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Exophthalmos Revealing A Carotid-Cavernous Fistula In A 77-Year-Old Patient

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Summary:
Carotid-cavernous fistula is an abnormal communication between the carotid arterial system and the cavernous sinus. The most frequent origin is congenital, due to arteriovenous malformations, but it can also be post-traumatic. This is a rare condition, which can be life-threatening and visually disabling. We report the case of a 77-year-old patient with a right carotid-cavernous fistula, whose ophthalmological complaint was revealing. Radiological examination was necessary to confirm the diagnosis and initiate urgent therapeutic management.

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I. Case report :

Carotid-cavernous fistula is an uncommon pathology. It is a therapeutic emergency whose symptomatology is suggestive, confirmed by imaging and which requires a rapid multidisciplinary approach.

We report the case of a 77-year-old female patient with type 2 diabetes on treatment, hypertension on treatment and glaucoma on treatment. She presented with ocular pain and reduced visual acuity dating back 10 days, without any traumatic context.

Ophthalmological examination of right eye reveals : finger-count visual acuity at distance, inferior serous chemosis with non-reducible, non-painful pulsatile axial exophthalmos (Figure 1); limitation of abduction and elevation (Figure 2; 3).

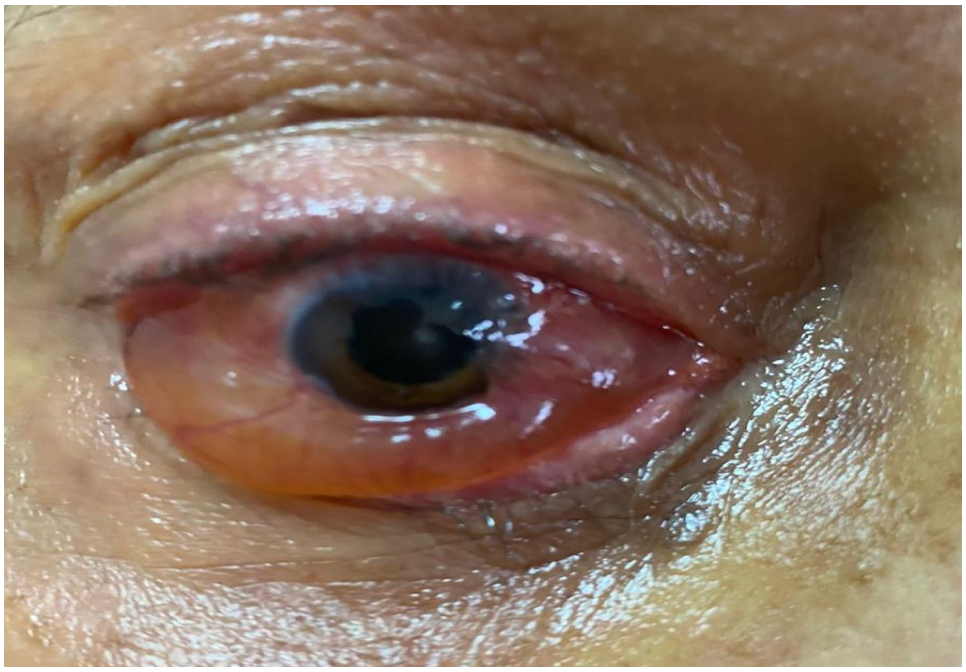

FIGURE 1 : Inferior serous chemosis with exophthalmos of the right eye.

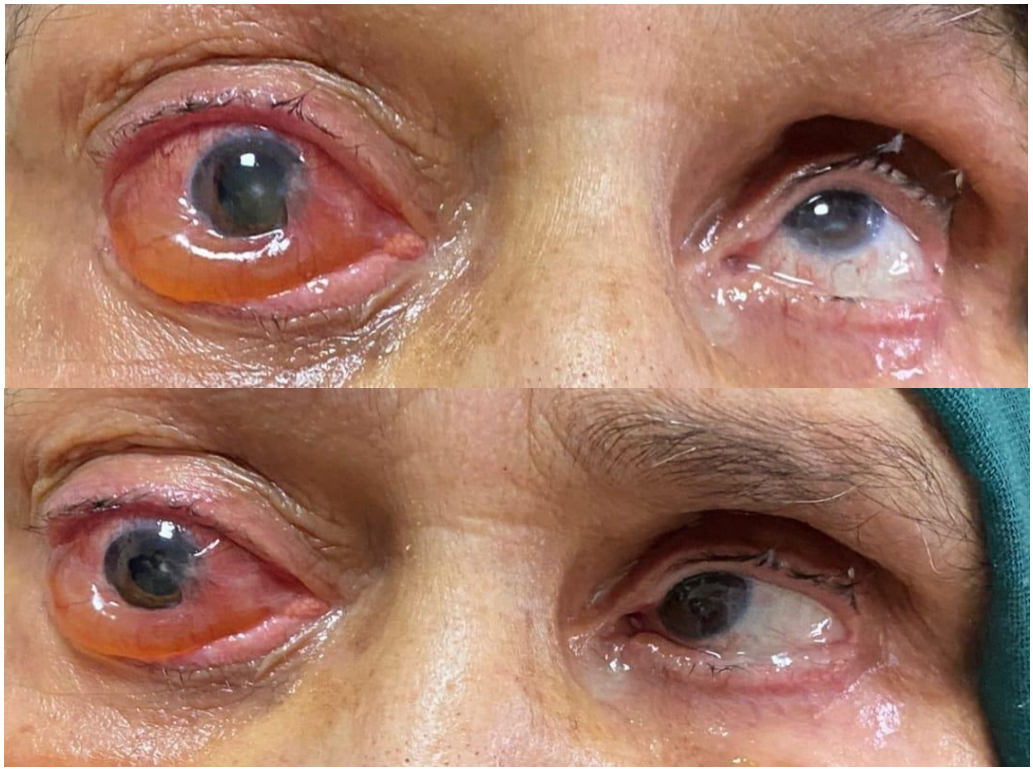

FIGURE 2 et 3 : Limited abduction and elevation of the right eye.

