Angiographic confirmation of the spontaneous resolution of two low-flow carotid-cavernous fistulae

Confirmación angiográfica de la resolución espontánea de dos fistulas carotidocavernosas de bajo flujo

Dear Editor,

We present two cases of low-flow carotid-cavernous fistulae with angiographic demonstration of spontaneous closure.

The first case is a 62-year-old woman with a history of type 1 diabetes mellitus and dyslipidemia, who was admitted to the emergency service for diplopia, eye pain and dizziness of 2 weeks duration. Exploration revealed paralysis of abduction with internal deviation of the eye, conjunctival hyperaemia, ocular hypertension (25 mmHg) and left eyelid palpebral oedema.

Due to suspicion of ischemic neuropathy of the VI cranial nerve, we performed a cranial magnetic resonance imaging (MRI), which showed an enlarged calibre of the left superior ophthalmic vein with no evidence of thrombosis. Contrast-enhanced intracranial MR angiography also revealed a slight early filling of the left cavernous sinus. Both findings were consistent with low-flow carotid-cavernous fistula. Cerebral angiography confirmed the diagnosis and also showed an input from the meningeal branch of the right meningohypophyseal trunk and drainage through the left ophthalmic vein (fig. 1).

Local measures with ocular occlusion were established and a new angiography was scheduled for 5 days later, objectifying spontaneous closure of the fistula. At discharge the patient presented a slight foreign body sensation, paresis of the external rectus with resolution of pain and of conjunctival injection.

The second case was a 67-year-old male with hypertension and congenital solitary kidney, who began with a sudden case of diplopia. This was treated with antiplatelet therapy for suspected ischemic sixth cranial nerve neuropathy. Given the persistence of the clinical signs and symptoms, we opted for a Botox injection to reduce diplopia. Three months later the patient began to present proptosis, complete ophthalmoplegia and pain in the left eye. The examination revealed a manifest loss of visual acuity in the left eye with exudative detachment of the retina. We performed a computerised tomography (CT) with contrast of the orbits, which showed exophthalmos with periorbital fat infiltration, enlargement of the rectus muscles and the superior ophthalmic vein. Suspecting endophthalmitis, we established broad-spectrum antibiotic therapy. A new MR revealed left proptosis with increased calibre of the superior ophthalmic vein associated with diffuse thickening of the extrinsic ocular muscles. The 3D-TOF MR angiography sequence without contrast showed abnormal hyperintensity in the posterior portion of the cavernous sinus. After contrast administration, it was possible to appreciate early filling of the cavernous sinus, and the 3D phase contrast (PC) sequence with flow-direction encoding gradient showed arterialisation or flow inversion in the superior ophthalmic vein. Carotid-cavernous fistula (fig. 2) was thus confirmed.

Cerebral angiography revealed the fistula, which depended on meningeal branches of both internal carotids and drained predominantly into the left posterior cavernous sinus and from there to the superior ophthalmic vein.

Topical ocular treatment and rest were established, and the patient was scheduled for a new angiography before endovascular treatment, which took place 9 days later with spontaneous resolution of the fistula. Clinical evolution was favourable and upon discharge he presented minimal proptosis and good vertical motility with mild paresis in abduction.

Carotid-cavernous fistula is defined as the subtype of dural fistula caused by a pathological union of arterioles and cavernous sinus on the walls of the dura mater. The cavernous sinus receives venous drainage from the superior and inferior ophthalmic veins and the superficial middle cerebral vein through the sphenoparietal sinus. The physiological drainage of the cavernous sinus is into the petrosal sinuses and pterygoid plexus. The most common aetiology is trauma in relation to skull base fracture, although spontaneous forms are not infrequent. In the latter group, there seem to be predisposing factors such as high blood pressure, intracranial surgery, Valsalva manoeuvres, sinusitis or even pregnancy.[1]

The clinical manifestations are variable and most symptoms stem from venous hypertension of the orbit by the flow inversion of the superior ophthalmic vein with symptoms of exophthalmos, chemosis and diplopia. The drainage of arterial blood in a posterior direction is associated with tinnitus and dysfunction of cranial nerves III, IV, V and VI. Drainage towards the sphenoparietal sinus and reflux to the cortical veins may cause neurological dysfunction and a risk of intracerebral haemorrhage. In the two cases presented, the clinical signs and symptoms were produced by anterior drainage with stasis of the superior ophthalmic vein and ocular symptoms, both with chemosis, proptosis, involvement of the sixth cranial nerve and pain. Our first case also presented increased ocular pressure. Vision is generally normal in only 11% while 25%present a significant reduction in visual acuity and another 25%reach the point of blindness. In rare cases, sudden death can occur due to massive haemorrhage.[11]

