DIRECT CAROTID-CAVERNOUS FISTULA: A CASE REPORT AND REVIEW OF THE LITERATURE

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ABSTRACT

A 21-year-old man presented with severe proptosis, chemosis, diplopia and an orbital bruit three weeks after a motor vehicle accident. The intraocular pressure was increased. The suspected diagnosis of a direct carotid-cavernous fistula (CCF) was confirmed by digital substraction arteriography. Placement of a covered stent in the internal carotid artery was performed with rapid resolution of the symptoms and normalization of the intraocular pressure. The epidemiology, pathogenesis, symptomatology, differential diagnosis, treatment and outcome of carotid-cavernous fistulas are discussed.

RÉSUMÉ

Un homme agé de 21 ans se présentait avec proptose, chémosis, diplopie et souffle de l'orbite trois semaines après un accident de moto. Le diagnostic d'une fistule carotidocaverneuse était confirmé par artériographie. Le patient a été traité avec succès par le placement d'un stent dans l' artère carotide. Après le traitement, les symptômes disparaissent. La tension de l' œil qui était trop haute redevient normale. Partant de ce cas, l' épidémiologie, la pathogénèse, les symptômes, le diagnostic différentiel, le traitement et les résultats des fistules carotido-caverneuses sont discutés.

SAMENVATTING

Een 21-jarige man presenteerde zich met een uitpuilend rood oog met dubbelzien en geruisen over de oogbol, drie weken na een moto-accident. De diagnose van een directe carotido-caverneuze fistel werd bevestigd met arteriografie. De patiënt werd succesvol behandeld door middel van een stent die in de arteria carotis werd geplaatst. De symptomen verdwenen na enige tijd. Ook de intra-oculaire druk, die aanvankelijk gestegen was, kreeg terug normale waarden. Op grond van deze gevalsbespreking worden de epidemiologie, de pathogenese, de manifestaties, de differentieel diagnose, de behandeling, het verloop en de uitkomst van de directe carotido-caverneuze fistels besproken.

KEY WORDS

Carotid-cavernous fistula - internal carotid artery - cavernous sinus - therapeutic embolization

MOTS-CLÉS

Fistule carotido-caverneuse - artère carotide interne - sinus caverneux - embolisation

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INTRODUCTION

A direct carotid-cavernous fistula (CCF) is a rare life-threatening disorder where a direct shunt arises between the internal carotid artery (ICA) and the cavernous sinus. (Fig. 1) We first describe a case of a traumatic direct CCF and the successful treatment with angiographic stenting. Then we will discuss the epidemiology, pathogenesis, clinical manifestations, differential diagnosis, treatment and outcome of this disorder.

CASE REPORT

A previously healthy 21-year old man had a motor vehicle accident with a fracture of the right maxillary sinus, which was left untreated. Three weeks later, he developed a painful red eye at the right side, swelling of the eyelids and photophobia. After two days, he experienced diplopia and he was referred to our outpatient ophthalmology clinic. The visual acuity was 10/10, also color vision with the Ishihara pseudo-isochromatic plates was normal. There was a striking right-sided periorbital edema and ptosis, a bruit was audible over the right orbit. Ocular motility testing revealed an abduction, elevation and depression deficit of the right eye. Pupillary reflexes were normal. Slitlamp biomicroscopy revealed tortuous dilated conjunctival blood vessels and Hertel exophthalmometry revealed a right proptosis of 4 mm. The intraocular pressure was increased to 21 mm Hg. Fundoscopy showed slightly dilated vessels. The diagnosis of a traumatic direct CCF was considered.

Fig. 1: Schematic representation of a direct CCF.

