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Unilateral Blindness as the Only Finding in a Patient with High Flow Carotid-Cavernous Fistula

The authors reported a patient with unilateral blindness of the left eye after trauma. Apart from paralysis of the 4th cranial nerve and mild optic nerve edema, there was no structural problem during the ophthalmologic examination. Brain CT scan, CT angiography and MRI demonstrated aneurysm in the left pericavernous region. The patient underwent angiography for coiling of the aneurysm, which detected large carotid cavernous fistula (CCF) mainly draining into the petrous sinus. Therapeutic embolization was performed with balloon and the CCF vanished completely. The most important points of this case are the blindness without exophthalmia as the only presentation of CCF which has not been reported as the only symptom and failure of CT-angiography and MRI evaluation for the diagnosis of CCF.

Keywords: Carotid-Cavernous Sinus Fistula, Blindness, Embolization, Therapeutic

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Introduction

Carotid-cavernous fistula (CCF) is an abnormal communication between the arterial and venous system which occurs within the cavernous sinus and is categorized according to pathological, hemodynamic or angiographic criteria.[1,2] This rare condition mostly occurs after head trauma.[1,2]

In angiography, lesions that directly receive their flow from the internal carotid artery (ICA) (direct type of CCF) are high-flow, and those that receive their flow from dural branches of the internal or external carotid system (indirect type) are usually low-flow fistulae.[1] The size, duration, location, venous drainage pattern and collateral vascular anatomy are very important factors for the clinical manifestations.

Venous drainage from the anterior region into the ophthalmic veins or posterior into the petrosal sinuses causes different symptoms in patients.[1] In cases that fistula drainage takes place into the inferior petrosal sinus, the orbital sign and symptoms are less severe.[3-6] The clinical manifestations are usually unilateral and ipsilateral and are often vague at the onset of occurrence.[7-9] Clinical manifestations are proptosis, exophthalmia, bruit, frontal headache and orbital pain, chemosis, extraocular palsy and diplopia, loss of visual acuity, 5th cranial nerve involvement and epistaxis.[10,11]

The 'gold standard' method for the diagnosis of CCF is cerebral angiography which is useful in identifying the fistula, evaluating venous drainage and assessing collateral circulation.[12,13] To our knowledge, there is no report of blindness without exophthalmia as the presentation of CCF in the literature or the failure of CT-angiography and MRI evaluation for the diagnosis of CCF. We report a very rare case of CCF that presented with unilateral blindness.

Case Presentation

An 11-year-old boy was admitted to our university-affiliated hospital for evaluation of blindness. He had no visual problem up to 4 years of age, when he suddenly

fell down and fractured his right forearm and lost consciousness for several hours. He was admitted and there was no serious problem in his evaluation. One month later, unilateral blindness occurred.

In spite of blindness, there were no retinal or structural problems (Fig. 1). Neurological examinations were normal except for mild left optic nerve edema and 4th nerve palsy. During one year, complete evaluation including ophthalmic examination, routine laboratory evaluation, brain CT scan, CT angiography and MRI evaluation of the patient was performed.

Ophthalmologic examination revealed no light perception (NLP) in the left eye, left optic nerve atrophy and left sixth nerve palsy. Routine laboratory evaluations were normal.

Brain CT scan demonstrated a round, well defined, low attenuated structure in the left parasellar region extending posteriorly over the petroclinoid region, which is suggestive of aneurysm (Fig. 2). CT angiography showed a deformed aneurysm measured approximately 15×20 mm, originated from the posterior aspect of the cavernosal part of the left internal carotid artery (ICA) (Fig. 3). MRI evaluation on T1, T2 and FLAIR images indicated round signal void in the left parasellar area suggesting aneurysm of the internal carotid artery (Fig. 4).

The patient was referred to the interventional radiology department for angiography and coiling of aneurysm. Angiography was performed by using angiography system GE innova 4100 with 3D capability. The complete cerebral angiography revealed a carotid cavernous fistula with aneurysmal dilatation of the C4 segment of ICA, venous drainage mainly into superior and inferior petrosal sinuses, and faint drainage toward the left ophthalmic vein (Fig. 5).

Right carotid angiography showed visualization of left hemisphere vessels via the anterior communicating artery by cross compression test.

