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Acquired Carotico-Cavernous Fistula Presenting As An Intractable Glaucoma: A Case Report

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The carotico-cavernous fistula is a specific type of dural arteriovenous fistula characterized by abnormal arteriovenous shunting between the internal carotid artery and the cavernous sinus. Young men are more likely than others to develop traumatic carotid-cavernous fistulas (CCFs), possibly because of their increased incidence of trauma in young patients. A relatively high incidence of traumatic CCF has been reported in patients with middle fossa basilar skull fractures.[1] We discuss a MRI and USG features of a case of bilateral traumatic caroticocavernous fistula in a 20 year old male patient due to injury sustained 1 year back. He was later treated by interventional embolization by coiling of fistula. Patient is now asymptomatic and shows resolution of pulsatile proptosis and glaucoma.

Keywords: Carotico-cavernous fistula, Glaucoma, pulsatile proptosis

The carotico-cavernous fistula (CCF) is a specific type of dural arteriovenous fistula characterized by abnormal arteriovenous shunting between the internal carotid artery and the cavernous sinus. These lesions may be classified based on the following: etiology (traumatic vs spontaneous), velocity of blood flow (high vs low), and anatomy (direct vs dural, or internal carotid vs external carotid). A traumatic CCF is one of the cause for loss of vision if left untreated. Young men are more likely than others to develop traumqtic (CCF), possibly because of their increased incidence of trauma in young patients. A relatively high incidence of traumatic CCF has been reported in patients with middle fossa basilar skull fractures.[1] We discuss a MRI and USG features of a case of bilateral traumatic caroticocavernous fistula in a twenty years old male patient due to injury sustained 1 year back.

Case Report

A twenty year old male patient referred from the Department of Ophthalmology for MRI brain & orbit and high frequency ultrasound examination (B-scan) of both

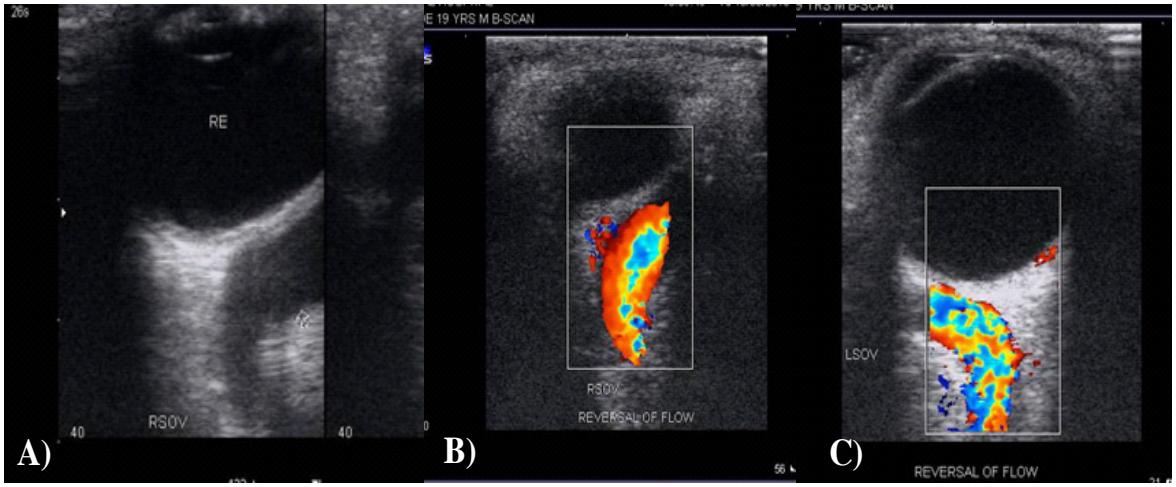

Figure 1: Color Doppler study of eye ball showing congested and dilated ophthalmic veins

