Direct carotid-cavernous fistula and glaucoma: case reports

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Abstract

A carotid-cavernous fistula is an abnormal communication between carotid artery system and the cavernous system. The causes that determines the onset of the fistula are trauma or spontaneous. The fistula receives venous blood from the eye and the brain, from the ophthalmic veins and the superficial middle cerebral vein. The abnormal shunt determines raised venous pressure on the cavernous sinus, determining ophthalmic veins stasis. It appear ocular congestion, raised intraocular pressure, exophthalmia, diplopia, decreased visual acuity or cranial nerves paralysis.

Glaucoma is a potential complication that appear in carotid-cavernous fistula from increased pressure in the episcleral veins.

Carotid-cavernous fistula diagnosis can be determined clinically and imagistically. This pathology is treated successfully in 90% of all cases, the mortality and morbidity rates are very low.

This study presents two case reports, diagnosed with direct carotid-cavernous fistula and glaucoma.

Keywords: direct carotid-cavernous fistula, glaucoma

Direct carotid-cavernous fistula appears in 70-90% of all cases with carotid-cavernous fistulae cited by Keltner et al. in 1987, Debrun et al. in 1988. (1) Direct fistula can be localized in any intracavernous portion of internal carotid artery. Iatrogenic fistulae are reported in transsphenoidal surgery of the pituitary gland or internal carotid artery lesion in endarterectomy or ethmoidal sinus surgery. Some direct fistulae can be caused by rupture of an intracavernous aneurysm. Ehlers-Danlos syndrome, arterial hypertension or artheriosclerosis were associated with carotid-cavernous fistula.(2)

Cerebral trauma causes 75% of carotid-cavernous fistulae at young patients, with pulsatile exophthalmia. 25% of carotid-cavernous fistulae are spontaneous, specially middle aged women with atherosclerosis. (3) Glaucoma is a potential complication of the direct untreated carotid-cavernous fistula, mentioned by Sugar and Meyer in 1940, and by Madsen in 1970, that appears in 30-50% of all cases.(3)

Case I

A 63 year old female patient, B.I., presented with ocular congestion that appears two weeks ago, retro-orbital mild pain and left eye diplopia.

Clinical exam revealed the 6th left nerve palsy, 3 mm exophthalmia, discreet orbital bruit and dilated conjunctival vessels. On Goldmann tonometry the intraocular

pressure was 22-25 mm Hg. During hospitalization were administered topical ocular hipotensors and the diurnal pressure curve was assessed, the pressure normalized between 16 and 18 mm Hg. The MRI-angiography revealed a left carotid-cavernous fistula; the patient was transferred in a neurosurgical clinic. All ocular symptoms and signs resolved in four weeks after intravascular closure of the fistula with a detachable balloon (Figure 1a, 1b). The patient continued the ocular hipotensors treatment during neurosurgical hospitalization. At eight weeks, the patient presents in the ophthalmological clinic for re-assessment, the intraocular pressure was between 14-17 mm Hg, without antiglaucomatous medication. The ophthalmoscopic exam didn't revealed pathological findings of the cup-disc ratio, that was 0.2-0.3, the visual field was normal as well, regarding to her age.

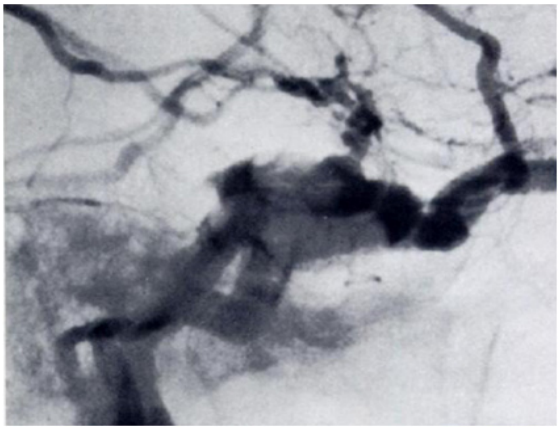

Figure 1a Cerebral angiography of the carotid-cavernous fistula

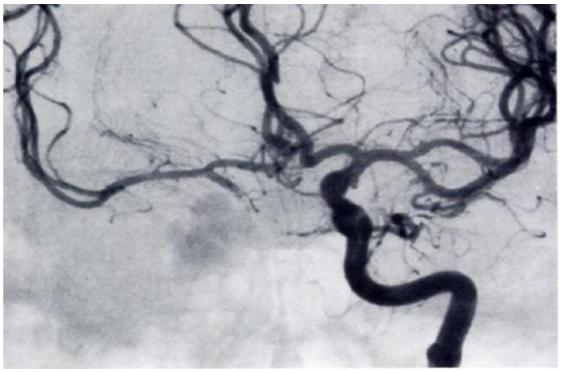

Figure 1b Intravascular closure of the fistula with a detachable balloon

Case II

A 25 year old male pacient, D.M., diagnosed with traumatic carotid-cavernous fistula, presented in the ophthalmological clinic with right eye chronic glaucoma. Clinical exam revealed a discreet right eye exophthalmia, dilated conjunctival vessels and the left anterior pole didn't present any pathological finding. On Goldmann tonometry, the intraocular pressure was between 32 and 38 mm Hg.