Considering different diagnosis, interventional treatment and balloon occlusion for CCF was considered. For balloon insertion, guiding catheter (Cordis Corp NJ USA) was placed into the distal cervical segment of the left internal carotid artery. Then microcatheter (magic) advanced with detachable

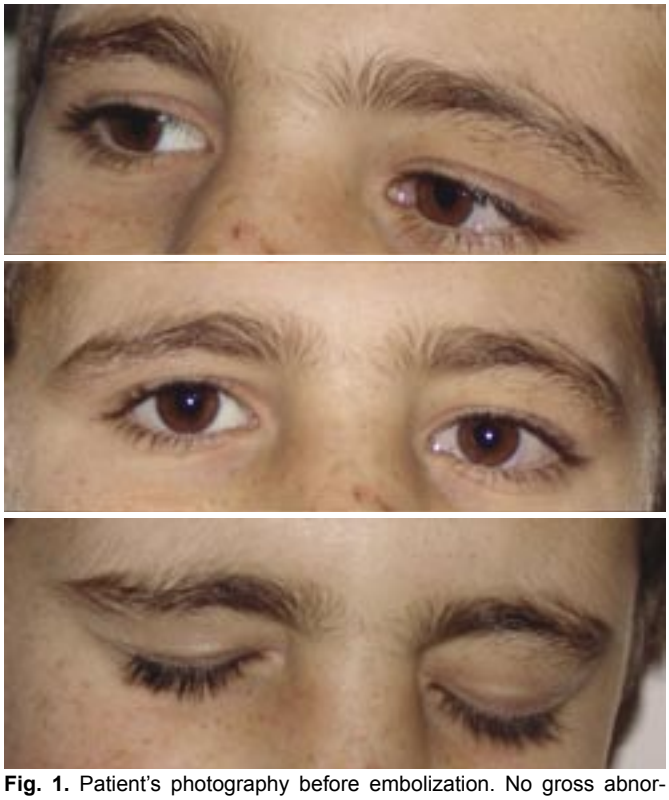

Fig. 1. Patient's photography before embolization. No gross abnormality is detected.

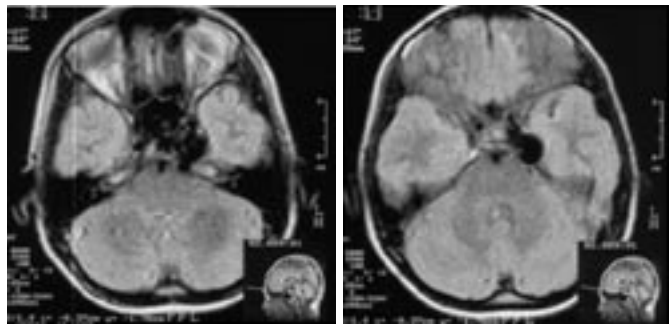

Fig. 2. CT scan reveals a round low-attenuated lesion in the left cavernous sinus compatible with aneurysm.

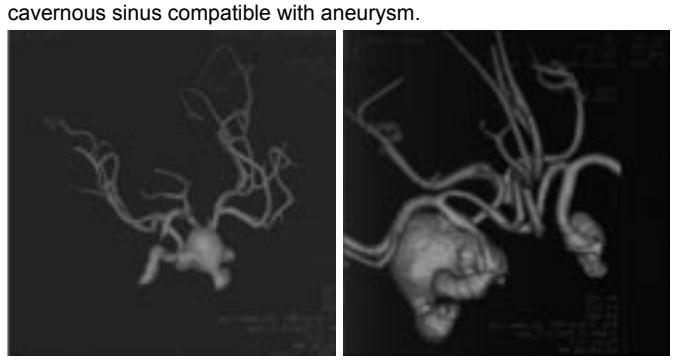

Fig. 3. CT angiography with 3D VRT images revealing giant aneu-rysm in the cavernous portion of the left internal carotid artery.

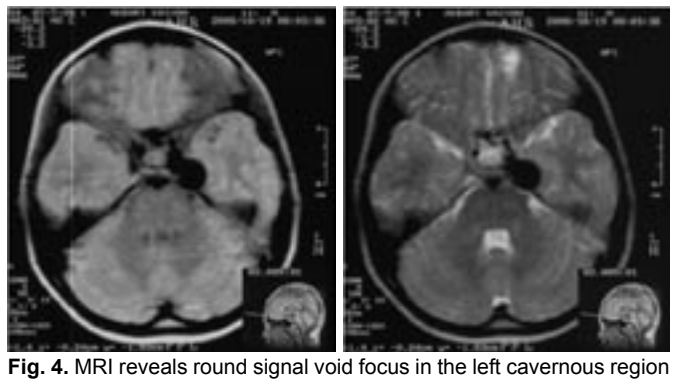

Fig. 4. MRI reveals round signal void focus in the left cavernous region compatible with aneurysm

balloon. We treated the patient with balloon which released at the site of fistula. The balloon was inserted at the site of fistula's opening with L shape manner, and control angiogram showed complete vanishing of CCF (Fig. 6).

Control angiography from the right carotid showed well visualization of both hemisphere vasculatures. There was no complication at the time of angiography. In the follow-up study, there were no changes in visual acuity.