Magnetic resonance angiography showed an enlarged, hyper-intense right cavernous sinus (Fig. 2). Digital substraction angiography (DSA) showed a rupture of the cavernous segment of the ICA 5mm proximally of the ophthalmic artery and early retrograde opacification of the superior ophthalmic vein and inferior petrosal sinus (Fig. 3). Intracerebral perfusion and collateral circulation showed no anomalies.
The patient was transferred to the interventional radiology department of the Ghent University Hospital, where the CCF was occluded with a 20mm long, 5mm diameter self-expandable covered stent (Symbiot®; Boston Scientific), delivered transfemorally via a 8F guiding catheter (Lumax®, Cook). Already a few hours after the procedure, chemosis and proptosis had improved considerably. The patient was discharged 2 days later. At the outpatient clinic two months later, all symptoms had disappeared and the eye motility had returned to normal. The control DSA at six months showed a patent stent and occlusion of the CCF (Fig. 4).

Fig. 2: Magnetic resonance imaging shows a widened, hyperintens cavernous sinus (arrow). Reprinted with permission (67).

Fig. 3: DSA shows a direct fistula between the intracavernous segment of the right ICA (black arrow) and the cavernous sinus (long arrow) with early and retrograde opacification of the superior ophthalmic vein (white arrowhead) and inferior petrosal sinus (black arrowhead).

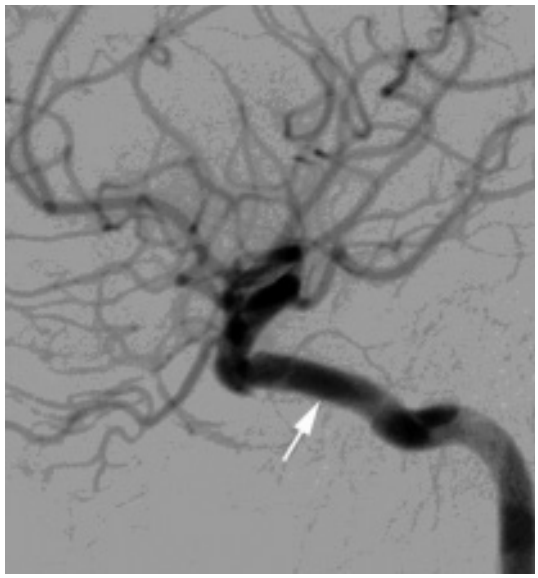

Fig. 4: Postoperative angiographic control: stented right ICA (arrow) with occlusion of the fistula.

DISCUSSION

Epidemiology and Pathogenesis

Abnormal communications between the cavernous sinus and the carotid artery can be classified by etiology (traumatic; spontaneous; iatrogenic), by flow dynamics (low flow; high flow) and by anatomy (direct versus dural; ICA versus external carotid artery (ECA) versus both). In direct CCFs there is a direct communication between the ICA and the cavernous sinus. An indirect CCF is defined as an arteriovenous fistula between the meningeal branches in the dura and the cavernous sinus. A clinically useful classification is the anatomical-angiographic classification by Barrow et al. in which fistulas are divided in four types. Type A fistulas are direct shunts between the ICA and the cavernous sinus (7) (Table 1).

The symptoms result from the abnormal communication between the carotid arterial and the venous cavernous structure, which causes a pressure gradient and consequently a flow through the fistula (Fig. 5). The pattern of venous drainage, either via anterior into the ophthalmic veins or posterior into the petrosal sinuses, dictates the clinical findings. In most cases there is a mixed anterior and posterior drainage. Anterior drainage through the valveless veins leads to the most dramatic symptoma-

Table 1: *The anatomical-angiographic classification of CCFs according to Barrow et al. (7)*

Category	*Definition*
Type A	Direct high-flow fistulas resulting from a tear between the ICA and the cavernous sinus; usually by trauma (A I) or by a ruptured aneurysm (A II)
Type B	Dural shunts between meningeal branches of the ICA and the cavernous sinus; spontaneous
Type C	Dural shunts between meningeal branches of the ECA and the cavernous sinus; spontaneous
Type D	Dural shunts between meningeal branches of both the ICA and the ECA and the cavernous sinus; spontaneous