A cerebral and orbital CT scan was ordered, revealing a right carotid-cavernous fistula with grade I exophthalmos (Figure 4).

On the advice of the multidisciplinary consultation meeting, embolization of the dural fistula via the superior ophthalmic vein was proposed.

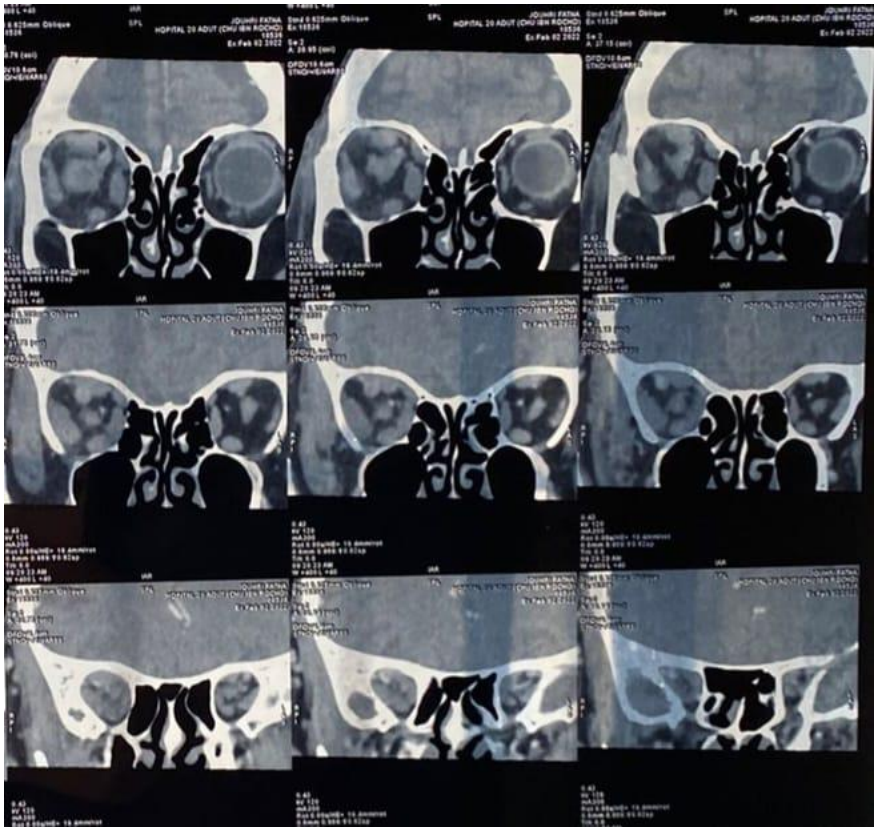

FIGURE 4 : Cerebral and orbital CT revealed a right carotid-cavernous fistula with grade I exophthalmos.

II. Discussion :

Carotid-cavernous fistulas (CCFs) are abnormal communications between the cavernous venous sinus and the carotid arterial system. Barrow's classification distinguishes 4 types of carotid-cavernous fistula (1):
- Type A: represents a direct communication between the internal carotid artery and the cavernous sinus. It most often occurs in a post-traumatic context and is rarely spontaneous.
- Type B: dural fistula between the meningeal branch of the internal carotid artery and the cavernous sinus.
- Type C: dural fistula between the meningeal branch of the external carotid artery and the cavernous sinus.
- Type D: type B+ type C. It should be noted that direct carotid-cavernous fistulas, classified as Barrow type A, are high-flow shunts, and occur three times more frequently than indirect types (2).

Delayed diagnosis is facilitated by discreet clinical signs. Exophthalmos (80%), dilated episcleral vessels, oculomotor paralysis, palpebral edema, chemosis, an audible vascular murmur (20%) on auscultation of the eyeballs, increased intraocular pressure (50%), fundus abnormalities such as stasis retinopathy or papilledema, and reduced visual acuity (20-30%) may be found (3-4).

Diagnosis is based on angioscan evidence of arterial enhancement within the cavernous sinus (5), dilatation of the superior ophthalmic vein with early enhancement is highly sensitive, while dilatation/enhancement of the superior pterygoid sinus and venous plexus and exophthalmos is highly specific (6).

Angiomagnetic resonance imaging (MRI) can also help with positive diagnosis, thanks to the "time-of-flight" sequence, which objectifies FCC with a sensitivity of 83% and a specificity close to 100% (7).

Treatment involves venous embolization, with retrograde access to the cavernous sinus via the superior ophthalmic vein or inferior petrous sinus. The principle is to directly occlude the venous collector by releasing coils in the cavernous sinus, resulting in a localized thrombogenic effect (8).

If left untreated, they can be life-threatening (cerebral or subarachnoid haemorrhage, epistaxis), as well as functional (ocular hypertonia, optic atrophy, reduced visual acuity) (9,10).

III. Conclusion :

Direct carotid-cavernous fistula is an uncommon entity. It is most often post-traumatic. The diagnosis should be made in the presence of uni- or bilateral exophthalmos with evidence of previous craniofacial trauma. Treatment must be multidisciplinary and rapid to avoid complications.

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