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A Continuous Red Eye.... Case Report about A Carotid Cavernous Fistula

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Abstract

Purpose: To present a case of indirect carotid cavernous fistula successfully treated with endovascular approach.

Case report: A 46-year-old man presented with a progressive, painless visual loss and conjunctival injection of the right eye with one month of evolution. On ophthalmology examination, the patient had best corrected visual acuity (BCVA) of 20/30 in right eye and an intraocular pressure (IOP) of 20mmHg. The patient still had pulsating proptosis and an orbital bruit on the auscultation. Slit-lamp examination revealed corkscrew episcleral vessels. The patient was submitted a magnetic resonance angiography that confirmed the presence of an indirect carotid-cavernous sinus fistula. The patient was submitted to endovascular remobilization by trans-arterial approach for closed the fistula with complete clinical and imaging resolution. Two months after the procedure, the visual acuity was 20/20 and the intraocular pressure was 8mmHg.

Discussion: Carotid cavernous fistulae are an uncommon disease and may be misdiagnosed when there are few signs and symptoms on presentation. Different subtypes of CCF cause different clinical signs, with varying speeds of onset and severity. The diagnosis is based on clinical findings and by imaging studies. However, the definite diagnosis is only possible with cerebral angiography. It is important to consider risks and benefits in relation to endovascular treatment mainly in indirect CCF.

Keywords: Red eye; Carotid cavernous fistula; Endovascular treatment

Introduction

Carotid cavernous fistula (CCF) is an abnormal communication between the cavernous sinus (CS) and the carotid arterial system. Anatomically these fistulas can be divided into 2 types, direct and indirect or dural CCF. Direct CCF, corresponding to 70-80% of all cases and they are characterized by a direct connection between the intra cavernous segment of the internal carotid artery (ICA) and the CS. 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 CCF are characterized by a communication between the CS and one or more meningeal branches of the ICA, external carotid artery (ECA), or both. These fistulae usually have low rates of arterial blood flow and almost always produce symptoms and signs spontaneously, without any antecedent trauma or manipulation. 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 [1,2]. The diagnosis is based on patient's signs and symptoms and imaging studies such as Computed Tomography scan (CT scan) or magnetic resonance imaging (MRI) of the orbits where the most specific sign is the dilation of the superior ophthalmic vein (SOV). However, definite diagnosis, fistula characterization and planning of endovascular treatment are only possible with cerebral angiography [2,3]. Most CCFs are not life threatening, but the involved eye is at risk. Spontaneous closure from thrombosis of CS is unlikely (especially those that occur after trauma or in high flow fistulas). Dural fistulas may undergo spontaneous closure, especially after diagnostic angiography. Main indications for treatment include glaucoma, diplopia, intolerable bruit or headache, and severe proptosis causing exposure keratopathy [2,4].

Case Report

A 46-year-old man with a past medical history of chronic liver disease with portal hypertension, presented with a progressive, painless visual loss and conjunctival injection of the right eye with one month of evolution. He was treated one month ago with antibiotic drops for a conjunctivitis without

improvements of symptoms. On ophthalmology examination, the patient had best corrected visual acuity (BCVA) of 20/30 in right eye and 20/20 in left eye and an intraocular pressure (IOP) of 20mmHg and 10mmHg, respectively. The patient still had pulsating proptosis and an orbital bruit on the auscultation. There was no restriction on eye movements or diplopia. Slit-lamp examination of the right eye revealed a superior swollen eyelid without pain, inferior chemosis and corkscrew episcleral vessels (Figure 1). Fundo scopy showed slight venous tortuosity. Slit-lamp examination and fundo scopy of the left eye was unremarkable. For clinical suspicion of CCF, the patient was submitted a magnetic resonance angiography that confirmed the presence of an indirect carotid-cavernous sinus fistula supplied by the right accessory meningeal artery and draining to the superior ophthalmic vein. This exam showed the asymmetry between the cavernous sinuses (Figure 2), the orbital muscles and the proptosis of the right eye (Figure 3). The exam still demonstrate dilation of the right superior ophthalmic vein (Figure 4). Antihypertensive eye drops was initiated in right eye and due to the symptoms presentation we decided referred the patient for diagnostic and therapeutic cerebral angiography. The patient was submitted to endovascular embolization (Figure 5 & 6) by trans-arterial approach for closed the fistula with complete clinical and imaging resolution. Two months after the procedure, the visual acuity was 20/20, the intraocular pressure was 8mmHg and the slit lamp examination was unremarkable (Figure 7).