the orbits. The patient initially presented to Ophthalmology department with mild blurring of vision. He had history of road traffic accident resulting in fracture of right maxillary bone & right zygomatic bone. Patient was operated by maxillo-facial surgeon by applying titanium plates for fixation of fractures. Patient was asymptomatic for around 8 months after injury. On clinical examination, his vision in right eye was 6/9 & IOP of right eye was 25mm of Hg. All other examination of right eye including ocular movements were within normal limit. Left eye examination was also within normal limit. As there was no obvious cause of raised IOP in right eye, he was advised for removal of titanium plates which were used to fix the fracture maxilla & zygomatic bone presuming that those plates might be causing pressure on episcleral veins/ venous plexus. Titanium plates were removed by maxilla-facial surgeon subsequently. Patient reported after 8 months with raised IOP of 28mm of Hg in right eye with few dilated circumciliary blood vessels & few dilated episcleral veins in right eye. On palpation of globe, there was no pulsation felt or on auscultation of orbit no bruit was heard. Patient was treated with the intraocular pressure lowering agents to prevent glaucomatous damage of optic nerve. But the patient did not responded to intraocular pressure lowering agents. At the present visit, more & more dilated episcleral veins became prominent along with few circumciliary blood vessels. This time while palpating right eye arterial pulsations were felt. At the same time, arterial pulsations were felt in left eye also with rest of the ocular examination in left eye was within normal limit. A provisional diagnosis of bilateral carotico-cavernous fistula was made, probably due to injury sustained 1 year back. Patient was referred to the department of Radio-diagnosis for confirmation of diagnosis.

High frequency ultrasound examination of the both orbits was done using Siemens Sonoline G60 S linear 7.5 MHz probe. Axial length was normal on both sides. Anterior and posterior chambers of the eyeballs were normal. Both superior ophthalmic veins were dilated and tortuous. On colour Doppler study, there was arterialisation of the flow within the superior ophthalmic veins with peak systolic velocity of superior ophthalmic veins approximately 30-40 cm/sec. Colour Doppler study showed a typical ying-yang flow pattern in dilated tortuous superior ophthalmic veins. **(Figure 1)**

MRI of Orbit (Plain & Contrast study) with MRA & MRV was done using Philips Achieva 1.5 Tesla high gradient MRI scanner. T1W, T2W, FLAIR axial and coronal images followed by post gadolinium T1W images were obtained using multiecho-multiplanar technique. Also MR angiography and MR venography was also performed. **(Figure 2)**

Clinical diagnosis of bilateral Carotico-cavernous fistula was confirmed on MRI and USG of orbit.

Discussion

Carotid-cavernous sinus fistula is an abnormal communication between the internal or external carotid arteries and the cavernous sinus. These lesions may be classified based on the following: etiology (traumatic vs spontaneous), velocity of blood flow (high vs low), and anatomy (direct vs dural, or internal carotid vs external carotid). The cavernous sinuses consist of extradural venous plexuses surrounded by a dural fold. The intra-cavernous internal carotid artery with its peri-arterial sympathetic plexus runs between the venules of the parasellar venous plexus. Carotid–cavernous fistula can result from traumatic laceration of the carotid artery or from rupture of an aneurysm into the surrounding venous sac establishing a direct arteriovenous fistula between the internal carotid artery and the venous spaces of the cavernous sinus. This results in short-circuiting of the arterial blood into the venous system of the cavernous sinuses. Direct carotid-cavernous sinus fistulae, which represent 70-90% of all carotid-cavernous sinus fistulae in

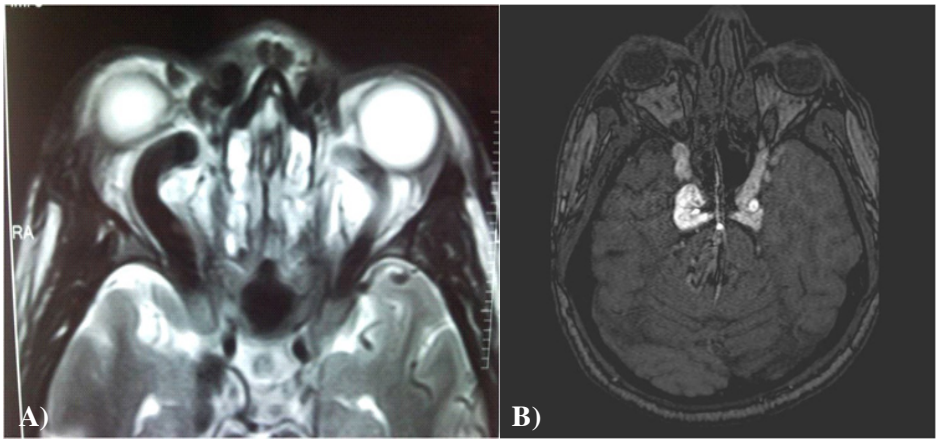

Figure 2: MRI brain and orbits showling congested ophthalmic vein and cavernous sinuses

