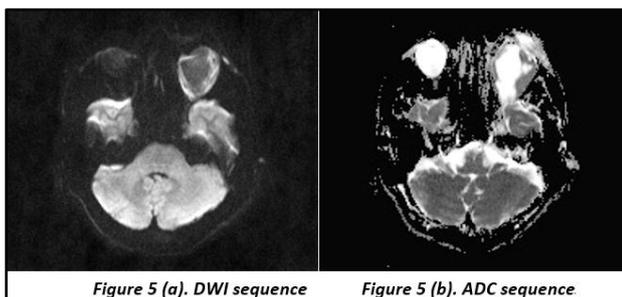


Figure 5 (a). DWI sequence **Figure 5 (b). ADC sequence**

Figure 5(a) and (b). The Lesion Shows No Diffusion Restriction on DWI Sequences

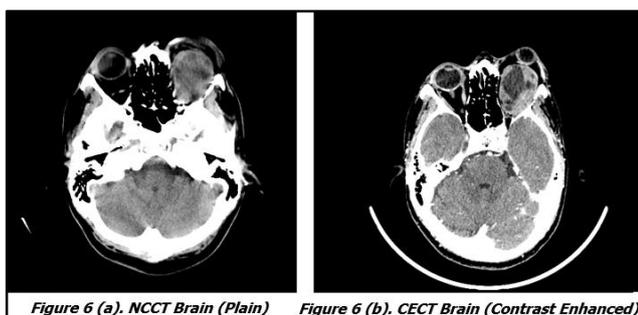


Figure 6 (a). NCCT Brain (Plain) **Figure 6 (b). CECT Brain (Contrast Enhanced)**

Figure 6(a) and (b). The Lesion Appears Isodense on NCCT Scan Brain and the Solid Component of the Lesion Shows Enhancement on Contrast-Enhanced CT Scan of Brain with Necrotic/Cystic Component-Appearing Hypodense

There is a well-defined, oblong shaped, heterogeneous solid cystic (necrotic) mass lesion in the intraconal compartment of the left orbit. The lesion measures 4.5 x 2.9 x 4 cm (TR x AP x CC) in size approximately. Medially, the lesion is causing bowing of lamina papyracea and displacement and stretching of the optic nerve medially and inferiorly. Fat plane between the lesion and the medial rectus muscle is maintained. Laterally, the lesion is abutting the lateral rectus muscle with loss of intervening fat plane. The origin of the tumour is from the terminal branch of cranial nerve VI.

DIFFERENTIAL DIAGNOSIS

- a. Orbital meningioma.
- b. Orbital neurofibroma.
- c. Optic nerve glioma.

Clinicoradiological Diagnosis

A well-defined, oblong shaped, heterogeneous solid cystic (necrotic) mass lesion noted in the intraconal compartment of the left orbit suggestive of a neoplasm arising from the VI cranial nerve (abducens nerve).

PATHOLOGICAL DISCUSSION

Histopathological examination revealed typical findings of schwannoma. Antoni A and Antoni B were identified. The tumour cells showed diffuse nuclear positivity for S100 protein.

DISCUSSION OF MANAGEMENT

The patient underwent lateral orbitotomy with complete removal of the tumour.

Postoperative Picture-



Figure 7. Postoperatively, the Patient had No Complaints of Diplopia and No Paresis of Cranial Nerve VI was Detected

DISCUSSION

Schwannomas are well-defined, encapsulated, slowly progressive benign tumours that develop as eccentric growths from the sheaths of peripheral nerves. They have a predilection for the head and neck region. In the orbit, they account for 1-2% of all tumours.^{3,4} Schwannomas of the abducens nerve are extremely rare. The tumour maybe located within the cavernous sinus or more often at the preoptine region.^{3,5}

Based on the work of Erdogmus et al, Irace et al proposed that the entry point of nerve fibres in the muscle is the zone where intraorbital schwannomas truly arise.^{6,7} The abducens nerve runs on the medial surface of the lateral

rectus muscle and innervates the middle third of the muscle with an average of three small branches entering the muscle.^{8,9}

The schwannomas arising from the sensory branches of trigeminal nerve are located in superior and medial quadrants of orbit. The schwannomas arising from the motor nerves usually arise at their respective myoneural junction. Those arising from the infraorbital nerve are in inferior quadrant.^{1,10}

They generally appear in patients between 20 to 70 years of age.¹¹ When small, they are mostly asymptomatic. They gradually grow in size and produce progressive painless ocular proptosis.^{12,13} They may have headache. The vision is usually not impaired unless the tumour is situated at orbital apex and compresses on the optic nerve.^{2,4}