The ophthalmoscopic exam on the right eye revealed a mild glaucomatous cupping, cup-disc ratio 0.6 and in the left eyes a normal aspect, without pathological cupping. The MRI-angiography revealed the presence of a carotid-cavernous fistula (Figure 2); the patient refused the neurosurgical treatment.

During hospitalization we administered antiglaucomatous topical medication, intraocular pressure was above normal level (32-34 mm Hg); we performed filtering surgery (trabeculectomy) for intraocular pressure decrease; after surgery the intraocular pressure level normalize, between 16 and 18 mm Hg. At six weeks after the surgery, the patient had under antiglaucomatous topical treatment values of intraocular pressure between 14 and 16 mm Hg. The ophthalmoscopic exam revealed on the right eye a glaucomatous cupping, the left eye was normal. We prescribed antiglaucomatous topical therapy to continue on right eye and periodical ophthalmological assessment.

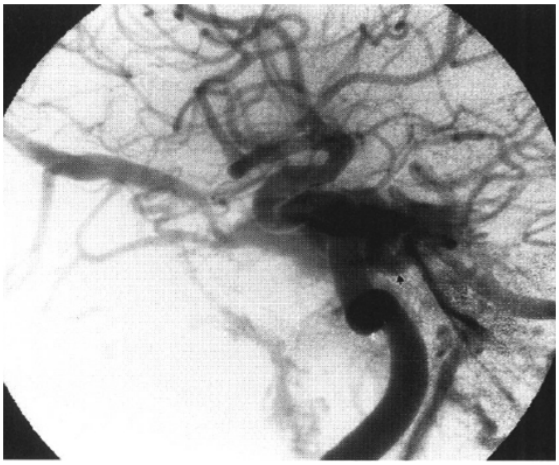

Figure 2 Cerebral angiography of the carotid-cavernous fistula in case 2

Discussions

Direct carotid-cavernous fistula has a frequency up to 90% of all carotid-cavernous fistulae and glaucoma is a potential complication of untreated direct carotid-cavernous fistula.

In 1970, Madsen mention untreated carotid-cavernous fistula and glaucoma association. (1) Henderson and Schneider find that glaucoma was diagnosed on 30-50% of all cases with untreated fistula. (1)

The most common form is determined by the raise of pressure in episcleral veins. In 1973,Spencer et al. mention cases of patients diagnosed with direct carotid-cavernous fistula and neovascular glaucoma, always associated with retinal chronic hypoxia and retinal neovascularization, after central retinal vein occlusion. (1)

Ocular findings of the first patient are related with blood redistribution from the orbital veins, determining decreased arterial blood flow at the cranial nerves level, venous stasis in anterior and posterior circulation in the eye and the orbit, raised pressure in episcleral veins. Ocular findings are localized unilateral, same path with the fistula, may be bilateral or in rare cases contralateral. Ocular finding path depends of venous blood drainage thru cavernous sinus, especially the intercavernous system and the basilar system, with thrombosis at the sinus level or the superior ophthalmic vein. Trauma can determine bilateral cavernous sinus fistula.

Carotid-cavernous fistulae (direct communications between internal carotid artery and the cavernous sinus) (4), determine a high blood flow, accompanied by unilateral or bilateral pulsatile exophthalmia, orbital bruit, ocular chemosis and diplopia. (5) Fistula associates with moderate or severe eyelid inflammation, cyanoses and palpebral derma thickening appear in chronic disease. (6) Ocular pulsation is caused by a pulsatile wave from the internal carotid artery level or the ophthalmic artery thru dilated ophthalmic veins. Orbital veins become tortuous and secondary congestion of the orbital soft tissue appears, increase of the venous pressure and decrease of the arterial perfusion, local hypoxia, that determines tissue edema and a certain degree of ophthalmoplegia, resulting in anterior segment ischemia.

Conjunctival chemosis appears in almost all patients diagnosed with direct carotid-cavernous fistula, preceeding the exophthalmia in most cases. (7) Dilation of the conjunctival vessels is the most obvious sign of this disease. Keratopathy is diagnosed frequently, associated with the exophthalmia. (8) Cornea can present an edematous aspect, due to secondary glaucoma or anterior segment ischemia. Diplopia is caused by common oculomotor nerves damage, ocular movement is limited, complete ophthalmoplegia can occur. Ophthalmoplegia cand be caused by the damage of one or multiple oculomotor

nerves due to compression or ischemia; the most affected oculomotor nerve is the abducens nerve, but also the trochlear nerve can be affected, which is situated in the profound layer of the lateral wall of the cavernous sinus. The decrease in the visual acuity produces immediately, being caused by optic nerve damage or it can appear late due to vitreous hemorrhage, retinal dysfunction, anterior ischemic optic neuropathy or corneal ulcer.