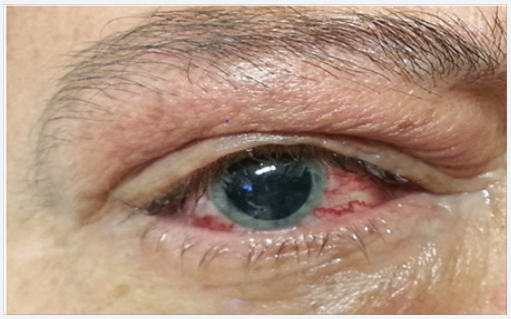

Figure 1: Right red eye demonstrate corkscrew episcleral vessels.

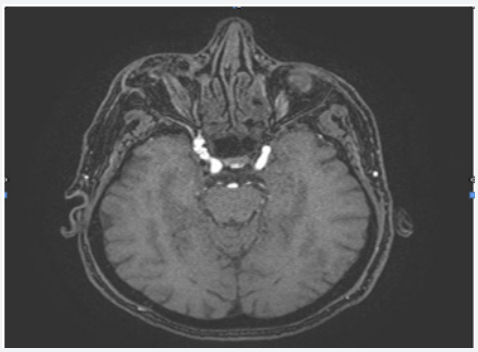

Figure 2: MRI: asymmetry between cavernous.

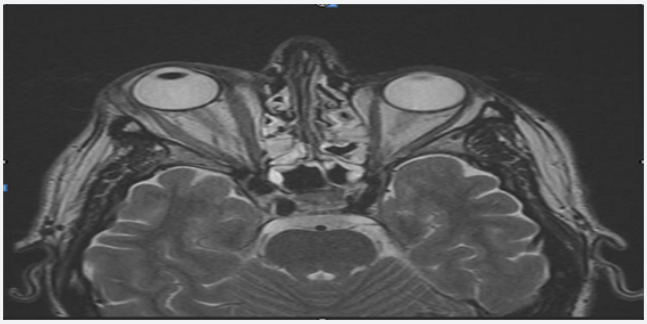

Figure 3: MRI: proptosis and enlargement of the extrinsic muscles.

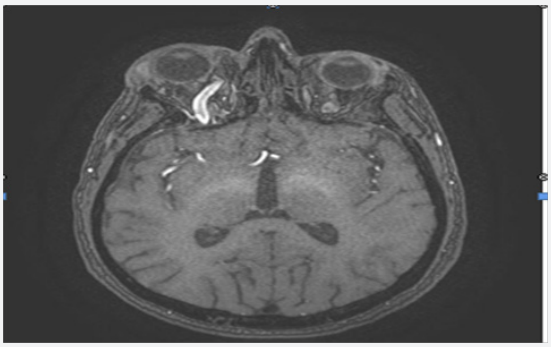

Figure 4: MRI: VOS dilated.

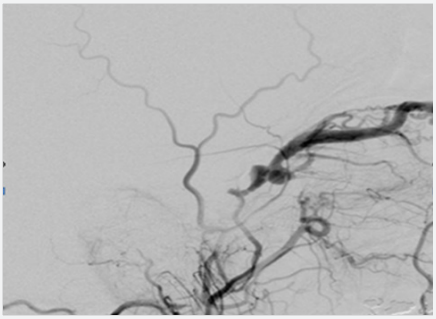

Figure 5: Cerebral angiography: VOS before embolization with cyanoacrylate.

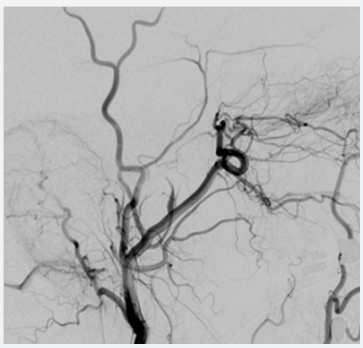

Figure 6: Cerebral angiography: VOS after embolization with cyanoacrylate.

