A Case Report of Orbital Venolymphatic Malformation - A diagnostic dilemma

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ABSTRACT

Introduction: Intermittent proptosis is a transitory position-dependent protrusion of one or both eyeballs beyond orbit. The common causes of proptosis in young females are tumors, inflammation, trauma , thyroid eye disease and vascular lesions. The case report highlights the importance of imaging in prone position.

Case: We report a rare case of a 24-year-old female who recently developed intermittent proptosis while bending forwards. The patient was evaluated clinically. Ultrasonography B scan, Computerised axial tomography (CT), Magnetic resonance imaging (MRI) were advised to rule out orbital varices, arteriovenous aneurysm, or any organic lesion. Probable diagnosis of venolymphatic malformation was made.

Conclusion: An intertwined approach was carried out with Ophthalmologist and Radiologist to arrive at the final diagnosis highlighting the importance of imaging in prone position.

Keywords: Intermittent proptosis, Orbital varices, Venolymphatic malformation.

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INTRODUCTION

The pattern of intermittent proptosis classically consists of enophthalmos with the head in the upright position, readily reducible orbital contents, pronounced exophthalmos developing rapidly due to venous stasis, which is induced by lowering the head, performing Valsalva manoeuvres, straining and coughing (Brauston et al.,1963) . At rest with the head elevated, the affected eye assumes a normal position, or is enophthalmic.

CASE REPORT

A 24-year-old female visited the ophthalmology outpatient department with complaints of a dragging sensation and bulging of the right eyeball while bending forward. The patient noticed a protruding eyeball for the first time two years back while performing daily chores. She gradually began to experience pain in her right eye, which spread to the ipsilateral forehead, but the pain was mild, worsened by stooping, coughing and relieved by standing. She also gave a history of diminution of vision concurrent with proptosis, which improved spontaneously as the protrusion regressed. There was no history of nausea, vomiting, fever, seizures, ocular trauma and surgery. In the left eye, there were no such complaints. The patient underwent a detailed ocular examination. The pupil was round, regular and reactive in both eyes. There was no restriction of movements in any gaze. Visual acuity in each eye was 20/20 on Snellen's chart. Examination of the left eye was within normal range. In the upright position, the right eye was in enophthalmos. Exophthalmometry showed 2.8 mm of enophthalmos and 5.9 mm of exophthalmos when bending forward, using Hertel's exophthalmometer. The proptosed eye had no pulsation, and there was no audible bruit. No perilimbal conjunctival abnormality was found. Slit lamp biomicroscopy revealed a normal iris pattern and anterior chamber depth. On direct and indirect ophthalmoscopy, the fundus was normal. Complete blood count (CBC), thyroid function, auto-antibody tests, renal function, and C-reactive protein were all part of the comprehensive diagnostic workup. Her USG B-scan in the supine position showed normal bilateral globes and no obvious venous malformation. The patient was referred to the radiology department for a Contrast-enhanced MRI [CE-MRI] and CT scan. CE-MRI was performed supine, and lobulated hyperintense lesions were seen. The radiologist recommended a contrast enhanced CT scan of the orbit [venous phase] in a prone position to correlate the clinical findings. CECT showed a lesion of approximately 28×9×15 mm in the superior aspect of the right extraconal space extending into the intraconal space posteriorly (Fig 1); innumerable slender vascular channels within the bulky right superior rectus muscle belly were seen. (Fig2,3). These CECT findings are suggestive of venolymphatic malformation, but the patient refused to undergo any intervention.

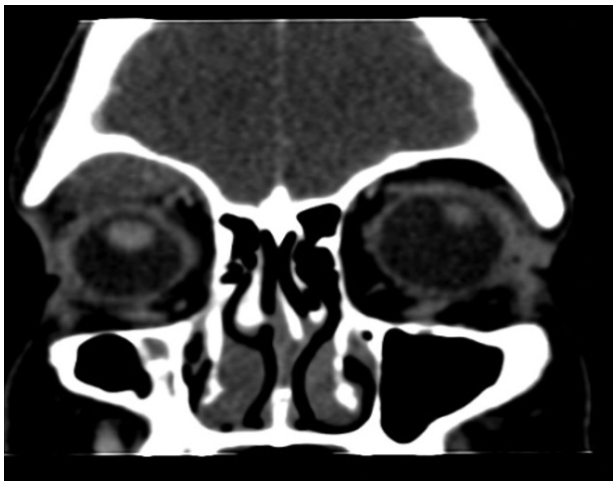

Fig 1: Contrast enhanced CECT (prone position-reformatted images)- Coronal view reveals a lobulated enhancing lesion in the anterosuperior aspect of the right orbit in the extraconal space. Note -inferiorly displaced right globe secondary to the mass effect.