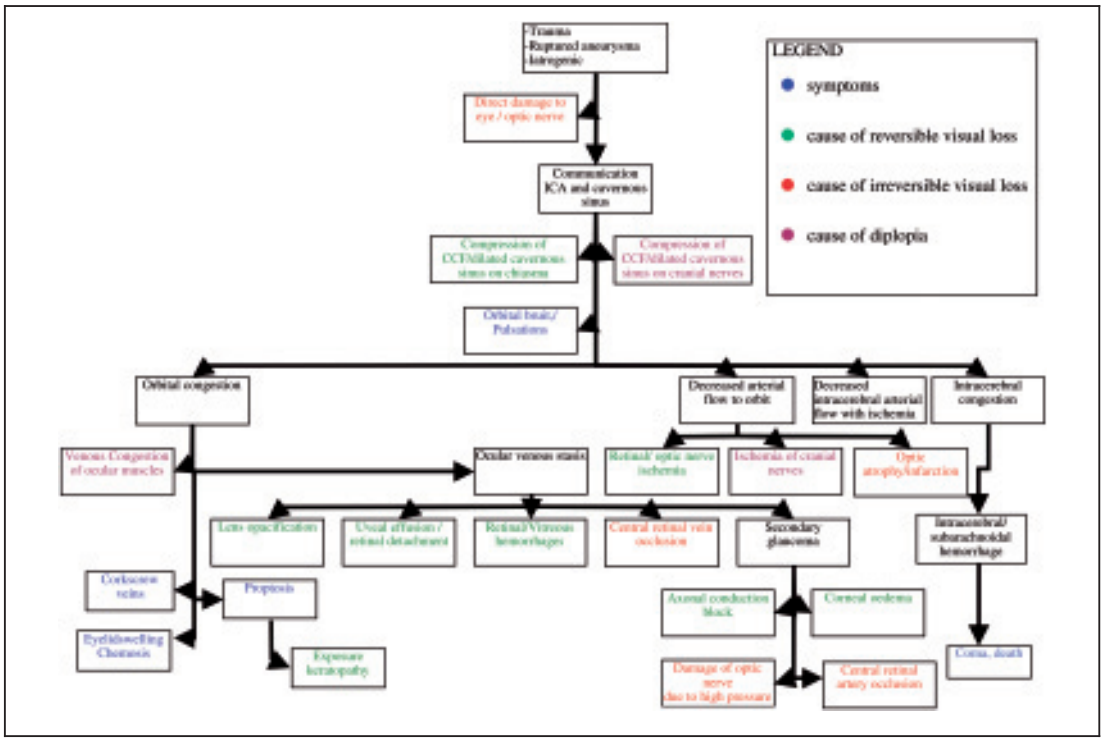

Fig. 5: Symptomatology of a CCF.

tology. The orbital manifestations are less severe when the fistula drains into the inferior petrosal sinus (11, 14, 45, 54).

Up to 76% of CCFs are traumatic. CCFs are seen in 0.2-0.3 % of craniofacial traumata (11, 45) (Table 2). The high percentage of traumatic CCFs can be explained by the anatomical relationship between the ICA, the cavernous sinus and the skull base. The cavernous sinus is unique because it is the only anatomic location in the body in which an artery is surrounded completely by a venous structure. Because the ICA is fixed to the surrounding dura of the skull base, it is exposed to shearing forces and penetrating injuries (22). A traumatic CCF occurs as the result of a laceration of the siphon of the ICA or one of its intracavernous branches, giving rise to a direct communication with the cavernous sinus. Most traumatic CCFs are of the high-pressure, high-flow type. Traumatic CCFs occur predominantly in young men (57%) because they suffer more frequently from head-traumata (45).

