Carotid-cavernous fistulas: from the pathophysiology and clinical symptoms to the radiologic findings.

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Learning objectives

First of all, to understand the anatomy as well as the pathophysiology related carotid cavernous fistulas and their symptoms.

To know the radiologic findings by vascular imaging studies (orbital Doppler, CT & MRI angiography and cerebral arteriography) about this vascular malformation.

To know a basic understanding about their endovascular treatment.
Background

INTRODUCTION. ANATOMICAL BASES.

Carotid-cavernous fistulas are anomalous communications between the arteries from the carotid system with the cavernous sinus. It constitutes, in this way, a pathology with an underlying structural alteration. For this reason it is essential to remember the anatomy of these structures as well as their anatomical relationships and vascular communications.

The cavernous sinuses are splits of the dura mater located on the medial slope of the middle cranial fossa, lateral to the sphenoid body. It is widely interrelated by connections to other veins and sinus within the skull. It previously communicates with the superior and inferior ophthalmic veins where the venous blood from the eyeball drains and also with the sphenoparietal sinus, where the middle superficial cerebral vein ends. Remember, ophthalmic veins communicate in front of orbit with frontal and angular veins. Behind, it communicates with the superior petrous sinus, which leads to the transverse sinus; and also with the inferior petrous sinus which continues with the sigmoid sinus. Eventually, it communicates medially with the contralateral cavernous sinus, through the intersigmoid or coronary sinus, and the basilar venous plexus too, above the clivus. (Fig 1)

As its own definition indicates, both external and internal carotid arteries through their branches may be involved in this pathology. The most important anatomically segment is the internal carotid artery intracavernous. This segment of the internal carotid artery (ICA) begins in the petrolingual ligament and ends in the proximal dural ring. The branches extending from this proximal to distal arterial segment are the meningohypophyseal trunk, inferolateral trunk, and McConnell's capsular artery. The meningohypophyseal trunk gives branches to the pituitary (inferior pituitary artery), the tentorium and the oculomotor nerves III and IV (tentorial artery) and the medial dura mater and V pair (dorsal meninges). The inferolateral trunk irrigates the inferior and lateral wall of the cavernous sinus, and the VI pair. Subsequently, it gives distal branches to the oval and spiny foramen, which are anastomosed with distal branches of the external carotid artery (emphasizing the middle meningeal artery). Finally, although inconstant, we find McConnell's capsular artery, which gives branches that perfuse the adenohypophysis and the medial dura mater (capsular and inferior pituitary arteries) and also the supero-medial dura mater (anterior capsular artery). It should not be forgotten that cavernous sinus also contains the following nerves: the closest to the carotid is the VI (abducens), superior and laterally the III (oculomotor) and inferiorly to it, IV (trochlear). Already more distant from the carotid artery is V1 (ophthalmic), inferolaterally, and inferior to the V1, appears V2 (maxillary). (Fig 2)
The carotid autonomic plexus is placed in close contact around the walls of the intracavernous carotid artery and contains the sympathetic nervous system. (Neurons afferent to the ciliary and pterygopalatine ganglia).

CLASSIFICATION

Once the anatomy is assimilated, carotid-cavernous fistulas (CCF) classification can be understood based on its pathophysiology. The most commonly used is Barrow’s classification, which subdivides them into 4 letters (A, B, C, and D) according to the arterial branch that communicates with cavernous sinus. (Fig 3)

TYPE A:
It is the CCF that is produced by a direct communication between cavernous internal carotid artery and cavernous sinus because of a solution of continuity of the arterial wall. It is the most frequent type, and its main cause is cranioencephalic trauma (approximately 80%). This is due to the cavernous internal carotid artery is strongly fixed by the dura when it enters and leaves the cavernous canal, immediately before the previous clinoids. Shear forces and traumatic shock waves tear the cavernous carotid artery, often at a single point of considerable size (2-5 mm), resulting in a high-flow fistula with massive arterialization of the cavernous sinus. The tear point is usually located in the proximal cavernous segment, adjective to the vicinity of the meniphero-physeal trunk (remember, the first intracavernous branch).
Occasionally arteriography has seen another adjacent tear, and rarely (approximately 5% cases), a massive tear in this arterial segment, with a complete theft and absence of vascularization of the distal branches of the cavernous internal carotid artery. Bilateral posttraumatic CCF, which is usually in the context of severe cranioencephalic trauma, is also possible, although rare (approximately 2%).
Other causes of this type of CCF are atraumatic (approximately 20%). In this sense, there are spontaneous CCFs, such as the rupture of an aneurysm in the intracavernous ICA or posttreatment, either surgical (transsphenoidal open surgery) or endovascular (for example, angioplasty of ICA in the siphon). There are rare diseases that predispose to a spontaneous atraumatic type A CCF that increase the weakness of arterial walls such as fibromuscular dysplasia, Ehler-Danlos syndrome or elastic pseudoxanthoma. Likewise, they will also predispose to situations that increase blood pressure (such as occlusion of the contralateral carotid artery)