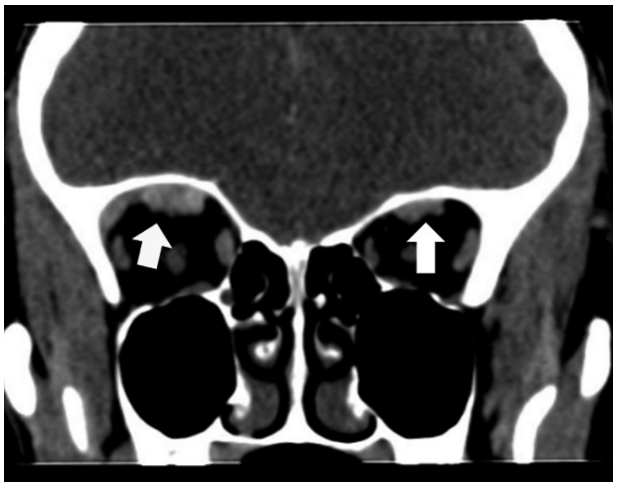

Fig 2: Bulky right superior rectus muscle (yellow arrow) as compared to the left side (white arrow). Multiple tiny enhancing leashes of vessels inseparable from the right superior rectus muscle are seen.

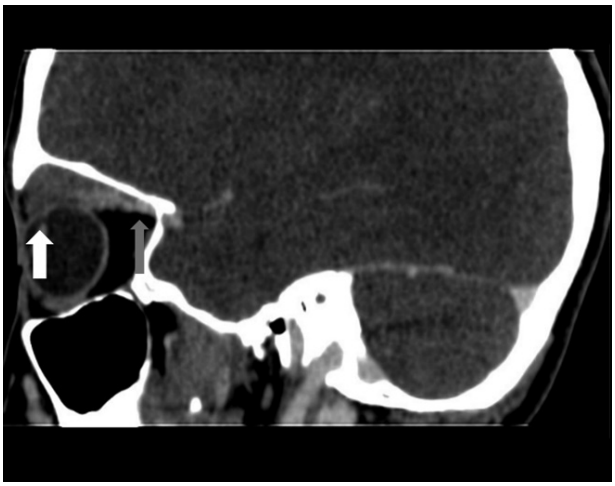

Fig 3: Sagittal reformatted image shows the entire anteroposterior extent of the lesion (white arrow). Multiple tiny enhancing leashes of vessels are seen predominantly towards the posterior aspect of the lesion (red arrow).

DISCUSSION

Intermittent proptosis is a condition characterised by alternating exophthalmos and enophthalmos, which develops gradually and is progressive in nature, with no gender and age predilection. Enophthalmos is usually present, but it is not essential for the diagnosis. Enophthalmos, if present, is apparent only when the head is in the upright position, and exophthalmos

develops almost instantaneous in one eye when venous stasis is induced by lowering the head forward, forced head rotation, hyperextension of the neck, coughing, forced exhalation with or without compression of nostrils or pressure on the Jugular veins(Issiaka et al.,2021). Exophthalmos is not a condition but a sign of a disorder(Mishra et al.,2009). The aetiology of unilateral exophthalmos is multifactorial, rarely represented by intra or extra-conical vascular masses, which account for approximately 10% of orbital tumours. Haemangiomas, venolymphatic malformations (VLM), and orbital varices constitute the maximum proportion of the tumour(Nariman et al.,2015). VLMs are rare benign tumour that accounts for 0.3 to 4 % of all orbital tumours. Orbital varix is a rare condition representing 1.3%-2% of all orbital tumours and is due to ectatic dilation of one or more orbital veins and proliferation of intraorbital venous elements(Pappas et al.,2018). This exophthalmos is non-pulsatile, positionally variable and sometimes painful. Orbital VLMs are also rare, benign, diffuse vascular lesions with a spectrum from venous to lymphatic characteristics(Russin et al.,2015). VLM may remain clinically unapparent or might manifest in childhood as a slow progression of proptosis, periorbital swelling and globe displacement(Smoker et al.,2008). These malformations usually enlarge slowly. Although benign due to their infiltration into orbital contents, gross total resection is frequently impossible(Topilow et al.,2020). Imaging techniques can make a precise diagnosis in such cases. The distensibility of varices during the Valsalva manoeuvre can be easily demonstrated in ultrasound. It is characterised by intermittent anechoic retrobulbar lesion, which shows intrinsic flow during the Valsalva manoeuvre(Walsh et al.,1944). Axial CT images show dilatation of veins. However, the distensibility can be seen during the manoeuvre that increases the venous pressure, such as scanning in the prone position, Valsalva manoeuvre or jugular vein compression. Varices can appear as smooth dilatation, which can be club-shaped or tangled and draining in the systemic circulation. However, they are not associated with any soft tissue mass, whereas in venous lymphatic malformation, there is an isolated heterogeneous hyperattenuating mass. The mass is multicompartmental with hypodense lymphatic and hyperdense venous components.

CONCLUSION

Intermittent proptosis due to Venolymphatic malformation may appear for the first time in adults. A methodical approach can narrow down the diagnosis. A contrast-enhanced CT scan in a prone position may help to clinch the diagnosis in challenging cases. Treatment is controversial because of the lesions variable and unpredictable natural histories. When possible, observation and conservative management are advised. Surgery can be advised in cases where lesions produce significant compression of the optic nerve, but complete excision is impossible for diffuse lesions, and recurrence is common.

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