

ORBITAL ARTERIOVENOUS FISTULA WITH SYMPTOMS CONTRALATERAL TO THE ARTERIAL SUPPLY. A CASE REPORT

FRANKEFORT N., MD*, SALU P., MD*,
VAN TUSSENBROEK F., MD**

ABSTRACT

Carotid-cavernous sinus fistulas presenting signs and symptoms contralateral to the arterial supply of the fistulas are not uncommon. We describe a thoroughly documented case of an orbital arteriovenous fistula with symptoms exclusively contralateral to the arterial source, a rarer entity. A carotid angiography performed on a patient who presented a red chemotic and proptotic left eye showed a shunt between the meningeal branches of the right internal carotid artery and a left orbital vein.

RÉSUMÉ

Les fistules du sinus caverneux de la carotide avec des signes et symptômes contralatéraux de la source artérielle ne sont pas infrequentes. Nous présentons un cas bien documenté d'une fistule orbitaire artério-veineuse avec exclusivement des signes contralatéraux à la source artérielle, une entité plus rare. Une angiographie de la carotide nous montre une fistule entre les branches meningeées de l'artère carotide interne droite et une veine orbitaire gauche, chez une patiente présentant un œil gauche rouge avec un chémosis et une proptose.

KEY WORDS

Orbital arteriovenous fistula, carotid angiography, contralateral symptoms

MOTS-CLÉS

Fistule orbitaire artérioveineuse, angiographie de la carotide, symptômes contralatéraux

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* Department of Ophthalmology, Academic Hospital Free University of Brussels, Brussels, Belgium.

** Department of Radiology, Academic Hospital Free University of Brussels, Brussels, Belgium.

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INTRODUCTION

Arteriovenous fistulas are characterized by abnormal shunting of blood circulation between the arterial and the venous system. They represent a group of disorders that are characterized by abnormal shunting from the external carotid artery, internal carotid artery, vertebralbasilar system, or spinal arteries into normal or abnormal venous channels. Peeters and Kröger (3) and De Keizer (1) classified different types of carotid-cavernous (CCF) and orbital arteriovenous fistulas. Fistulas with ocular features can also be differentiated as spontaneous or traumatic, or as low- and high-flow fistulas. The calibre of the artery involved will dictate the type of lesion, as well as the signs and subsequent treatment.

Orbital arteriovenous fistulas are low-flow fistulas that are located only in the orbit itself but which induce the same ocular features as do dural and direct CCF (1). Pathological connections can be found between the branches of the ophthalmic artery and anterior cerebral fossa veins as well as between the recurrent meningeal artery and superior or inferior ophthalmic veins

(4). These arteriovenous shunts are quite rare, and most are a part of more extensive intracranial or facial arteriovenous malformations (2). Orbital arteriovenous fistulas, dural and direct CCF can present the same clinical features with orbital signs to the homolateral as well as at the contralateral side. This unusual manifestation of findings solely contralateral to the arterial source has been reported for direct CCF (1) and less common for dural CCF (6), but in case of orbital arteriovenous fistulas less cases have been presented. We report the clinical and radiological findings in a patient with an orbital arteriovenous fistula fed by a right artery, which presented exclusively left-sided signs and symptoms.

CASE REPORT

An 84-year old woman was sent by her family doctor to our out-patient department of ophthalmology for a lasting and increasing redness of the left eye since three weeks, in spite of treatment with local antibiotics. The patient couldn't tell us when the symptoms at her left eye began and what she was really feeling be-



Fig 1. Photography of the patient on the day of her first examination, showing a chemotic and proptotic left eye.

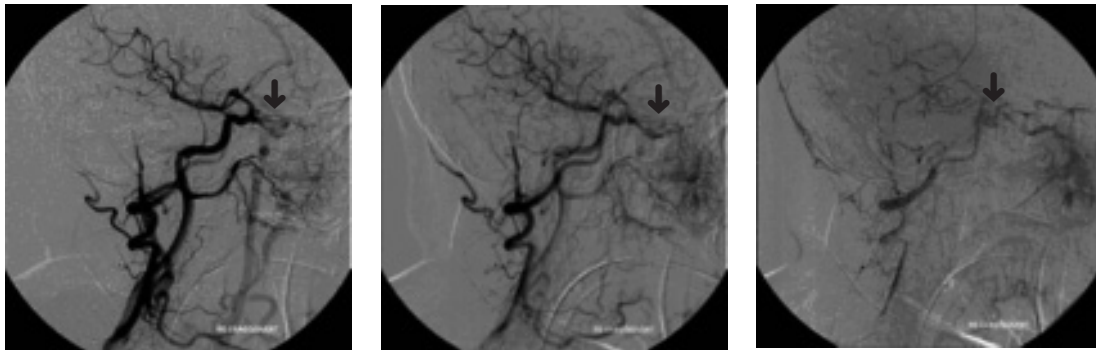


Fig 2, 3 and 4. Carotid angiography: Selective injection of the right carotid artery showing the shunt between the meningeal branches of the right internal carotid artery and a left orbital vein (arrows).

