International Journal of Advances in Medicine
Saini V et al. Int J Adv Med. 2022 Mar;9(3):353-355
http://www.ijmedicine.com
pISSN 2349-3925 | eISSN 2349-3933
DOI: https://dx.doi.org/10.18203/2349-3933.ijam20220441

Case Report

Orbital varix presenting with enophthalmos and intermittent proptosis: a case report

Varun Saini, Shruti Bhattacharya*, Usha K. Raina, Jawahar Lal Goyal

Department of Ophthalmology, Guru Nanak Eye Centre, Maulana Azad Medical College, New Delhi, India

Received: 03 November 2021
Accepted: 29 November 2021

***Correspondence:**
Dr. Shruti Bhattacharya,
E-mail: shrutibhattacharya1993@gmail.com

ABSTRACT

We report the case of a 22-year-old male, presenting with right enophthalmos and intermittent exophthalmos observed after valsalva maneuver. Orbital imaging revealed a retrobulbar vascular lesion with multiple phleboliths suggestive of orbital varices. On magnetic resonance angiography, no obvious feeder vessel could be delineated. We decided to manage the patient conservatively, in the absence of complications such as proptosis with corneal exposure, optic nerve compression, orbital hemorrhage. Orbital varix is a low flow vascular anomaly which generally presents with an intermittent exophthalmos. Development of enophthalmos associated with orbital varices as in our case, is very rare.

Keywords: Orbital varices, Enophthalmos, Proptosis intermittent

INTRODUCTION

Orbital varices are vascular hamartomas consisting of a plexus of low-pressure, low-flow, thin-walled vessels that communicate with normal orbital veins.[1] Orbital varices are characterized by intermittent and non-pulsatile proptosis, which can may be precipitated or accentuated by increasing venous pressure through coughing, straining, the valsalva maneuver, assuming a head-down position or external compression of the jugular veins.[2] However, rarely, orbital varices can lead to enophthalmos by causing atrophy of orbital fat.[3] We could find reports of only three such cases in the literature.[4-6] We propose to report a case of orbital varices with enophthalmos and intermittent proptosis in a 22 year old male patient.

CASE REPORT

A man aged 22 years presented with complaints of sunken appearance of right eye since childhood, associated with outward bulging of eyeball appearing on straining and bending head down. Proptosis was not associated with any pain nor diplopia. There was no relevant personal or family history.

This patient had a right 4mm enophthalmos measured with Hertel's exophthalmometer (12 mm right eye, 16 mm left) and right intermittent, non-pulsating proptosis observed during Valsalva manoeuver or bending forward (18 mm to the right eye, 16 mm to the left) (Figure 1 A-D). The vertical palepebral aperture was 13mm in right eye and 11mm in left eye. Uncorrected visual acuity of both eyes was 20/20. Examination of the anterior segment, pupils, fundus and ocular motility were normal. The intraocular pressure measured with applanation tonometry was 14 mmHg at each eye. In addition, a bluish vascular mass was seen in the soft palate on the right side going into the right tonsillar fossa and involving the uvula (Figure 2 A).

Contrast enhanced computed tomography (CECT) through orbit revealed an ill-marginated soft tissue lesion in right retrobulbar space involving both intraconal and extraconal compartment and showing increase in size on prone scan, likely orbital varix (Figure 2 B). CECT through neck revealed an ill-marginated infiltrating enhancing lesion in right tonsillar fossa, showing multiple phleboliths and dilated tortuous vascular channels, extending to right masseteric and parotid space as well as to right

infratemporal fossa, likely slow flow veinous vascular malformation.

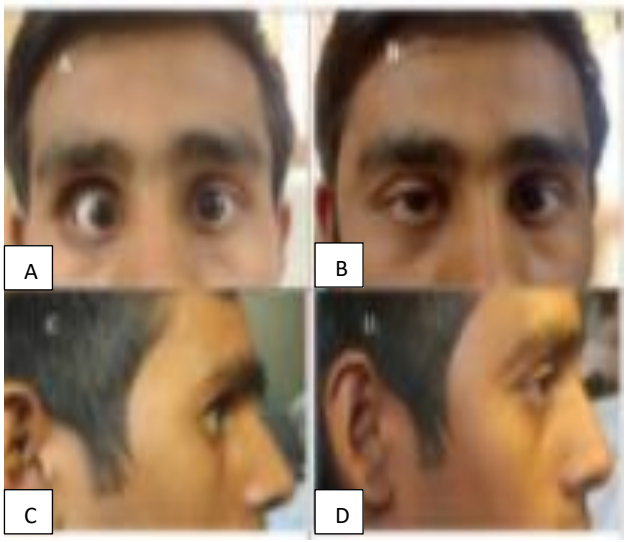

Figure 1 (A-D): Right eye enophthalmos- (front view) and (side view); right eye transient proptosis following valsalva maneuver- (front view) and (side view).

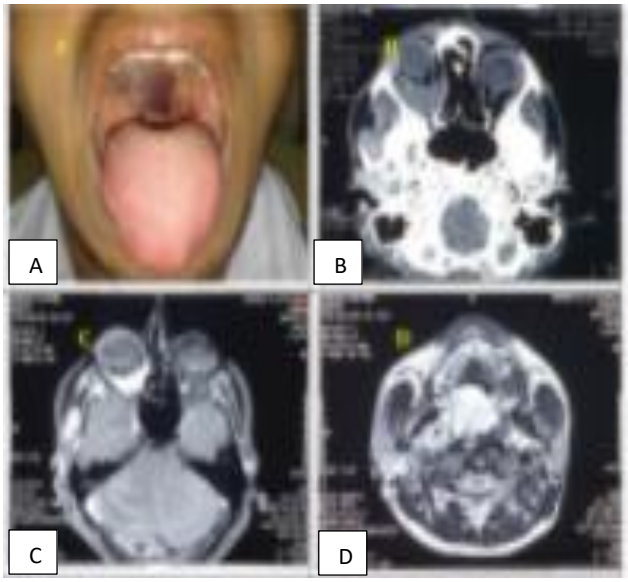

Figure 2 (A-D): Bluish vascular mass in the soft palate going into the right tonsillar fossa and uvula; CECT scan through orbit showing ill-marginated soft tissue lesion in the right orbit; gadolinium enhanced MRI showing enhancing soft tissue lesion in the right orbit; gadolinium enhanced MRI of enhancing soft tissue mass lesion in the right massetric space and infratemporal fossa.

