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CASE REPORT : CENTRAL RETINAL VEIN OCCLUSION SECONDARY TO CAROTID-CAVERNOUS FISTULA

Ophthalmology

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ABSTRACT

A 50 years old male Patient with Complaints of sudden onset diminution of right eye vision since 15 days. Diminution of vision was profound, painful and progressive in nature. Patient had history of dull aching pain, redness and watering in right eye since month. Patient is a known diabetic since last 4 years and on treatment. On examination patient was found to have engorged episcleral vessels associated with mild proptosis and increased intraocular pressure. His fundus examination showed flame shaped haemorrhages in all quadrants of the fundus suggestive of central retinal vein occlusion. Investigations (OCT) revealed macular oedema due to central retinal vein occlusion and CT angiography showed slow dural carotid-cavernous fistula. Improvement of ocular symptoms were achieved when patient was given intravitreal Anti-Vascular endothelial growth factor and antiglaucoma medication . Along with carotid massage as adviced by Neurologist.

KEYWORDS

Dural carotid-cavernous fistula, central retinal vein occlusion , Carotid massage

INTRODUCTION

The cavernous sinus is a network of venous channels traversed by the intracranial portion of the internal carotid artery. The carotid-cavernous fistula (CCF) is an abnormal communication between cavernous sinus and carotid arterial system (1,2). It is a type of arteriovenous fistula. CCFs are classified as either direct or dural (indirect). Direct CCFs are high-flow fistulas with a direct connection between the internal carotid artery (ICA) and the cavernous sinus. Dural CCFs are low-flow fistulas resulting from communications of cavernous arterial branches and the cavernous sinus.

As arterial blood under high pressure enters the cavernous sinus, the normal venous return to the cavernous sinus is impeded and this causes engorgement of the draining veins, manifesting most dramatically as a sudden engorgement and redness of the eye of the same side.

The mixing of venous and arterial blood leads to a venous hypertension transmitted to the orbital content. The classic clinical triad is pulsatile exophthalmos, ocular bruit and episcleral venous engorgement (2). Fremitus, chemosis, ocular nerve palsy, increased intraocular pressure (IOP) and retinal hemorrhages may also occur, with all manifestations usually being ipsilateral to the fistula (1,2,3).

There are many in literature where the association of carotid-cavernous fistulas with CRVO are reported (7-9). The chances of severe visual loss if the fistula is not treated was emphasized by Drs' Pollock and Miller. Isabelle Brunette, MD, and Dan Boghen, MD, reported a patient with a spontaneous carotid-cavernous fistula that subsequently developed CRVO, and suggested that this complication may be very common. In many literatures it was observed that CRVO might be the main cause of visual loss in patients with a CCF (8)

CASE REPORT

50 years old male came to Ophthalmology clinic with diminution of vision in right eye since 15 days. Diminution of vision was profound, painful and progressive in nature. Patient had history of dull aching pain, redness and watering in right eye since month. There was no history of trauma. There was no diplopia. Patient is a known diabetic since last 4 years and on treatment.

On examination, patient's visual acuity in right eye was 20/200 and in left eye was 20/30. Patient was found to have engorged episcleral vessels associated with mild proptosis and increased intraocular pressure. His fundus examination showed flame shaped haemorrhages in all quadrants of the fundus suggestive of central retinal vein occlusion. Investigations revealed macular oedema due to central retinal vein occlusion and CT angiography showed slow dural carotid-cavernous fistula.

Although most dural fistulas are not life-threatening, some may cause severe ocular morbidity. Indications for treating the vascular abnormality include progressive ophthalmoplegia, progressive proptosis with exposure, uncontrolled elevation of intraocular pressure, retinopathy or optic neuropathy, and abnormal cortical venous drainage noted on MRI.2

The treatment of dural carotid-cavernous fistulas may require medical and surgical modalities. Medical treatment is used primarily to control intraocular pressure. Surgical treatment options primarily include percutaneous embolization of spontaneous DCCFs. This may be performed from a transarterial or transvenous approach(3,4). The transvenous approach accesses the superior ophthalmic vein cutaneously. Platinum coils, detachable balloons and liquid acrylic agents have been used for embolization.