Spontaneous CCFs are less common, and most often caused by the rupture of intracavernous carotid aneurysms. They develop most often in middle-aged women (77%). About 2-9% of intra-cavernous carotid aneurysms are complicated by a direct CCF, whereas aneurysmal CCFs account for about 20% of direct CCFs (41, 45, 72).
A direct CCF may also be iatrogenic in origin. Development of a CCF has been described after transsphenoidal surgery for pituitary adenomas, treatment of trigeminus neuralgia and thrombectomy of the carotid artery.
Patiens with collagen diseases, such as Ehlers-Danlos Syndrome type 4 (EDS IV) (major abnormality in collagen type III) have a greater risk to develop a CCF because of the abnormal and fragile walls of their veins and arteries. Patients with atherosclerotic blood vessels also have a greater risk to develop CCFs (1, 11, 15, 20, 25, 34, 37, 45, 52, 62).
Indirect CCFs arise spontaneously by rupture of small dural arteries, and rarely by trauma. (35) These fistulas are of the low-pressure, low-flow

Table 2: *Causes of direct CCFs. Reprinted with permission (45).*

Cause	*Fistulas (n=100)*
Motor vehicle accident	54
Ruptured aneurysm	22
Fall	10
Sports injury	6
Gunshot wound to the head	5
Iatrogenic	2
Knife stab to orbit	1

type. This kind of CCF is often associated with atherosclerosis, arterial hypertension, diabetes mellitus and collagen diseases. They are more frequent in postmenopausal women and during childbirth (7, 11).

Symptomatology

CCFs mostly present with ocular symptoms, the severity can vary widely. This variation is not always correlated with the fistula gradient (22, 24). Because other more critical injuries often require clinical attention, a traumatic CCF frequently can be overlooked initially. The manifestations are usually unilateral and ipsilateral, but may be bilateral or even contralateral because of the connections between the two cavernous sinuses. Symptoms are often abrupt in onset and rapidly progressive as the fistula arises (10, 24, 30).

An orbital bruit is a common symptom (up to 80%), and can be heard by the patient and objectively determined by the physician. Subjective bruits are a result of bone conduction from posterior drainage (22, 46, 65). Exophthalmia is present in 72% of the patients but its extent is highly variable (from 3 to 16 mm) (11, 45). Pulsating exophthalmia has been described but is rare (19). Chemosis and arterialization of the episcleral veins are present in 55% of patients: epibulbar loops (corkscrew-like tortuous vessels). Eyelid edema with eventually exposure keratopathy may be present. (10, 19, 45)

Diplopia is present in 60-70% of the cases, and is caused by direct mechanical compression of the cranial nerves, a decreased arterial flow to the cranial nerves or congestion of the eye-muscles. Abducens palsy occurs in 49-85% of patients due to the free floating location of the 6th nerve within the cavernous sinus. The oculomotor and trochlear nerve are less commonly damaged because of their localization in the lateral wall of the cavernous sinus (11, 19, 45).

Visual impairment of some degree affects 60-90% of patients and is progressive if the fistula is not closed. Total visual loss has been described in as many as 25 % of patients but may be reversible. Reversible vision loss can result from retinal dysfunction due to ischemia produced by stagnant-flow anoxia, a mechanical, axonal conduction block as a result of intraocular pressure due to secondary glaucoma, compression of the optic nerve or chiasm by a distended cavernous sinus, opacification of the media by venous congestion and vitreous hemorrhage, uveal effusion, retinal detachment, lens opacification, corneal edema or corneal ulcers from proptosis-related exposure. Irreversible visual loss is due to optic neuropathy including optic nerve atrophy, infarction and trauma to the nerve sustained during the original head injury in patients with a traumatic carotid cavernous fistula, thrombosis of the ophthalmic veins with retinopathy and/or uveal effusion. It has been reported that a CCF may increase the risk of a central retinal artery occlusion due to the high intraocular pressure (2, 19, 43, 61, 66).

Retro-orbital pain and pain in the eye occur in 50-60% of patients with a CCF (45).

Raised pressure in the episcleral veins causes a rise in the intraocular pressure. This is present in 30-50% of cases with a CCF that are not treated. Acute angle-closure glaucoma has been described (19, 45). Massive epistaxis is a rare complication and is due to bleeding in the sphenoid sinus (31, 48).

Finally, an untreated CCF may become life-threatening. Death can result because of subarachnoidal or intracranial hemorrhage, venous infarctions and brainstem congestion. This is more frequent in patients with a complete hemodynamic steal phenomenon and a poor collateral circulation (22, 56, 73).