The differential diagnosis should be carried out considering thyroid ophthalmopathy, infections, inflammatory pseudotumour, retrobulbar haemorrhage and Tolosa-Hunt syndrome. With infraorbital masses, orbital ultrasound is useful for differential diagnosis, observing an "arterialised" centrifugal flow in the superior ophthalmic vein.[2]

CT and CT angiography show a thickening of the superior ophthalmic vein and may show asymmetry and early filling of the cavernous sinus. They also make it possible to identify a thickening of the extraocular muscles and an increase of the orbital fat density, which can sometimes mimic retrobulbar cellulitis with infectious myositis. Magnetic resonance (MR) can confirm the diagnosis through MR angiography sequences. Dynamic sequences with echo gradient after gadolinium injection can reveal a filling in the arterial phase of the superior ophthalmic vein and the homolateral cavernous sinus. The phase contrast sequence (3D-PC) with flow encoding can show the centrifugal flow of the superior

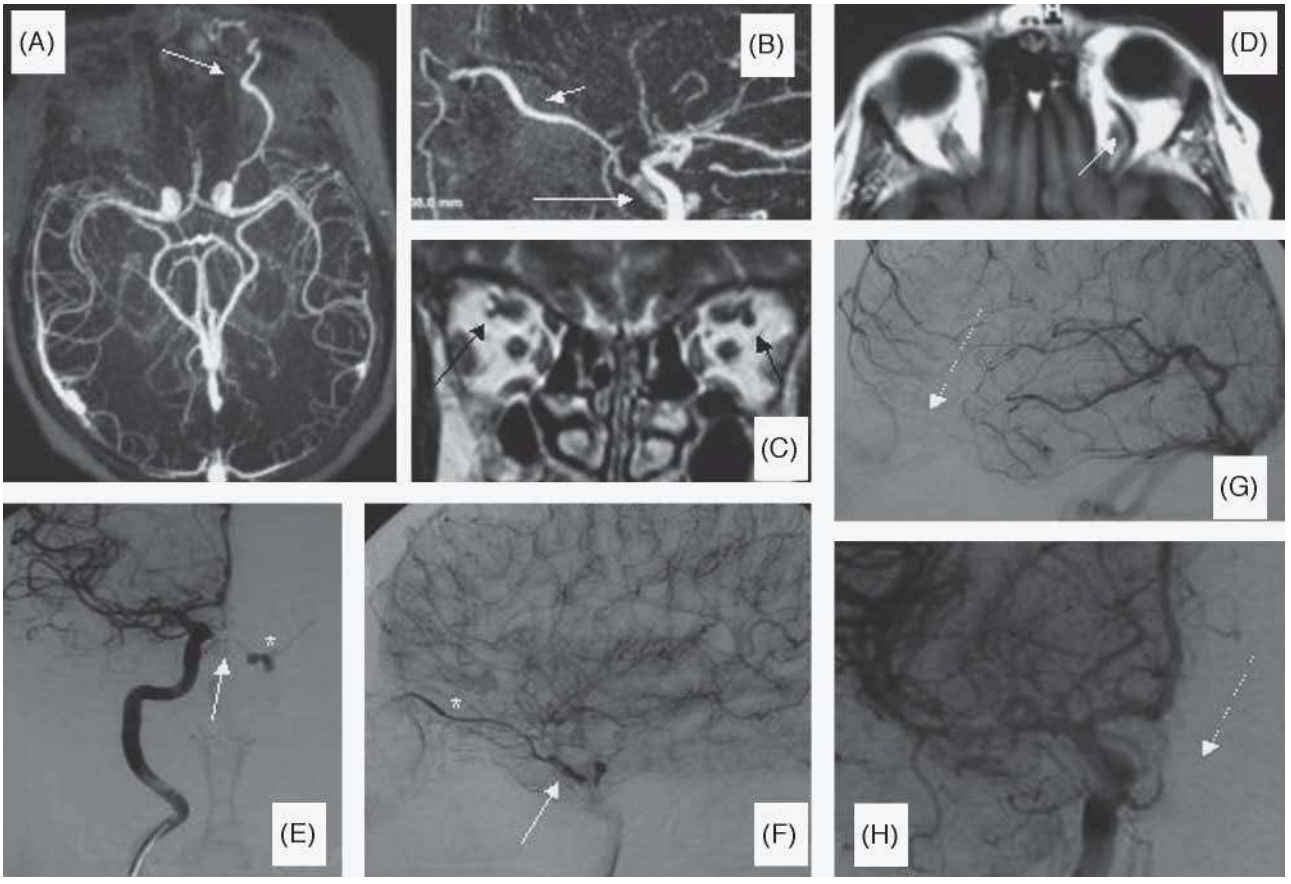

Figure 1 A: Contrast-enhanced magnetic resonance angiography, axial plane. B: sagittal plane. Early filling of the left cavernous sinus and the superior ophthalmic vein. C and D: T1-weighted sequences showing asymmetry and enlargement of the left superior ophthalmic vein. Images of the right carotid artery obtained by digital subtraction angiography. E: diagnostic image showing afference from the right meningohypophyseal trunk. F and G: lateral projection images of the drainage into the left superior ophthalmic vein. H: resolution of the clinical condition observed in E.

ophthalmic vein with the characteristic arterialisation of the process. In our cases, the suspected diagnosis was clarified by MR, which identified characteristic alterations in the cavernous sinus and superior ophthalmic vein.

