



Painful Blindness: A Diagnosis not to be Missed

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Abstract

A 62-year-old female was admitted with a history of left sided headache, mild proptosis and left ocular redness occurring during the last weeks. A brain MRI showed findings suggesting a carotid fistula. The cerebral angiogram highlighted an indirect Barrow type D left carotid-cavernous fistula which was treated with embolization. The following day, the patient complained of left retro-orbital pain and loss of vision. The examination showed the following findings (in the left eye): mydriatic nonreactive pupil, vision loss, conjunctival chemosis and an intraocular pressure of 33 mmHg. The new angiogram showed an almost complete closure of the fistula and the brain MRI demonstrated a choroidal detachment along the medial wall of the left ocular globe which provoked acute angle closure glaucoma. Considering that an early diagnosis and treatment are crucial to the prognosis, neuro-ophthalmologists should consider this entity in the differential diagnosis of a patient with worsening ocular symptoms after a carotid-cavernous fistula embolization.

Keywords: Carotid-Cavernous fistula; Choroidal detachment; Angle closure glaucoma; Embolization; Angiography cerebral

Introduction

Carotid cavernous fistulas (CCFs) are anomalous connections between the carotid arterial system and the cavernous sinus and they are normally classified in two main categories, depending on the arterial system involved. The first group, direct or high flow shunts (type A), are characterized by a direct connection between the internal carotid artery and one or more of the venous channels within the sinus; the second group are indirect or low flow shunts, which arise from the dural branches of internal, external carotid system or both (type B, C, D respectively) [1]. The symptoms and signs produced by these lesions depend on several factors but the majority of them are ocular, usually ipsilateral to the fistula. Endovascular embolization currently represents the first therapeutic modality for CCFs because it is associated with high occlusion and low complication rates [2].

In this report, we describe a patient with loss of vision and acute angle closure glaucoma (AACG) secondary to choroidal detachment, a very infrequent complication after carotid cavernous fistula embolization.

Case Report

A 62-year-old female was admitted to emergency care with a history of left sided headache, mild proptosis and left ocular redness occurring during the preceding four weeks. On admission, the ophthalmologic examination revealed a subtle exophthalmos with epibulbar congestion and some tortuous corkscrew-type blood vessels in the left eye (Figure 1). The visual acuity was 20/25 Oculus Dexter (OD) and 20/20 Oculus Sinister (OS), with normal colour vision in the Ishihara pseudo-isochromatic plates. The pupils were isocoric and reactive to light and there were no limitations in the ocular motility, ptosis nor bruits. The funduscopy examination showed slightly dilated retinal veins and optic disc swelling without retinal hemorrhages in OS. Intraocular pressure was 18 mmHg OD and 23 mmHg OS. Medical history was significant only for hypertension and she denied suffering any head trauma.

Laboratory tests, which included thyroid profiles such as T3, T4, TSH and TSH receptor antibody were all within normal ranges. A cranial MRI and a 3D time-of-flight (TOF) MR angiography revealed proptosis, an enlarged left superior ophthalmic vein (SOV) and a dilated left cavernous sinus with multiple abnormal flow voids which filled in early during arterial phase. Digital subtraction angiography (DSA) confirmed an indirect Barrow type D left carotid cavernous fistula originating from both arteries, the right external carotid (through internal maxillary artery) and from the right internal carotid (through meningo-hypophyseal trunk) (Figure 2A and B). No aneurysms were identified. The patient underwent transarterial embolization with detachable coils resulting in an

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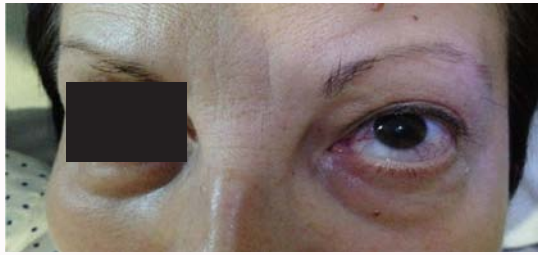


Figure 1: External photograph showing a mild exophthalmos, redness and dilated episcleral vessels in the left eye.

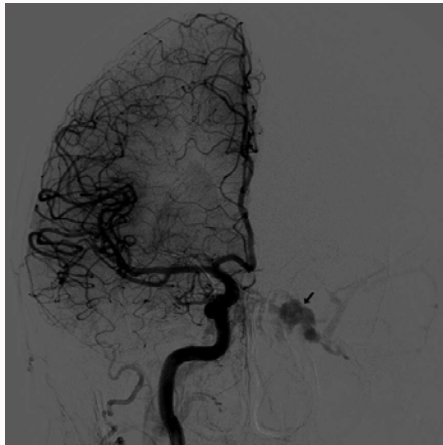


Figure 2a: Right internal carotid angiogram showing an indirect Barrow type D left carotid cavernous fistula. Early filling of the left cavernous sinus (arrow) fed by branches from the right internal carotid (through meningo-hypophyseal trunk).