Diagnosis and differential diagnosis

The patient's history and the clinical examination often point to the diagnosis: a direct CCF is suspected in every patient with a painful red eye, chemosis , exophthalmia and orbital bruit, especially after a previous head trauma (26). The differential diagnosis should include posterior scleritis, endocrine exophthalmia, idiopathic inflammation of the orbit, retrobulbar bleeding, cavernous sinus thrombosis, malignancy of the orbit and arteriovenous malformations of the orbit/cavernous sinus (19, 74). In patients with CCF, fundoscopy may be normal, but often dilated retinal veins can be observed. Furthermore, ipsilateral optic disk swelling, intraretinal hemorrhages, vitreous hemorrhages and retinal detachment are associated with CCF. In the long run proliferative retinopathy and a cupped optic disk may develop (19, 36).

Intraocular tumors, scleritis and myositis or endocrine myopathy can be excluded with ultrasonography. In some cases, ultrasonography may demonstrate a dilatation of the superior ophthalmic vein (19).

Carotid and transcranial color Doppler imaging gives two-dimensional structural information and a hemodynamic assessment of the orbital vessels simultaneously. In patients with CCF, an irregular color mosaic flash in the markedly enlarged cavernous region may be discerned. The bruit is synchronous with the heart rhythm, and decreases with compression of the ipsilateral carotid artery. In addition, the orbital sonogram may demonstrate a reversed flow direction in the superior ophthalmic vein, sometimes called the 'ophthalmic steal phenomenon'. Color Doppler imaging should be used as prime investigation and for follow-up (19, 21, 23, 38, 77).

The diagnosis of CCF can be confirmed by more invasive diagnostics. Contrast-enhanced CT, CT-angiography and magnetic resonance angiography imaging show enlarged tortuous superior ophthalmic veins (the "hockey stick sign") in patients with CCF (76). Other radiographic findings with variable prevalence include lateral bulging of the cavernous sinus wall, edematous fat in the orbit and enlargement of extraocular muscles (3, 16, 32, 44, 67) (Fig. 2).

Jugular venous oxygen saturation monitoring is the management of patients with severe head injury. High values of the oxygen saturation may indicate a situation of arteriovenous shunting (12, 13, 17, 50).

Selective DSA of the carotid arteries remain the golden standard for the evaluation of the fistula prior to embolization. The exact localization of the lesion may be refined by injecting 2 to 3 mL/s of contrast into the ipsilateral intracavernous carotid artery, together with manual compression of the more proximal carotid artery in the neck, the so-called Mehringer-Hieshima manoeuvre. Furthermore, the Huber manoeuvre may help in identifying the upper extent of the fistula and can demonstrate double-hole traumatic fistulas and complete carotid artery transsection. This manoeuvre involves the injection of the ipsilateral vertebral artery, with lateral-projection DSA using manual compression of the affected carotid artery during the injection. The retrograde siphon filling of the cavernous sinus is then evident (3, 10, 16, 22, 32, 60, 67) (Fig. 3).

In patients with EDS IV, invasive angiography should be avoided except in extreme circumstances. The inherent vascular fragility makes the procedure hazardous. In patients with EDS IV, Doppler imaging and magnetic resonance arteriography are the procedures of choice for initial investigation (25, 37, 68).

Treatment

Treatment of a direct CCF is mandatory for immediate orbital symptom relief and to prevent the development of intracranial venous hypertension. Most direct CCFs can be treated electively, urgent treatment is necessary only in patients with progressive visual loss, corneal exposure, severe proptosis with pain and high intraocular pressure, intolerable bruit, epistaxis, sphenoid sinus aneurysm, severe retro-orbital pain, cortical venous drainage or coma (2, 22, 25). When the fistula is small and asymptomatic, further treatment is not indicated. The goal of treatment is the occlusion of the fistula while

maintaining carotid artery patency, which is mandatory when the collateral blood flow via the contralateral ICA is insufficient.