Visual acuity decrease pathology is explained by retinal dysfunction, decreased perfusion pressure in ophthalmic artery and increased venous pressure. The result is chronic hypoxia of the retinal cells. The eye fundus exam reveals dilated veins with spontaneous pulsation, optic disc swelling, retinal hemorrhage, venous stasis retinopathy or venous occlusion. Sever pain and local inflammation can suggest ophthalmic vein thrombosis, which can be diagnosed by magnetic resonance imaging or ocular echography.

Ocular and orbital pain, facial hyperesthesia can be caused by compression of the maxillary and the ophthalmic trunks of trigeminal nerve at the cavernous sinus level.

Glaucoma is a potential complication which can appear in direct carotid-cavernous fistula. (1) The most common form of glaucoma is caused by increased pressure in the episcleral veins as we presented in the two cases. (8) Intraocular pressure is moderately raised (24 mm Hg), but cases cited have intraocular pressure up to 50-60 mm Hg, the normal level being 10-21 mm Hg, the fistula often associates with central retinal artery occlusion. Orbital congestion is associated with glaucoma, intraocular pressure in these cases being severely raised and the medical treatment is difficult. Neovascular glaucoma can appear in patients with carotid-cavernous fistula, following chronic retinal hypoxia, retinal neovascularization or after central retinal vein occlusion. In rare cases, some patients develop angle closure glaucoma by raised pressure in the orbital veins which determines choroidal and iris congestion with forward pulling of the iridolenticular diaphragm, determining angle closure.

The diagnosis is based on the clinical findings: rapidly developed conjunctival chemosis, exophthalmia and ocular congestion. Patient can present a trauma in the past, am intracavernous aneurysm rupture or diplopia.

The paraclinical exams used to establish the diagnosis are: tonometry, CT scan, MRI-angiography, ocular echography, cerebral angiography (which helps identifying the presence of the fistula).

The optimal treatment of direct fistula is the closure of the abnormal communication with preserving of the internal carotid artery. There are many surgical options for closure: direct surgical closure of the affected intracavernous portion of the internal carotid artery, isobutyl-2-cyanoacrilate embolization, particles of polyvinyl alcohol or a detachable balloon placed in the cavernous sinus, in the arterial or venous path, thru inferior petrous sinus or superior ophthalmic vein, directly in cavernous sinus. (9) Once the balloon has reached the cavernous sinus it is inflated and detached determining fistula closure. The complications of this therapeutically method include transitory or permanent cranial nerve palsies, visual field loss or pseudo-aneurysm development. (10) It was mentioned in the literature spontaneous closure of fistula in 3 days up to 18 months in 10-60% of all patients. (1)

After the successfully closure of the fistula, almost all symptoms and ocular signs disappear or improve. (11) Ocular pulsation, eyelid edema, conjunctival chemosis, conjunctival vessels dilation, stasis retinopathy, optic disc swelling and raised intraocular pressure disappear immediately. Exophthalmia decreases but it may not disappear entirely. Similarly, the patients with ophthalmoplegia can have a normal motility and diplopia can disappear. The only symptom of the direct carotid-cavernous fistula that don't improve is the visual acuity loss due to optic neuropathy, that's why the treatment must be commenced as urgent as possible that the irreversible optic nerve lesions don't appear.

Glaucoma necessitated rapid treatment; ocular hypotensors were administered in the first case or the surgical treatment (trabeculectomy) in the second case. The therapy target was to normalize the intraocular pressure and to reduce ischemia and ocular necrosis.

Beta adrenergic antagonists, carbonic anhydrase inhibitors and alpha 2 agonists have reduced the intraocular pressure in the first case. Pilocarpine and prostaglandin analogues are not efficient in this case. Prostaglandin analogues treatment can be associated with episcleral venous dilation. Filtering surgery was efficient in the second case and made a stable, uniform decrease of the intraocular pressure, when the medical treatment cannot be administered, the patient refused it.

In the second case, the trabeculectomy was the surgical method we choose: a scleral flap was created, two-thirds thickening, a rectangular shape (six by four mm), 1,5 mm advance in clear cornea, the profound scleral flap was excised, approximately 2 by 2 mm at the limboscleral junction (the gray zone), tissue that contain the trabecular meshwork. (12)

When the trabeculectomy doesn't provide an efficiently result, we recommend the artificial drainage systems or the trabeculectomy augmented with antimetabolites (5-fluorouracile or mytomicin C).

Conclussions

Direct carotid-cavernous fistula doesn't necessitates rapid treatment, it can be delayed; the untreated fistula lead to progressive ocular symptoms.

In months or years, this disease evolves to exophthalmia, chemosis and total visual acuity loss due to glaucoma, ischemic optic neuropathy. In the first case, ocular symptoms necessitate paraclinical exams which showed the carotid-cavernous fistula, surgically treated, the ocular symptomatology disappear after the surgical treatment.

The other case with carotid-cavernous fistula necessitates filtering surgery for ocular pressure balance and visual parameters normalize because the patient refused the neurosurgical treatment and doesn't accept the anti-glaucomatous therapy.

Even if the direct carotid-cavernous fistula doesn't present vital risk, it can produce intracerebral hemorrhage, ischemic optic neuropathy, secondary glaucoma and primary open angle glaucoma, diseases that without treatment determine irreversible visual loss.

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