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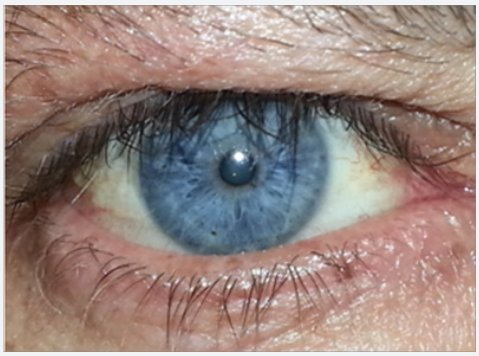

Figure 7: Right eye two months after embolization.

Discussion

CCF are an uncommon disease and may be misdiagnosed when there are few signs and symptoms on presentation. Several classifications of CCF exist depending on anatomy, etiology and patho physiology. The most used classification has been proposed by Barrow et al. [5] and is based on the arterial supply of these communications. Type A CCF results from direct communication between ICA and CS, type B results from a communication between dural ICA branches and CS, type C results from communication between dural ECA branch and CS and type D results from a communication from dural branches of ICA and ECA branches to CS. CCF type A is the only type with direct communication between the ICA and the CS, therefore high flow is usually present. A compilation of different series shows that brain trauma is responsible for up to 80% of cases, the others occurring spontaneously such as aneurysm rupture or weakness of the vascular wall in cases of collagen deficiency. While acute forms occur more often in young men, spontaneous forms tend to occur in middle-aged women [5-7]. Different subtypes of CCF cause different clinical signs, with varying speeds of onset and severity, which is usually higher in direct forms. Pathogenesis involves CS dilation and retrograde drainage (cortex and orbit), with increased vascular volume and enlarged orbital muscles, increased capillary transudation, elevated episcleral venous pressure, and impairment of the optic nerve and retina due to stasis. In direct CCF, these events lead to a more acute presentation with more abundant signs and symptoms [8]. Indirect forms, however, have a milder or minimal presentation. This is possibly due to the fact that in many cases, drainage of dural shunts often occurs to the basal and petrosal sinus, and only in the presence of thrombosis there is flow back into the orbit [2,5]. Clinical signs range from mild paresis and dysfunctions (nerves III, V, VI) without orbital congestion to pulsatile proptosis (milder in indirect forms). Characteristic symptoms include pulsatile exophthalmos, murmur and venular dilation with chemosis (Dandy's triad) [9,10]. Diagnostic suspicion is stronger in cases with a variety of characteristic signs. However, the gold standard for any type of CCF is cerebral angiography. It identifies the type, location and size of the connection, examines the vascular surroundings and co-existing alterations, especially ischemic effects on the cortex. Initial evaluation is done with CT scanor MRI. However, normal results in these tests do not exclude the diagnosis. Images can assess proptosis, CS and SOV dilation and enlargement of the extrinsic muscles [11].

Emergency endovascular treatment of CCF is indicated in the presence of pseudo aneurysm, large CS varix, venous drainage toward the cortex, thrombosis extending beyond the fistula, increased intracranial pressure, altered mental status, proptosis and/or progressive loss visual acuity, brain haemorrhage, and ischemic stroke [2,4,12]. In other situations, the severity of the condition versus the possibility of spontaneous resolution should be taken into account, especially in dural forms [2]. The endovascular approach is the current procedure of choice, either through an arterial or venous access. Different natural or synthetic occlusive materials can be used. Proptosis, chemosis and keratopathy should be managed according to severity. Lubricants and topical antibiotics (when are often necessary. For glaucoma, drugs that reduce the production of aqueous humour are indicated (beta-blockers, alpha-adrenergic agonists, carbonic anhydrase inhibitors), while drugs promoting drainage are ineffective (pilocarpine and prostaglandin analogues) [11]. This case illustrate an indirect CCF that was treated with the endovascular approach. We decided to treat because the patient presented with proptosis and progressive loss visual acuity. This reinforces the importance to considering the literature and the signs and symptoms of our patients in order to obtain the best results.

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