Discussion

Carotid-cavernous fistula (CCF) which is a pathologic shunt between the carotid artery and cavernous sinus may be the cause of visual loss, and is mainly classified as direct and dural types. Direct CCF is a shunt between the internal carotid artery and the cavernous sinus[14-17] and this type of CCF is mostly traumatic. Symptoms of CCFs are mostly ocular and their severity can vary widely. The presentations are usually unilateral and ipsilateral, but may be bilateral or contralateral due to the communication between the two cavernous sinuses. The symptoms are often vague in onset and then progress when the fistula arises. Our patient showed two interesting points in comparison with other similar cases; first the bizarre presentation and second the misleading imaging.

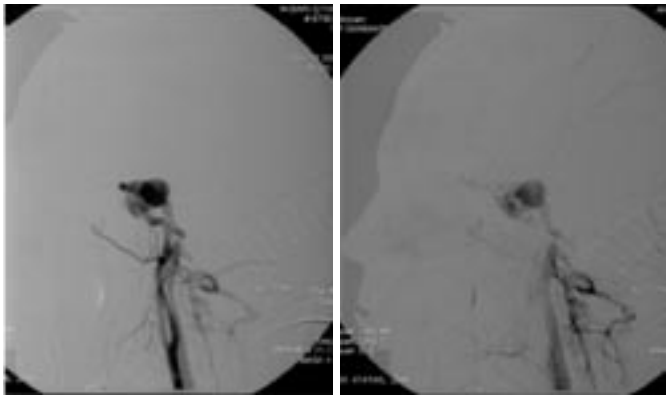

Fig. 5. Angiography shows aneurysmal dilatation of the C4 segment of the left internal carotid artery and fistulous connection to the cavernous sinus and venous drainage to petrosal sinuses and also to the superior ophthalmic vein.

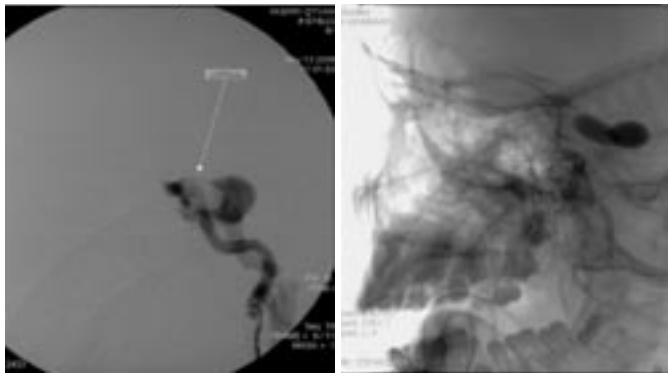

Fig. 6. Balloon insertion at the CCF neck to occlude the fistula opening. The arrow points to the site of CCF closure with balloon and the completely occluded flow to the venous sinuses.

The most common symptoms and signs of direct CCF include pulsatile tinnitus, periorbital bruit, proptosis and chemosis.[18-22] The causes of visual loss as Das et al. mentioned include "reverse venous flow from the fistula, arterial flow into the superior ophthalmic vein, increased intraocular venous pressure, venous stasis retinopathy and ischemic optic neuropathy".[2] But our patient presented with gradually developing visual loss and headache, without other remarkable orbital findings. The cause of such unusual presentation is related to cavernous sinus anatomy. Cavernous sinus is a network of anatomically separated sinusoids instead of an actual vein.[23] When a fistula develops between the ICA and the cavernous sinus, the high flow and pressure within the venous drainage pathways increase and flow reversal occurs in the venous tributaries to the cavernous sinus. The clinical signs and symptoms are directly related to the site of venous drainage.

The reversed and increased flow in anterior drainage of cavernous sinus into superior and inferior ophthalmic veins can lead to ocular symptoms including exophthalmia, chemosis and decreased visual acuity. However, our patient did not have exophthalmia or scleral injection due to main drainage toward the petrosal sinuses and no remarkable drainage into the ophthalmic veins.

Visual deterioration results from a combination of reduced arterial perfusion and venous hypertension accompanied by glaucoma. The decrease in retinal perfusion and the rise in intraocular pressure may result in optic atrophy as it happened to our patient.

Bizarre symptoms and signs in this patient mislead the clinicians and masked the high flow CCF till high venous pressure induced optic nerve atrophy. In spite of the dedicated value of CTA and MRI in the vascular lesions in this case, failure of both techniques in the detection of CCF is remarkable. We treated the patient mainly to protect the right eye, to reduce venous pressure in the right cavernous sinus and to prevent any possible similar event for the contralateral eye and other complications that may later affect the patient.

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