

Case report

An arteriovenous malformation presenting with proptosis – a case not to be missed

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The Journal of the College of Ophthalmologists of Sri Lanka 2022; 28: 86-87

Key words: proptosis, thyroid eye disease, arteriovenous malformation

Introduction

Proptosis is one of the classical presentations of wide range of clinical conditions such as thyroid eye disease, vascular, infective, inflammatory, cystic, neoplastic and traumatic aetiologies. Some of them being sinister needing early diagnosis and prompt investigation followed by immediate treatment. Arteriovenous malformation is well known to present with proptosis, yet could be missed in imaging of brain due to masking features suggesting other causes. Here we describe such a presentation with proptosis which clinically mimics thyroid associated orbitopathy, presenting mainly with bilateral proptosis, lid retraction and lid lag but a suspicion. The high IOP and negative thyroid profile abnormalities made us to proceed with brain imaging which revealed arteriovenous malformation.

Clinical case

74 years old male presented with bilateral proptosis (Figure 1) and gradual loss of vision over 6 months. Examination revealed, bilateral proptosis being more prominent on left with lid retraction and lid lag. Visual acuity was 6/24 in right eye and NPL on the left. Intraocular pressure (IOP) on left was 40 while it is 38 on right. Gonioscopy finding showed open angles. Fundus examination revealed full cupping on left and 0.8 cup to disc ratio on right, with clear disc margins. Right eye visual field suggested glaucomatous optic nerve damage.

Basic blood investigations including thyroid profile were within normal range. MRI brain and orbit revealed an arteriovenous malformation in the parietotemporal region of the brain (Figure 2 and 3). Digital subtraction angiography (DSA) confirmed it (Figure 4).



Figure 1. Bilateral proptosis seen in the patient.

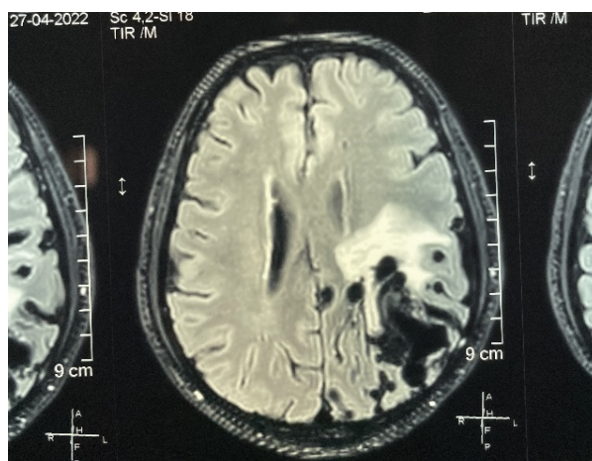


Figure 2. Axial view of the MRI showing temporo-occipital arteriovenous malformation.

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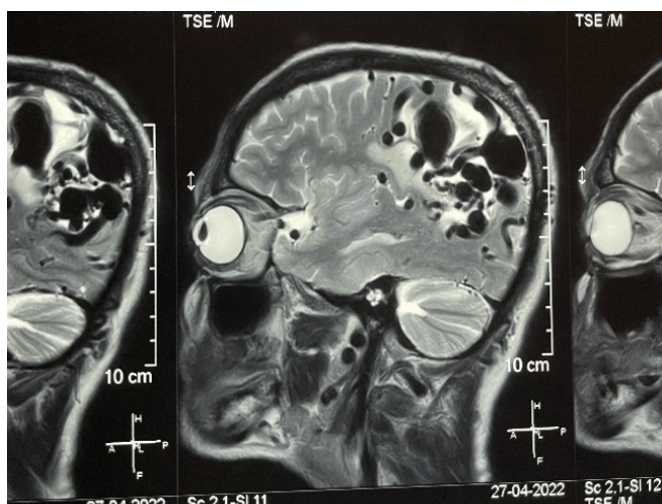


Figure 3. Sagittal view of the MRI showing temporo-occipital arteriovenous malformation.

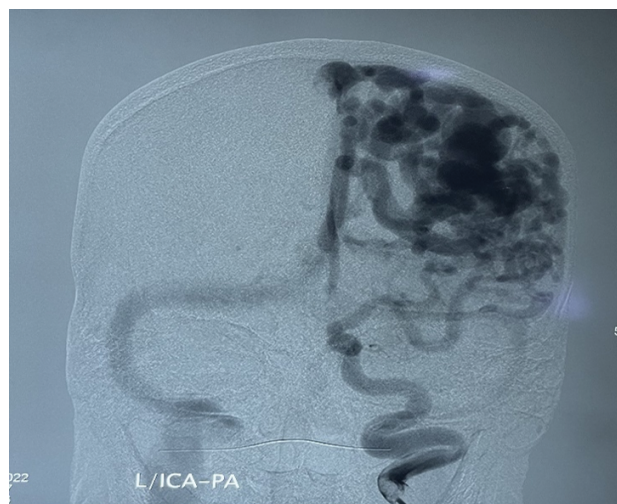


Figure 4. DSA of the patient showing the arteriovenous malformation.

Discussion

Abnormal protrusion of any organ is given the name proptosis. But, it is generally applied to the eyeball. Proptosis due to endocrine causes is implied by exophthalmos. This clinical presentation in a patient can be due to any force acting to displace the globe forward, being any anatomical structure found within the orbit.

The contents of the orbit are mainly globe with its extraocular muscles, vascular structures, nerves, adnexae and connective tissues along with bony architecture around.

The orbital venous blood is drained into cavernous sinus via superior and inferior ophthalmic veins. In this patient arteriovenous malformation also communicates with the cavernous sinus system causing high pressure inside them. Thus, exerted pressure impedes with venous drainage of orbit. Therefore, dilated and engorged orbital veins cause bilateral proptosis.

Intraocular pressure is the force acting on the outer coats of eyeball by its contents. Aqueous humour is produced by ciliary body, circulates from posterior chamber to anterior via pupil and drained through trabecular meshwork and uveoscleral pathway at angle. Thus, drained fluid passes through episcleral veins which in turn goes to ophthalmic veins, then to cavernous sinus system.

Our patient had high intraocular pressure due to back pressure from cavernous sinus impairing his aqueous flow into systemic veins. Persistent high pressure inside the globe has led to his glaucomatous optic nerve damage.

B/L proptosis with prominent lid signs leads to the clinical diagnosis of Thyroid Eye Disease. But AVM of the brain is a rare cause of proptosis. In this case, suspicion of brain pathologies due to high IOP, made us to do MRI brain. AVM might be draining to the sinus system which would have raised the back pressure to the ophthalmic veins explaining the reason for high IOP.

Management

A tentative diagnosis of end stage glaucoma was made while awaiting imaging and patient underwent left eye cataract surgery and trabeculectomy. With MRI, neurosurgical referral was done. Patient is awaiting embolization of AVM in near future while being followed up for IOP control.

Conclusion

This case denotes how crucial it is for ophthalmologists to consider the potential of brain pathologies like AVM as a cause of proptosis and to include brain imaging for definitive diagnosis on selected cases that could save lives.

References

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