An arteriography is required to classify and characterise fistula haemodynamics. The Barrow classification[3] divides fistulae according to their flow, putting them into four groups that go from Type A (or high-flow) to Type D (or low-flow). Direct or Type A fistulae have a direct trauma origin due to arterial fracture in 80%of cases and spontaneous in 20%; they generally correspond to a ruptured carotid aneurysm[4] and are more rarely associated with arteriovenous malformations, atherosclerotic lesions, arterial hypertension and connective tissue diseases. They affect young adults and have high flow and florid symptoms. These fistulae require embolic treatment, given that they very rarely close spontaneously.

Types B, C and D fistulae are called indirect or dural and most often affect perimenopausal women. They tend to be spontaneous from fistula of small calibre arterioles, with scarcely florid symptoms and may be spontaneously obliterated.

A new angiographic classification has recently been described, with a prognostic factor based on the type of arterial afferents and the location of venous efferents. This new classification divides fistulae into:

- Proliferative type: large number of arteriovenous shunts with relatively fast flow. The venous phase shows preserved anterograde flow into the superior petrosal sinus. The diffuse proliferative subtype envelops the entire sinus, while the posterior proliferative is located only in the posterior portion of the cavernous sinus.
- Restrictive type: minimal arterial afferents that can be delineated. The venous phase shows obliteration of anterograde flow in the inferior petrosal sinus and opening of the retrograde flow to the superior ophthalmic vein and/or cortical veins.

This classification relates the angiographic type to patient symptoms. Proliferative types, which introduce posterior permeable drainage, are often oligosymptomatic. Restrictive types, by presenting a cortical reflux and deteriorated venous drainage, are more often associated with ocular symptoms and cerebral haemorrhage.[5]

The two cases reported presented low-flow fistula with flow inversion of the superior ophthalmic vein and contrast stasis in