Improvement of ocular symptoms were achieved when patient was given intravitreal Anti-Vascular endothelial growth factor and antiglaucoma medication . He was referred to Neurosurgery department, where he was advised carotid massage. His visual acuity improved & IOP was well controlled with treatment

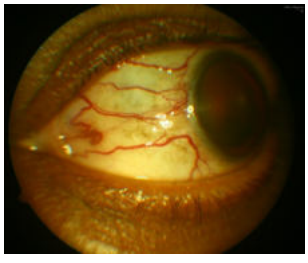

FIGURE:1
Figure 1 showing conjunctival injection with cork-screw vessels

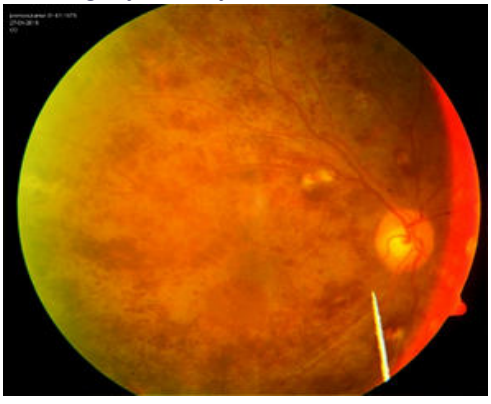

Figure 2: showing Right eye Central retinal vein occlusion with scattered retinal haemorrhages with tortuous retinal veins in all quadrants

INVESTIGATIONS

- Routine Blood Investigations – wnl
- Coagulation profile – wnl
- Carotid doppler-wnl
- Glaucoma workup – open angles , disc - BE - cup disc ratio : 0.4
- Oct CMT : macular edema CMT : 320 microns with LE: Within normal Limits.
- Pateint was advised CT angiography which showed slow dural carotid cavernous fistula .

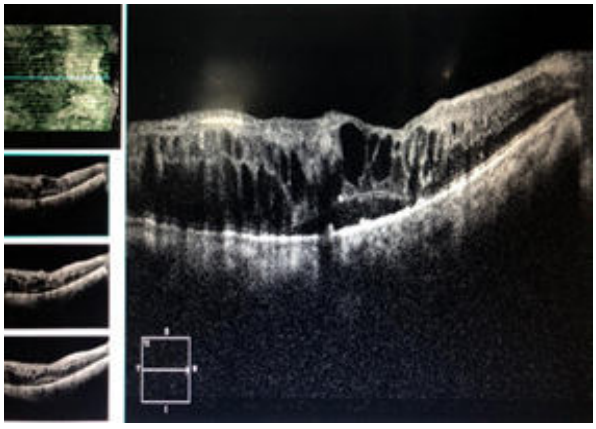

Figure 3: A Ocular Coherence Tomography RE Showing Cystoid Macular Oedema

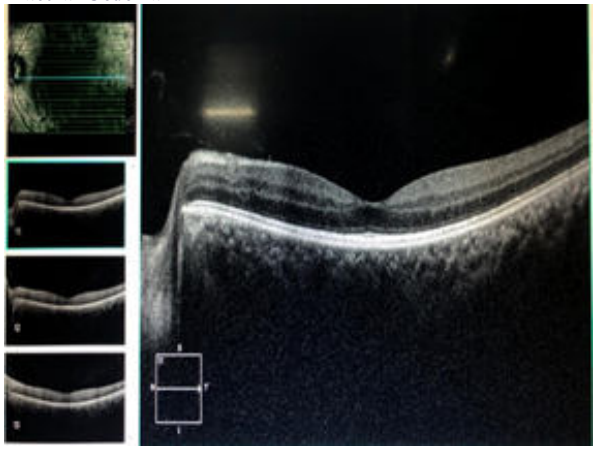

Figure 3: B Ocular Coherenece tomography of Left eye: Normal CMT

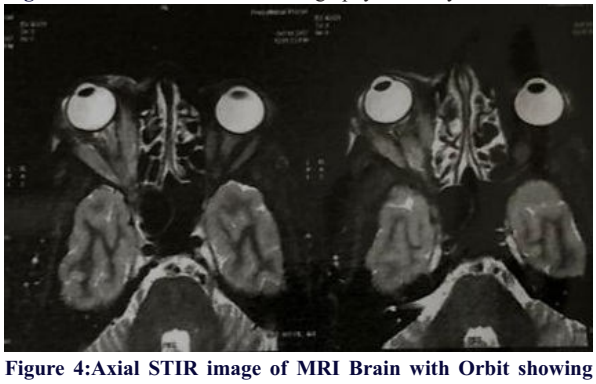

Figure 4:Axial STIR image of MRI Brain with Orbit showing Proptosis Of the right globe with bulky extraocular muscles showing hyperintense signal

DISCUSSION

Central Retinal Vein Occlusion-

CRVO arise from either thrombosis of the central retinal vein at the level of, or posterior to, the lamina cribrosa. It raises the venous pressure, slows arterial blood flow .(6)

The incidence of retinal vein occlusion increases over the age of 50 and is generally associated with glaucoma(10,11)

Risk factors-

A study on CRVO found that hypertension, diabetes and open-angle glaucoma are risk factors for CRVO (12).