Orbital schwannoma on MRI is usually hypointense on T1W images and hyperintense on T2W images. It can enhance homogeneously or heterogeneously. The intraorbital abducens schwannoma is situated in lateral intraorbital quadrant and medial to lateral rectus muscle. It displaces the optic nerve medially and upwards.^{14,15} CT scan is also helpful to determine the tumour extent and whether it has spread to the periorbital tissues or paranasal sinuses.¹⁶ The schwannoma can be approached by lateral or superior orbitotomy.^{8,13} Superior orbitotomy provides direct access to superior, medial and lateral quadrants of orbit to ensure safe and complete excision of the lesions. The nerve attachment at the myoneural junction has to be visualised to preserve the integrity of the nerve. But, complete excision is necessary to prevent recurrence. Gamma knife surgery has also been tried in patients of intraorbital schwannoma.^{17,18}

FINAL DIAGNOSIS

The lesion located in the intraconal compartment of the left orbit with characteristic imaging findings and loss of intervening fat plane between the lesion and lateral rectus muscle noted arising from the terminal branch of the VI cranial nerve and histopathological findings suggest intraorbital schwannoma of the abducens nerve.

CONCLUSION

Intraorbital abducens nerve schwannoma is an extremely rare clinical entity. It usually presents as gradually progressive painless proptosis. Complete excision of the tumour should be the goal as residual tumour can potentially regrow.

ACKNOWLEDGEMENT

We thank our patient who gave consent for photographing and submitting the work for publication.

REFERENCES

- [1] Lida Y, Sakata K, Kobayashi N, et al. Orbital abducens nerve schwannoma: a case report and review of the literature. *NMC Case Rep J* 2016;3(4):107-109.
- [2] Kim KS, Jung JW, Yoon KC, et al. Schwannoma of the orbit. *Arch Craniofac Surg* 2015;16(2):67-72.
- [3] Bhaganagare AS, Bidkar VC, Rodrigus E, et al. Orbital intraconal abducens nerve schwannoma: a case report and review of literature. *Asian J Neurosurg* 2015;10(1):61.
- [4] Erlich SA, Tymianski M, Kiehl TR. Cellular schwannoma of the abducens nerve: case report and review of the literature. *Clin Neurol Neurosurg* 2009;111(5):467-471.
- [5] Jakobiec FA, Font RL. Peripheral nerve sheath tumours. In: Spencer WH, ed. *Ophthalmic pathology*. Philadelphia, PA: Saunders 1986:2616-2626.
- [6] Erdogmus S, Govsa F, Celik S. Innervation features of the extraocular muscles. *J Craniofac Surg* 2007;18(6):1439-1446.
- [7] Irace C, Davi G, Corona C, et al. Isolated intraorbital schwannoma arising from the abducens nerve. *Acta Neurochir (Wien)* 2008;150(11):1209-1210.
- [8] Rato RM, Correia M, Cunha JP, et al. Intraorbital abducens nerve schwannoma. *World Neurosurg* 2012;78(3-4):375.e1-4.
- [9] Mascarenhas L, Magalhaes Z, Honavar M, et al. Schwannoma of the abducens nerve in the cavernous sinus. *Acta Neurochir (Wien)* 2004;146(4):389-392.
- [10] Cantore G, Ciappetta P, Raco A, et al. Orbital schwannomas: report of nine cases and review of the literature. *Neurosurgery* 1986;19(4):583-588.
- [11] Garg R, Dhawan A, Gupta N, et al. A rare case of benign isolated schwannoma in the inferior orbit. *Indian J Ophthalmol* 2008;56(6):514-515.
- [12] Vachata P, Sames M. Abducens nerve schwannoma mimicking intrinsic brainstem tumor. *Acta Neurochir (Wien)* 2009;151(10):1281-1287.
- [13] Feichtinger M, Reinbacher KE, Pau M, et al. Intraorbital schwannoma of the abducens nerve: case report. *J Oral Maxillofac Surg* 2013;71(2):443-445.
- [14] Malone JP, Lee WJ, Levin RJ. Clinical characteristics and treatment outcome for nonvestibular schwannomas of the head and neck. *Am J Otolaryngol* 2005;26(2):108-112.
- [15] Wang Y, Xiao LH. Orbital schwannomas: findings from magnetic resonance imaging in 62 cases. *Eye (Lond)* 2008;22(8):1034-1039.
- [16] Rootman J, Goldberg C, Robertson W. Primary orbital schwannomas. *Br J Ophthalmol* 1982;66:194-204.
- [17] Kakizaki H, Zako M, Miyagawa T, et al. Myoneural junctions of extraocular muscles: distances from the orbital rim and widths. Part 2. *Ophthalmologica* 2006;220(2):87-93.
- [18] Hayashi M, Chernov M, Tamura N, et al. Gamma knife surgery for abducens nerve schwannoma. Report of 4 cases. *J Neurosurg* 2010;113 Suppl:136-143.