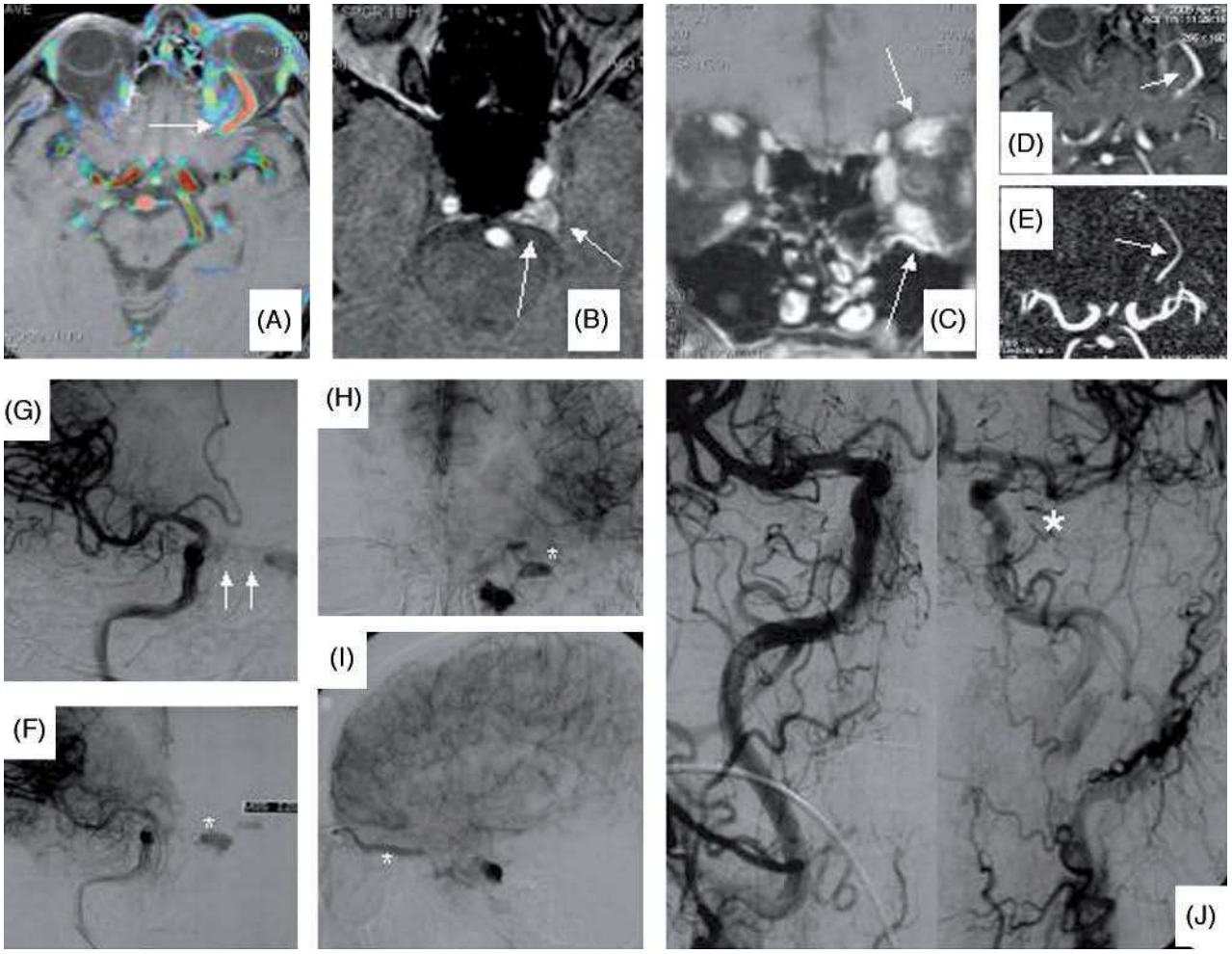

Figure 2 A: Phase contrast-enhanced (3D-PC) magnetic resonance angiography (MRA) sequence showing arterialisation of the left superior ophthalmic vein. B: MRA with and without contrast: abnormal hyperintensity of the posterior section of the cavernous sinus. C: fat-suppressed T1: increased calibre of the superior ophthalmic vein and diffuse thickening of the extrinsic ocular muscles. Angiography images with digital subtraction show afferences from the meningeal branches of both internal carotids. F and G: right internal carotid artery (ICA) in anteroposterior projection (AP). H: ICA in AP showing the primarily posterior affectation of the left cavernous sinus. I: the same ICA in lateral projection. J: angiographic composition showing the spontaneous resolution.

that vein. Using the second classification, the cases met the criteria for restrictive type fistula. Signs of poor prognosis include increased intracranial pressure, rapidly progressive proptosis, decreased visual acuity, neurological deterioration, haemorrhage and transient ischemic attacks. High-risk angiographic findings include venous drainage to cortical veins, thrombosis distal to the fistula, pseudoaneurysms and long cavernous sinus varix. None of these findings were observed in our cases.