most series, are characterized by a direct connection between the intra-cavernous segment of the internal carotid artery and the cavernous sinus. These fistulae usually have high rates of arterial blood flow and most commonly are caused by a single traumatic tear in the arterial wall.

Dural carotid-cavernous sinus fistulae are formed by a communication between the cavernous sinus and meningeal branches of the internal carotid artery, external carotid artery, or both. These fistulae develop without any antecedent trauma or manipulation. They usually have low rates of arterial blood flow and almost always produce symptoms and signs spontaneously. The lesions may represent congenital arteriovenous malformations, which develop spontaneously or in association with atherosclerosis, systemic hypertension, collagen vascular disease, pregnancy, and during or after childbirth[2].

Most of the patients with a direct carotid-cavernous sinus fistula experience progressive ocular complications if the fistula is left untreated. Increasing proptosis, conjunctival chemosis and visual loss occur over months to years, with central retinal vein occlusion and secondary glaucoma representing the most severe ocular complications.

Young men are more likely than others to develop traumatic carotid-cavernous fistulas (CCFs), possibly because of their increased incidence of trauma in young patients. A relatively high incidence of traumatic CCF has been reported in patients with middle fossa basilar skull fractures[1]. Past history of significant trauma and fracture of base of skull is important in the diagnosis of post traumatic direct carotico-cavernous fistulas. Patients may present with redness of the eye, diplopia, buzzing or swishing sounds, decreased vision, bulging eye and facial pain in the distribution of the first (and rarely the second) division of the trigeminal nerve.

Computed tomography (CT) scan, magnetic resonance imaging (MRI), and orbital ultrasound often help to confirm the diagnosis, demonstrating extraocular muscle enlargement, dilation of one or both superior ophthalmic veins, and enlargement of the affected cavernous sinus. In all carotid-cavernous sinus fistulae the definitive diagnostic test is cerebral arteriography with selective catheterization of the internal and external carotid arteries on both sides, so that all arterial contributions to the fistulae can be visualized. Intra-arterial subtraction angiography is generally the preferred technique. Because of the invasive nature of the procedure and radiation hazard magnetic resonance imaging is now a days is preferred imaging modality. Non invasive imaging and multiplanar technique as well as MR angiography and MR venography helped MRI to replace the conventional imaging modalities in diagnosis of carotico-cavernous fistulas.

On MRI, dilated superior ophthalmic veins, hypertrophy of the extra-ocular muscles and Enlarged cavernous sinus with a convex shape to the lateral wall is well demonstrated. Dilated superior ophthalmic veins are demonstrated as a dark signal due to flow voids secondary to flowing blood. High resolution ultrasound of the orbits using high frequency ultrasound transducers allows real time imaging of the eyeballs and retro bulbar region. Colour Doppler study demonstrates dilated and tortuous superior ophthalmic veins with arterialisation of the flow within them. The optimal treatment of a direct carotid-cavernous sinus fistula is closure of the abnormal arteriovenous communication with preservation of internal carotid artery patency[3, 4].

Once a fistula is closed, symptoms and signs usually begin to improve within hours to days. The rate and extent of improvement are associated with the severity of the signs and the length of time the fistula was present. The patient was later treated by interventional embolization by coiling of fistula. Patient is now asymptomatic and shows resolution of pulsatile proptosis and glaucoma. The abnormal episcleral veins are now not seen in both eyes.

Conclusion

The present case report shows the importance of early diagnosis and management of post-traumatic carotico-cavernous fistula in individuals who present with pulsatile

proptosis and progressively worsening glaucoma. Delay in diagnosis and treatment would lead to permanent loss of ocular vision. The case report also emphasises the characteristic B-scan and MRI findings in brain and orbit in a suspected case of carotico-cavernous fistula.

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