TYPE B

It is the CCF that is produced by an indirect communication between the branches of the internal carotid artery that supply the cavernous sinus. These branches have been mentioned previously (dorsal meningeal branch, inferolateral trunk and the capsular artery).
TYPE C

It is the CCF that is produced by an indirect communication between the dural branches of the external carotid artery that supply the cavernous sinus. Among them, it is worth mentioning the caliber of the middle meningeal artery that crosses the spiny hole. The accessory meningeal artery through the foramen ovale, and the ascending pharynx artery may also be involved.

TYPE D

It is the CCF that is produced by an indirect communication between the dural branches of both carotid arteries that supply cavernous sinus. It has been subclassified in D1 if it is a FCC with unilateral supply or D2 if it has bilateral supply.

FCC type B, C, and D are low-flow and are also known as dural arteriovenous fistulas. Its cause is not yet well established. Some are congenital and others acquired. Among the acquired are more frequent in postmenopausal women. Factors that have been associated with this type of fistulae are venous sinus thrombosis, hypertension, diabetes, atherosclerosis, collagen diseases, pregnancy, otitis and sinusitis. So are surgery and trauma.

PHYSIOPATHOLOGY AND CLINIC

The formation of arteriovenous communication causes **increased venous pressure** and inversion of the flow in the superior ophthalmic vein and in the inferior ophthalmic vein with the consequent ingurgitation of the latter. This fact produces typical clinical manifestations: chemosis, proptosis, pulsatile noise and increased intraocular pressure (promoted by the difficulty of draining the aqueous humor through the Schlemm's canal). **Pulsatile noise**, synchronized with systole, occurs in 50% of patients and is due to the entry of turbulent arterial flow in the dural sinus. Also on rare occasions in the long term, there is a decrease in visual acuity because of difficulty in retinal perfusion by increasing venous and intraocular pressure.

These clinics may manifest in the contralateral eye by a single unilateral CCF, without necessarily having a bilateral CCF, due to the communication between the two cavernous sinuses through the circular sinus. Another factor promoting this fact is thrombosis of the ipsilateral ophthalmic vein.

On the other hand, the arterio-venous flow will not always tend to go to the ophthalmic veins, it depends on the permeability of the drainage routes of the cavernous sinus and also on the site of the fistula. Thus, since the pathophysiological mechanism is the increase of the venous pressure, we can find other clinical manifestations less frequent depending on the route of drainage of the anomalous flow:
- If there is a posterior drainage to the petrous sinuses, there will be tinnitus (generated by the actual sound of the turbulent flow in the vicinity of the ipsilateral ear). He may also present ophthalmoplegia due to involvement of oculomotor pairs, as well as V1 or V2 neuralgia. If mydriasis or palpebral ptosis exists, the III pair is likely to be affected.

- If there is a drainage anterior, but towards the sphenoparietal sinus, the increase of venous pressure will be transmitted towards the superficial middle cerebral vein. This situation is the most dangerous, since it implies neurological dysfunction of the involved cortex and risk of subarachnoid hemorrhage or subdural hematoma.
**Fig. 1:** Cavernous sinus. Venous drainages.


**Fig. 2:** Cavernous sinus. Internal anatomy and relationships.

Fig. 3: Barrow's CCFs classification.

Findings and procedure details

RADIODIAGNOSIS:

We are going to detail imaging CCF signs from five most representative cases, diagnosed and treated, of our hospital that were retrospectively collected since 2009.

The gold standard to diagnose this pathology is cerebral arteriography. However, given its disadvantages as an invasive test, we can use other imaging tools that show suspicious signs.

**Eyeball US doppler** will show inversion of the direction of flow in color Doppler as well as a low resistance arterial flow (arterialized flow) similar to the other arteriovenous fistulas in the spectral Doppler.