In Blue Mountain Eye Study it was evident that history of a systemic vascular occlusive event like stroke or myocardial infarction, increased the risk of retinal vein occlusion(8).

Other systemic illnesses like blood dyscrasias, hypercoagulability state can lead to CRVO.

Treatment-

Primary management of CRVO consists of treatment of underlying systemic medical illnesses.

Secondary management consists of treating the ocular complications of CRVO like macular edema and glaucoma.

Macular Edema

The treatment modalities for macular edema include intravitreal injections of anti-Vascular endothelial growth factor or steroids.

Glaucoma

Glaucoma can arise as a complication of CRVO. Patients with CRVO need to be closely monitored for signs of glaucoma as it generally develops later after 2-3 months.,90 day glaucoma due to neovascularisation.

It was recommended by the CVOS study group to wait until iris, angle or retinal neovascularization developed before initiating PRP. Using anti-VEGF agent can delay laser PRP.

SURGICAL MANAGEMENT

Though there are different surgical options available, the course is variable. Recent surgical options consist of decompression of the central retinal vein by radial optic neurotomy and cannulation of the central retinal vein followed by infusion of tissue plasminogen activator.

CRVO is a potential complication of carotid cavernous fistula with potentially devastating visual consequences. Risk factor modification and treatment of any underlying systemic diseases is very important.

Carotid Cavernous Fistula

Carotid cavernous fistula (CCF) is characterised by abnormal communications between the carotid artery and the cavernous sinus.CCFs can be classified as direct and indirect, depending on whether they are supplied by the internal, external or both carotid artery branches. (13,14). Both forms can cause similar characteristic ophthalmological symptoms and signs. Direct CCF are usually high flow and present with sudden-onset symptoms, whereas indirect CCFs are generally associated with low flow and milder symptoms. A review of the literature showed that central retinal vein occlusion in carotid cavernous fistula may be more common than it was considered before. In fistulas draining directly into the ophthalmic veins, the diagnosis may be straightforward, presenting with symptoms of pulsatile proptosis, diplopia, marked episcleral congestion and chemosis, and a red eye. However, in posterior dural draining CCF symptoms may be mild and difficult to interpret (white-eye syndrome). In both cases, an orbital bruit may be audible with careful auscultation. Venous and arterial stasis causes retinal and choroidal changes such as conjunctiva arterialization, anterior segment ischemia with neovascular changes, papilledema, retinal venous dilatation, retinal hemorrhage, and central retinal vein occlusion, which require appropriate treatment Orbital venous stasis and extraocular muscle engorgement may cause restricted ocular motility associated with diplopia. Alternatively, ophthalmoplegia/diplopia may also be caused by decreased perfusion and ischemia of cranial nerves in the intracavernous sinus

The increase of IOP in CCF is mainly caused by increased episcleral and vortex vein pressure. In other cases, glaucoma may be caused by iris neovascularization. Therapy options depend on the gravity of the conditions. In patients with dural draining CCF and without ocular or cerebral signs, close observation may be adopted . Occasionally, dural draining fistulas may favorably evolve to spontaneous closure, and this may especially occur after diagnostic angiography.

In patients with dural CCF with low ocular or cerebral risk, a manual carotid-jugular manipulation/compression technique has also been adopted

CONCLUSION:

Carotid cavernous fistulas (CCFs) are rare potentially sight-threatening abnormal connections between carotid artery and cavernous sinus which could present with multiple symptoms, subtle in nature. Owing to the risk of vision loss in involved eyes, the ophthalmologist must be aware of the various clinical features of CCF.Treatment options must be carefully considered and adopted in a timely manner, owing to the risks of vision loss associated with high IOP, vascular stasis, and ischemia of the retina and other ocular structures. A multidisciplinary approach of neuroradiologists, neurosurgeons, and ophthalmologists is advisable in complex cases.

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