cause of an Alzheimer disease. At the clinical examination we observed a very red chemotic and proptotic left eye, and a normal right eye (*figure 1*). Her best corrected visual acuity was 3/10 in the right eye and 2/10 in the left eye; she had no relative afferent pupillary defect, and there was an important restriction of the eye motility left. Left exophthalmos was present (Hertel exophthalmometer measured 16mm on the right and 24mm on the left eye). Slit lamp examination showed markedly dilated and tortuous conjunctival vessels in the left eye with clear corneas, normal anterior chambers and irides, and cataracta incipiens bilaterally. Intraocular pressure on the left was 40 mmHg with applanation tonometry. Fundoscopy revealed dilated and tortuous retinal veins at the left eye, with normal optic nerves, maculas and periphery bilaterally. No associated ocular bruit was found.

A **computerized tomography (CT) scan** of the brain and left orbita showed an important unilateral left hypertrophy of the ocular muscles with exophthalmos. That explains the restriction of ocular motility.

Because of suspicion of an acute myositis or a Graves' ophthalmopathy an endocrinological checkup was performed and a treatment with corticoids was started. We associated 500 mg intravenous acetazolamide and local timolol 0.50% bid in order to decrease the intraocular pressure.

After 2 days of treatment, there was a normalisation of the intraocular pressure and a decrease of the conjunctival chemosis. Because of the negative endocrinological results we decided to perform a bilateral carotid angiogra-

phy in order to exclude an arteriovenous fistula. The **carotid angiography** performed on the left side showed a delayed venous drainage of the cerebral sinus with thrombosis of the left internal jugular vein and collateralisation via the occipito-vertebral veins; carotid angiography performed on the right side showed a shunt between meningeal branches of the right internal carotid artery and a left orbital vein with a slow flow, thus a contralateral connection with fistula formation (*figures 2, 3, 4*). **Treatment** by way of endovascular techniques was impossible, because of the too small calibre of the vessels. A conservative treatment was chosen.

The patient left the hospital with a visual acuity of 5/10 at the left eye, normalisation of the intraocular pressure, mild conjunctival redness and decreased exophthalmos (*figure 5*), but still dilation of the left retinal veins.

DISCUSSION

This patient presented clinical signs of chemosis, proptosis with dilated and tortuous conjunctival veins, ocular hypertension and exophthalmos of the left eye that may have represented an active Graves' ophthalmopathy, but the endocrinological investigation did not support such a diagnosis. A carotid angiography was performed for excluding an arteriovenous fistula. Some authors, as De Keizer (1) and Timothy et al. (6), accentuated in their reports the importance of injecting both carotid systems in the angiographic exploration of suspected fistulas. In our case the injection of the contralateral side of the signs revealed the presence of an orbital arteriovenous fistula.



Fog 5. After treatment: decreased conjunctival chemosis, injection and exophthalmos

The hemodynamic characteristics of arteriovenous fistulas in the orbit are comparable to that of orbital arteriovenous malformations or carotid-cavernous sinus fistulas. Increased venous pressure in the orbit and signs of orbital congestion, such as proptosis, dilation of conjunctival and retinal vessels, ocular hypertension, dilation of the superior ophthalmic vein and enlargement of the extraocular muscles can be found in all three conditions (2). Orbital fistulas are usually low-flow fistulas and typically occur spontaneously due to degeneration of vessels from hypertension, atherosclerosis and other vascular diseases. Blunt head injury can lead to shearing of intracavernous arteries, causing the development of a fistula. The origin of the fistula is unclear in this patient because of Alzheimer disease. There was no history of trauma and the orbital arteriovenous fistula could occur spontaneously.

Different cases are reported of direct CCF with signs predominantly or exclusively contralateral to the arterial source (5, 6). Théron et al.

theorized in their report that contralateral flow is the result of thrombosis of the ipsilateral cavernous sinus and/or its communicating venous channels (5). In our case there was a thrombosis of the left internal jugular vein who could cause the spontaneous orbital arteriovenous fistula and/or aggravate the clinical signs.

The management of low-flow fistulas like dural and orbital fistulas is a conservative treatment, rather than actively using vascular intervention (1). If visual impairment becomes progressive, then use of selective embolization and detachable balloon occlusion should be considered, who was impossible in our patient due to the too small calibre of the vessels. The clinical evolution was favourable as consequence of the steroid therapy as well as the treatment of glaucoma with topical beta-blockers and intravenous acetazolamide, who could be the cause of the possible successive closing of the fistula by a decrease of the carotid arterial pressure in the shunt caused by hemodynamic disorders such as hypotension (1).

CONCLUSION

We presented an unusual case of a spontaneous, or traumatic, orbital arteriovenous fistula with manifestations that were exclusively contralateral to the arterial supply.

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Correspondence and reprints:
FRANKEFORT Nathalie
Depart. of Ophthalmology
Free University of Brussels
Laarbeeklaan 101
B - 1090 BRUSSELS
Tel: 0032 (0)2 / 477.61.17
Fax: 0032 (0)2/ 477.61.41