**Craniofacial CT non enhanced** provides important data regarding the cause of the CCF. If fracture traces are evident in the craniofacial skeleton in the context of a CCF-compatible clinic, the etiology will be traumatic (Fig 4). Other signs will appear as proptosis and more subtly increase the caliber of the superior ophthalmic veins (Fig 5). It is also important to consider that acute intracranial hemorrhages in this context may be cause to bleeding by leptomeningal venous ingurgitation associated with CCF, although this finding is not usually so early and it is more frequent to associate intracranial bleeding post trauma itself.

On **CT enhanced (CT angiography)**, an ipsilateral cavernous sinus enhancement in the early arterial phase will be identified, which will frequently be proportional in intensity to the CCF flow. if CCF is high flow Direct, type A, it is expected to observe a marked enhancement than if the fistula is of low flow (indirect, types B - D).

**Non enhanced MRI** will reveal increase the size of the oculomotor muscles with intraorbital and periorbital edema (Fig 6), changes that will appear hyperintense in T2-weighted sequences. Ophthalmic veins ingurgitation or other structures to which the fistula drains can be identified due to the vascular void phenomenon (Fig 7) will also be more accurately identified than CT non enhanced. A thrombosis in the ophthalmic veins may be identified as absence of signal vacuum in the affective venous segment.

**MRI enhanced** (with gadolinium), the hyperintensity of the affected cavernous sinus as well as the drainage routes may be identified more sensitively than with CT (Fig 8, Fig
9): ophthalmic veins or less frequently cortical veins and other dural venous sinuses that will associate an increase in the caliber of these structures.

Both TC and RM angiography may often not be able to identify the origin of a direct or non-direct CCF. In this sense all the above mentioned signs are indirect, that is to say, they are evident radiological manifestations produced by a CCF, but they are not images that show us the fistula as such. That is why if both are normal, do not rule out the diagnosis of this pathology.

For this reason **arteriography is the only tool that confirms the existence of CCFs.**

It must carry out with the presence of clinical manifestations that suggest it or to confirm the diagnosis and also to classify it, determining arterial feedings and venous drainage.

This procedure is used with caution in patients with renal failure, uncontrolled arterial hypertension or clinically unstable, considering the risk-benefit balance for each particular case.

Standard cerebral arteriography, which must be selective and bilateral of the main branches of the supraaortic trunks. Studies in the venous phase will reveal drainage routes from the cavernous sinus and explain the clinical manifestations of the patient, as detailed above.

It is important to take into account that all the necessary projections must be made for a correct evaluation of the arterial contributions and the venous drainage.

For studying CCF more common (Type A), it is well accepted the following steps:

- Posteroanterior and lateral projections of the ipsilateral ICA: they allow to identify the FCC, as well as the presence of theft in territory distal to the same. It also delineates the cavernous sinus and venous drainage (**Fig 10**).

- Lateral projection of ipsilateral ECA.

- Contralateral ICA study with compression of ipsilateral ICA. It is used to assess the permeability of the Willis polygon, especially the anterior communicating arteries. Having good collateral is essential if you want to sacrifice ipsilateral ICA.

- Study of ipsilateral vertebral artery (VA), better with ipsilateral ICA compression. This procedure allows a better evaluation of the vascular lumen of this artery as well as the collaterality of the posterior communicating arteries.
In case of difficulty to identify the location of the fistula, to use a balloon guide catheter, and inflate it in the different segments of the intracavernous ICA, followed by angiographies it is well established:

- Ball proximal to FCC: complete occlusion of ACI and cavernous sinus void of contrast.
- Ball distal to the FCC: complete occlusion of ACI and massive filling of the cavernous. (Image if you can).

It should be remembered that arteriography gives us additional information as pseudoaneurysms proximal or distal to the fistula, as well as in the contralateral ICA.

Remembrance that in B, C and D CCF cases, pay special attention to the venous drainage that is involved in the most serious clinical manifestations, such as neurological dysfunction and hemorrhage. The presence of defects of repletion within the dural sinuses, a sign related to venous thrombosis, is also evaluated in this phase, which is related to these indirect fistulas.

DIFFERENTIAL DIAGNOSES

There is a wide spectrum of differential diagnoses. In this sense, other causes (endocrine, infectious, inflammatory and neoplastic) should be considered at a first clinical stage compatible with CCF.