Gadolinium enhanced MRI revealed multilobulated enhancing soft tissue mass lesion showing multiple phleboliths in the right massetric space and right infratemporal fossa with extensions into the adjacent neck spaces and oral cavity (Figure 2 D). Extension was also seen into the right orbit and extraconal space, through the inferior orbital fissure (Figure 2 C). On MRI angiography, the branches of right external carotid artery were seen in close proximity to the lesion, however no obvious feeder vessel could be delineated.

DISCUSSION

Enophthalmos is a relative posterior displacement of a normal-sized globe in relation to the bony orbital margin. It is due to a change in the volumetric relationship between the rigid bony orbital cavity and its contents. It can occur either due to expansion of the orbital cavity with no change in the volume of the orbital contents, or due to decrease in volume of the orbital soft tissue. Enophthalmos due to orbital cavity expansion occurs in fractures of the orbital bones and orbital varices with bone erosion. Enophthalmos due to loss of orbital contents include orbital fat atrophy following severe inflammation or infection, trauma, orbital varices, external beam irradiation, orbital metastases, and surgical resection of an orbital mass.[7] Enophthalmos can be diagnosed by Hertel's exophthalmometric value below 14 mm.[3] Bilateral comparative inspection is more important and a difference of 2 mm or more between two eyes, on Hertel exophthalmometry is suggestive of enophthalmos.

Orbital varices are characterized by intermittent proptosis which occurring during coughing, straining, the valsalva maneuver, assuming a head-down position or external compression of the jugular veins. These maneuvers raise the veinous pressure causing the variceal vessels to fill up and distend. This pushes the globe forward, causing proptosis. The varix may eventually cause atrophy of orbital fat, which allows the globe to sink back into the orbit when the vessels are not distended.[4] Enophthalmia in orbital varices may be due to orbital cavity expansion by bone erosion or orbital fat atrophy by pressure effect.[4,7] In this patient no defect in the bone walls was seen on imaging, so the cause of enophthalmia was the atrophy of the orbital fat.

Orbital varices can be treated by surgical excision, embolization under radiological supervision or carbon dioxide laser surgery.[8,9] The indications of treatment include orbital hemorrhage, optic nerve compression, severe proptosis causing corneal exposure or unsightly appearance. Orbital varices require careful monitoring because they can be progressively enlarged, thrombosed or transformed with intravascular endothelial hyperplasia.[2,10] Since our patient had normal vision and no complications, we did not plan any intervention. But we decided to keep under close observation the orbital varix as well as the neck mass.

CONCLUSION

Orbital varices are characterized by intermittent proptosis, but rarely they can cause enophthalmos, as in our patient. Surgical treatments or embolization are indicated for severe complications such as diplopia, optic nerve

compression, hemorrhage, proptosis with corneal exposure or significant aesthetic damage. In the absence of such complications, careful monitoring should be done because of the risk of aggravation and transformation.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Wright JE, Sullivan TJ, Garner A, Wulc AE, Moseley IF. Orbital venous anomalies. Ophthalmology. 1997;104:905-13.
2. Kanski JJ. Orbit. Vascular malformations. In: Kanski JJ, editor. Clinical ophthalmology, a systematic approach, 5th ed. Edinburgh: Butterworth Heinemenn, Elsevier Ltd. 2003;557-89.
3. Athanasiov PA, Prabhakaran VC, Selva D. Non-traumatic enophthalmos: A review. Acta Ophthalmol. 2008;86:356-64.
4. Haritoglou C, Hintschich C. Progressive enophthalmos in association with an orbital varix. Klin Monatsbl Augenheilkd. 2003;220(4):268-71.
5. Chung CF, Lai JS. Enophthalmos caused by an orbital venous malformation. Hong Kong Med J. 2009;15(4):299-300.
6. Aydin A, Velioglu M, Ersanli D. Orbital varix presenting with enophthalmos. A case report. J Fr Ophtalmol. 2010;33:344.
7. Cline RA, Rootman J. Enophthalmos: a clinical review. Ophthalmology. 1984;91:229-37.
8. Kremer I, Nissenkorn I, Feuerman P, Ben-Sira I. Congenital orbital vascular malformation complicated by massive retrobulbar hemorrhage. J Pediatr Ophthalmol Strabismus. 1987;24:190-3.
9. Takeshi A, Uozumi T, Kiya K, Yano T, Sumida M, Yoshikawa S et al. Embolisation of orbital varix. Neuroradiology. 1994;36:487-9.
10. Shields JA, Shields CL, Eagle Jr RC, Diniz W. Intravascular papillary endothelial hyperplasia with presumed bilateral orbital varices. Arch Ophthalmol. 1999;117:1247-9.

Cite this article as: Saini V, Bhattacharya S, Raina UK, Goyal JL. Orbital varix presenting with enophthalmos and intermittent proptosis: a case report. Int J Adv Med 2022;9:353-5.