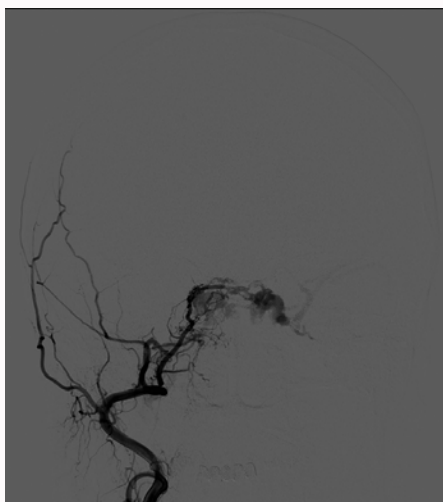


Figure 2b: Right external carotid angiogram, lateral view, shows the CCF feeding by internal maxillary artery.



Figure 3: External photograph (front view) showing a clinical worsening with proptosis, ocular redness, conjunctival chemosis occurring 24 hours after embolization of left CCF.

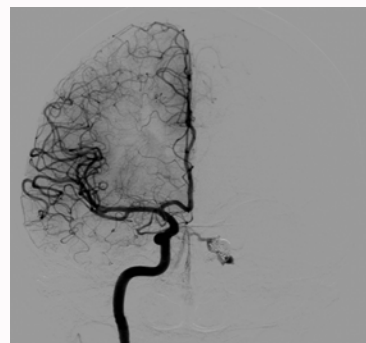


Figure 4: Right internal carotid angiogram (front view) shows little and early filling of the left cavernous sinus through a small meningo-hypophyseal trunk.

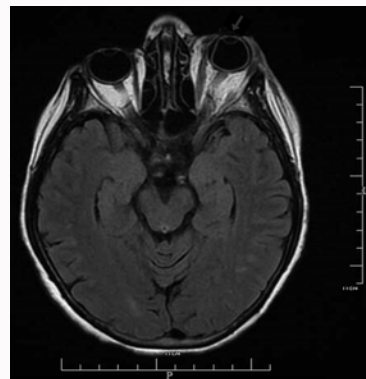


Figure 5: Axial Flair MRI image showing the choroidal detachment along medial wall of the left ocular globe extending up to the ciliary body (arrow).

almost complete closure of the fistula and with no complications. Twenty four hours later she complained of moderate to severe retro orbital pain, loss of vision in the left eye, nausea and vomiting. At that moment, the ocular examination revealed amydratic nonreactive left pupil, vision reduced to only light perception, limitation of horizontal ocular motility, conjunctival chemosis, severe proptosis and an intraocular pressure of 33 mmHg (Figure 3). Gonioscopic findings demonstrated a closed angle in the OS with a normal OD anterior chamber angle. A new cerebral DSA was immediately

performed showing little and early filling of the left sinus cavernous through a small meningo-hypophyseal trunk with no flow in the SOV, suggesting a possible thrombosis at that point (Figure 4). Given that these findings did not explain all the symptoms (such as severe loss of vision), a cranial MRI was carried out which revealed a left thrombosed SOV as well as a choroidal detachment along the entire medial wall of the left ocular globe, extending up to the ciliary body and provoking its anterior displacement and consequently an acute angle closure glaucoma (Figure 5). Topical instillation of 0.5% timolol maleatedrops, 750 mg of oral acetazolamide per day and

left canthotomy-cantholysis were necessary to control intraocular pressure, which lowered to 17 mmHg the following day. At a three month follow-up visit, the patient had improved significantly with no ocular symptoms and her visual acuity was 20/25 OS. A new DSA showed a complete closure of the CCF.

Discussion

Endovascular procedures involving the use of different embolic materials are one of the main therapeutic interventions for CCFs, with success rates between 70-78% for closing indirect carotid cavernous fistulas [2]. Different complications can be present up to 5% of the procedures, being iatrogenic choroidal detachment and subsequently acute angle closure glaucoma (AACG) an extremely rare complication and no more than a few cases have been described in the literature. The majority of them developed several hours after the embolization (as in our patient) however, AACG can also occur after angiography and endovascular manipulation without embolization and may even develop several weeks after the intervention [3-5]. The pathophysiological hypothesis is unclear but three different mechanisms have been proposed: 1) the hemodynamic changes produced by the CFF itself (venous stasis and orbital congestion); 2) the SOV thrombosis due to the embolization and 3) the absence of sufficient collateral drainage. All together could lead to increased pressure and permeability in the vortex veins, transudation into the supra choroidal space (normally virtual) and a choroidal separation. Subsequently, the detachment produced a forward shift of the iris-lens diaphragm, closing of the anterior chamber with failure of the aqueous humor circulation and finally, an AACG [4-6]. The worsening in our patient occurred over night, in the darkness, which favors pupil dilation, thus further narrowing the angle and increasing the propensity to develop an AACG. Neuro imaging techniques, such as brain MRI, can help us in the differential diagnosis showing the choroidal detachment as a semilunar or ring shaped area of increased signal on both T1 and T2 weighted images [7].

Considering that an early diagnosis and treatment are crucial to avoid deterioration of choroidal and retinal circulation, neuroophthalmologists should consider this entity in the differential diagnosis of a patient with vision loss after CCF embolization. Medical management, canthotomy-cantholysis, iridotomy, choroidal drainage are useful treatments to decrease intraocular pressure if the CFF has been previously closed.

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