Intraorbital and intraocular lesions (frontal sinus mucocele, fibrous dysplasia, hemangioma, osteoma, ocular neoplasias, dysthyroid ophthalmopathy, conjunctivitis) may mimic the ophthalmologic clinic of the CCF, except for vascular murmurs.

When non-invasive radiological studies identify an increase in caliber of the ophthalmic veins, it will allow the clinician physician to orient the symptoms towards vascular cause and rule out the previous options. Cerebral arteriography will confirm the diagnosis.

TREATMENT

Typically, symptomatic FCCs are candidates for scheduled treatment. Nevertheless, there are several symptomatic situations in which urgent treatment is required.

- ACUTE HEMIPLEJIA without associated cerebral hematoma. This situation is caused by a phenomenon of theft in ACI distal to the CCF and insufficient collateral circulation

- HERNIATION OF THE CAVERNOSUS SINUS within sphenoidal sinus. Contains risk of EPISTAXIS AND / OR MASSIVE HSA.
- DRAINAGE OF CORTICAL VEINS: High risk of acute cerebral bleeding.

METHODS OF TREATMENT

Conservative.

- Employed in CCF with little symptom and low risk.
  - Drugs against ocular symptoms and autocompression of the carotid artery and the jugular vein ipsilateral to the FCC are used. This maneuver can lead to side effects such as symptomatic bradycardia or ischemia (due to carotid stenosis or embolism).

Surgical:

- Employed as a second option for endovascular failed treatment.
  - High morbidity due to damage in the cranial nerves involved and also to leave residual fistulous communications.

Endovascular.

- It will be based on the type of fistula.
  - The goal in the type A CCF is to occlude the defect of the ACI preserving the permeability of this artery. This purpose can be achieved through several options (Table 1). In weird cases, the FCC has a considerable defect requiring ipsilateral ICA occlusion for its treatment.
  - The objective in indirect CCF is to discontinue fistulous communications and reduce pressure within the cavernous sinus. In order to get this, two procedures may be used essentially (Table 1).

- Embolization of the cavernous sinus using detachable platinum coils (Fig 11) is used instead of detachable balloons for:
  1. High rate of formation of pseudoaneurysms using detachable ballons.
  2. Major risk of embolism using detachable ballons too.
  3. Softness and easy repositioning of the platinum coils. This allows a more adequate reconstruction of the ICA and to treat more complex CCF.
Fig. 4: Coronal non enhanced CT. Fractures in the left middle cranial fossa affecting the sphenoid body (yellow arrows).

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**Fig. 5:** Axial non enhanced CT. Subtle ingurgitation of the superior ophthalmic left vein (yellow arrow).

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Fig. 6: MRI T2 weighted: left exophthalmos. Left preseptal cellulitis (yellow arrow).

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**Fig. 7:** Orbital MRI T1 weighted. Ingurgitation of ophthalmic veins can be identified by means of vascular void.

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Fig. 8: IRM 3D reconstructions angiography. Direct CCF. Cavernous sinus enhancement (curve arrow) and ingurgitacion and also enhancement of the superior ophthalmic left vein (straight arrow).

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**Fig. 9:** IRM T1 weighted with gadolinium. Previous case. Ingurgitation and enhancement of the superior ophthalmic left vein it is easily identified on this sequence.

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**Fig. 10:** Standard cerebral angiography: left ICA selective catheterization. Anteroposterior (left) and lateral (right) views. Presence of left CCF type A (White circle. High flow: fast filling of CS) which drains to superior ophthalmic vein (double arrow), angular vein (straight arrow) and facial vein (curve arrow), as well as to the inferior petrous sinus (arrowhead).

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**Table 1**

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Fig. 11: Cerebral angiography. Post-embolization controls by contrast injection catheterizing the internal carotid left artery. FIRST CONTROL (LEFT). Left cavernous sinus is partially embolized. Persistent CCF, with drainage to ophthalmic veins (arrowhead). LAST CONTROL (RIGHT). Left cavernous sinus thrombosed, without contrast evidence in superior ophthalmic vein. (straight arrow). CCF excluded. End of procedure.

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Conclusion

Carotid cavernous fistula is a pathological entity that is often associated with traumatic history and whose gold standard imaging tool is cerebral arteriography, that also allows endovascular treatment with better outcomes and less complications than surgery.
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