The treatment of choice for direct CCFs is transarterial embolization with detachable balloons (3, 5, 18, 27, 33, 34, 45, 46, 53-55). A latex or silicone balloon is mounted on a microcatheter and introduced in the cavernous carotid artery via the percutaneous approach of the femoral artery. Direct puncture of the carotid artery can also be used. The procedure is generally performed under general intubation analgesia. The high-flow shunt carries the device to the venous site of the fistula, where the balloon is then inflated and detached in situ.

A complete obliteration can be achieved in 80-90% of all direct CCFs. The carotid artery blood flow can be preserved in up to 75% of patients (14). The size of the cavernous sinus and of the fistula may affect the success rate of the procedure.

Prematurely deflation or dislocation of the balloon may lead to recurrence of the fistula or to the development of a pseudo-aneurysm. In a study of Debrun et al. 24 of 54 patients developed a pseudoaneurysm after transarterial embolization with detachable balloons (18). Recurrent CCFs or pseudoaneurysms can be treated with additional detachable balloons, coils (GDC or Guglielmi Detachable Coil) or, if the circle of Willis is patent, with occlusion of the ICA (39, 46, 48, 49, 69).

Complications of detachable balloon embolization of CCF are not uncommon: increased venous stasis, orbital congestion, cerebral ischemia (3%), cerebral infarction (4%) and permanent neurological damage (3%) have been reported. Furthermore, protrusion or shifting of the balloon into the lumen of the ICA may result in a significant stenosis, causing a transient ischemic episode or stroke in 6% of cases (45, 46). Up to 20% of patients experience transient oculomotor nerve palsy, possibly by direct pressure of the balloon on the cranial nerves (18, 27, 75).

The trans-arterial embolization procedure is unsuccessful in 5 to 10% of the cases (28). Failure occurs because it may be impossible to guide the balloon into the involved ICA segment due to intimal flaps, the fistula orifice may be too small to allow entry into the venous compartment, the fistulous communication may be too small to allow balloon inflation or sharp margins of bony fractures or foreign bodies may rupture the balloon during inflation. If the communication is large, it may be useful to inflate an additional balloon to stop the residual flow, but this is not always possible because the first balloon partially blocks the orifice (3, 28).

An alternative for balloon embolization are detachable platinum coils, either as a primary agent or in combination with balloons. A microcatheter is navigated into the cavernous sinus, where the coils are delivered to pack the cavernous sinus at the fistula site. Advantages of detachable coils are the ability to control their placement and to easily retrieve, reposition, or exchange them if necessary. However, in some cases the anatomy of the involved compartment of the cavernous sinus may hinder efficient and correct packing of the coils, leaving a partially patent CCF. Re-do interventions are therefore common in coil embolization of CCF. In cases where a ruptured aneurysm of the cavernous carotid siphon is the etiology of the CCF, coiling of the aneurysm often effectively occludes the fistula. A major disadvantage of coiling is its high cost (6, 8, 34, 63, 70, 72, 75).

Liquid embolizing agents (e.g. N-butyl-2-cyanoacrylate, isobutyl-2-cyanoacrylate, ethyl vinyl alcohol copolymers) can not be used in high-flow CCFs, because of the potential risks for cerebral infarction by the uncontrolled escape of the polymers during its injection and deposition. Protective devices (balloons, coils and stents) could play a positive role in preventing migration of the liquid embolic agent into the patent artery. These techniques are however largely experimental (4, 57).

Only recently, covered stents suitable for delivery in the cavernous segment of the carotid siphon became available. Covered stents reconstruct the carotid siphon and do not exert mass effect on cranial nerves. However, covered stents may be technically difficult to deliver, especial-

ly in elderly patients with tortuous vessels. Moreover, covered stents require chronic anti-sludge therapy with acetylsalicyl acid. During and immediate post-procedural, anticoagulation is mandatory, which in a posttraumatic setting may be hazardous. (35, 64) Although the two years follow-up angiographic control in this patient (and in another two successful cases) proved stent patency, longterm results by other investigators are required to define the role of covered stents for CCF. Unfortunately, for unknown reasons, the production of the self-expandable covered stent used in this case has been ceased (Fig. 4).