The conservative treatment for carotid-cavernous fistulae is based on jugular-carotid and ophthalmic compression and is indicated in patients with mild disease. Faced with failure of conservative treatment or with signs of poor prognosis, the treatment of choice is endovascular therapy through the arteries, with fistulae embolized via coils or liquid embolic agents (preferred in type A fistulae); and intravenous treatment via the inferior petrosal sinus or facial vein, embolizing the cavernous sinus with coils.[6,7]

Reviewing the literature, we found a general agreement on the benign course and spontaneous resolution of a considerable percentage of carotid-cavernous fistulae. Although there are no statistical studies that relate benign evolution with angiographic characteristics, they are presumed to occur more frequently in indirect low-flow fistulae. The rates vary between 22%[8] and 33%,[9] but there are series that report up to 73%.[10] In our cases, the evolution was benign and the resolution, which was spontaneous, was confirmed by angiography.

We therefore believe that every patient with a diagnosis of low-flow carotid-cavernous fistula obtained via imaging techniques should be given an opportunity for spontaneous resolution.

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"Santiaguiño" sign in multisystem atrophy

Signo de santiaguiño en la atrofia multisistémica

Dear Editor,

Multisystem atrophy (MSA) is an adult sporadic neurodegenerative disease that presents as a variable combination of autonomic dysfunction symptoms, Parkinsonism, cerebellar ataxia and pyramidalism.[1] It currently falls within the category of synucleinopathies, since its anatomopathological markers are oligodendroglial alpha-synuclein inclusions.[2] Magnetic resonance imaging (MRI) studies of patients with MSA with a cerebellar origin show a hyperintense lesion in the shape of a cross, called the "*hot cross bun*" sign, at the centre of the pons. This sign reflects the degeneration of the median raphe and the

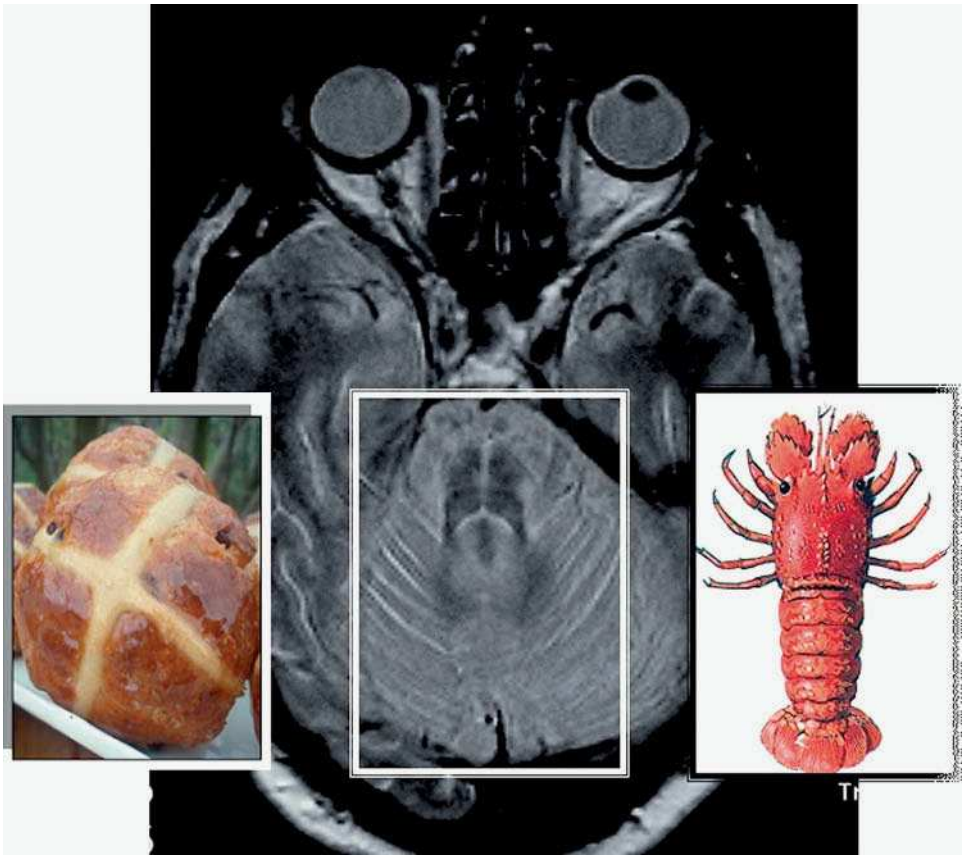

Figure 1 T2 axial MRI section: the pontine hypersignal resembles the *hot cross bun* sign (left), but when observed in association with the atrophy of the cerebellar vermis and hemisphere, it mimics the silhouette of a *Santiaguiño* (right).