If a transarterial approach is not feasible or safe, a transvenous access to the cavernous sinus is another therapeutic option. Usually, the cavernous sinus may be reached via the valveless inferior petrosal sinus from the jugular vein. In patients with drainage in the anterior direction, the inferior petrosal sinus approach may not be appropriate because of a lack of communication between the anterior and posterior compartments of the cavernous sinus. Then a direct cannulation of the superior ophthalmic vein should be performed. when access via the facial vein is not possible (9, 29, 40, 42, 51). Great care must be given to the exact localization of the shunt when using the approach via the inferior petrosal sinus because closure of the posterior segment of the cavernous sinus in the absence of fistula obliteration will result in a diversion of flow to the superior ophthalmic vein, thus increasing the intraorbital pressure. On the other hand, occluding the anterior drainage may increase cortical or petrosal venous drainage and cause cerebral or brain stem hypertension (56, 64, 71).

Very rarely, if both the transarterial and transvenous approach fail, pterional craniotomy with a Dolenc approach is a possibility, but this approach is technically difficult (11, 59, 76).

In patients with EDS IV, an conservative approach should be preferred because of the high risk of dissection or rupture of the carotid artery by the intra-arterial procedure. In addition, there is an increased risk of postoperative hemorrhagic complications. The intervention should focus on simple vessel ligation rather than reconstruction, therefore balloon embolization is usually the treatment of choice. A combination with sacrificing the ICA or surgically trapping may be necessary. Recently, some authors state that a transvenous approach with Guglielmi detachable coils could have a better outcome (15, 20, 25, 34, 37, 52, 62).

Finally, secondary glaucoma and ischemic retinopathy may require medical treatment and photocoagulation therapy, respectively. In situations where direct treatment of the fistula cannot be achieved, secondary treatment of the ocular manifestations has only marginal success. Medical management of elevated intraocular pressure in the presence of a CCF will only transiently relief symptoms (10, 11).

Outcome

After successful occlusion of the CCF, the orbital symptoms gradually disappear, and in the presence of elevated ocular pressure, resolution to normal intraocular pressure within 72 hours can be expected. In some cases intraocular pressure normalization needs several months. The orbital congestion resolves usually after several weeks, reconstitution of cranial nerve function may lag several weeks (19). Diploplia almost always improves but in some cases, operative shortening of the lateral rectus muscle may be necessary (10, 45).

When the CCF is left untreated, vision is eventually lost in 89% of patients. In 1981, before the use of intra-arterial procedures, Palestine et al. reported progressive visual loss in 28% of patients, even with carotid artery surgery. More recently, visual acuity was preserved or improved in 94% of cases after treatment with detachable balloons in patients with preoperative visual loss. There are only some case reports of reversal of blindness after endovascular treatment (2, 10, 46, 47, 58).

Large studies with a long follow-up time ($>$ 10y) have not been performed, but after occlusion of the CCF, most patients remain asymptomatic (46). Recurrence of the fistula is possible, Luo et al. report that the recurrence rate is highest in patients in which the initial treatment was delayed and with large

fistula. This may reflect the fact that detachable balloons may be unstable in these large dilated cavernous sinuses. There is no evidence that the outcome is influenced by the number of embolization attempts (22, 48). Although orbital ultrasonographic follow-up is frequently performed, the importance of long-term follow-up of patients with CCF is still unclear. An angiographic control is advocated early after the interventional procedure, to exclude the formation of a pseudoaneurysm or a recurrent CCF (19) (Fig. 4).

CONCLUSION

In conclusion, interventional neuroradiology offers several transarterial and transvenous endovascular techniques to embolize the different types of direct CCFs. Short-term clinical results are excellent when the diagnosis is made in an early stage and the patient is referred to a specialized centre. Although the experience with covered stents is still limited, this non-occlusive technique seems a promising alternative for embolization in specific cases.

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