Carotid-cavernous fistula

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- Home-Based Therapy Utilizing Intermittent Manual Compression of the Carotid Artery and Internal Jugular Vein in the Management of Carotid-Cavernous Fistula

The carotid-cavernous fistula (CCF) and the cavernous sinus dural arteriovenous fistula (dAVF) are two distinct vascular pathologies involving abnormal connections in or around the cavernous sinus.

Carotid cavernous fistula (CCF) are abnormal communications between the internal carotid artery (ICA) or the external carotid artery and their branches and the cavernous sinus [1)]

General information

Key concepts

● direct (high flow, from ICA) or indirect (low flow, from meningeal branches)

● classic triad (more common with direct CCF): chemosis, Pulsatile exophthalmos, ocular bruit

● risk of SAH is low. The major risk is to vision

● the natural history of low flow CCF is up to 50% spontaneous thrombosis

History

The carotid-cavernous fistula was one of the first intracranial vascular lesions to be recognized.

The paper of Lang et al. focuses on the historical progression of our understanding of the condition and its symptomatology-from the initial hypothesis of ophthalmic artery aneurysm as the cause of pulsating exophthalmos to the recognition and acceptance of fistulas between the carotid arterial system and cavernous sinus as the true etiology. Lang et al. also discussed in 2016 the advancements in treatment from Benjamin Travers' early common carotid ligation and wooden compression methods to today's endovascular approaches [2)].

Epidemiology

Incidence and Prevalence: Carotid-cavernous fistulas are considered rare, with an estimated annual incidence of 0.17 to 0.37 per 100,000 persons. Prevalence rates vary depending on the population studied and the diagnostic criteria used.

Age and Gender: CCFs can occur at any age but are most commonly diagnosed in adults, particularly those in their 40s and 50s. There is a slight predilection for females, with some studies suggesting a female-to-male ratio of approximately 3:1.

Etiology: CCFs can be classified as spontaneous or traumatic. Traumatic CCFs often result from head trauma, such as motor vehicle accidents or sports injuries, while spontaneous CCFs may arise from various causes, including connective tissue disorders, fibromuscular dysplasia, atherosclerosis, or idiopathic reasons.

Associated Conditions: Carotid-cavernous fistulas may be associated with certain conditions or predisposing factors, such as Ehlers-Danlos syndrome, fibromuscular dysplasia, hypertension, atherosclerosis, or previous head or neck surgery.

Geographical Distribution: While there are no specific geographical patterns associated with CCFs, studies have reported cases worldwide, indicating that it is not limited to any particular region.

Diagnostic Advances: Improved imaging techniques, such as digital subtraction angiography (DSA), magnetic resonance angiography (MRA), and computed tomography angiography (CTA), have led to more accurate diagnosis and potentially higher reported incidence rates in recent years.

Overall, while carotid-cavernous fistulas remain relatively rare, improved diagnostic methods and treatment options have led to better recognition and management of this condition.

Classification

Carotid-cavernous fistula classification.

Etiology

Motor vehicle accidents, falls and other crush injuries contribute to the incidence of basilar skull fractures and the formation of some of the CCFs.

Clinical Features

Classic triad: chemosis, pulsatile exophthalmos, ocular bruit.

In the setting of a carotid cavernous fistula, flow often reverses into the superior ophthalmic vein, thereby producing evidence of orbital congestion as well as secondary increased intraocular pressure (related to the problems with ocular venous outflow). This flow reversal results in the myriad ophthalmic manifestations of a carotid cavernous fistula.

CCFs present with symptomatology resulting from venous congestion, hypertension, thrombosis, hemorrhage, neural compression, and/or ischemia from vascular steal.

With anterior venous drainage, increased venous pressure results in an increase in intraocular pressure leading to loss of vision, ocular pain, glaucoma, and retinal hemorrhage. Manifestations in the orbit include chemosis, exophthalmos, periorbital pain, and blepharedema. With posterior venous drainage, increased pressure in the cavernous sinus and vascular steal can result in cranial nerve deficits manifesting as ophthalmoplegia, diplopia, ptosis, or anisocoria. Most daunting is cortical symptomatology such as hemorrhage and seizures resulting from retrograde drainage into the superficial middle cerebral vein or the posterior fossa via the petrosal vein.

Diagnosis

CT or MRI

Pproptosis

Enlarged superior ophthalmic veins

Extraocular muscles may be enlarged

Orbital oedema

May show SAH/ICH from a ruptured cortical vein

Angiography (DSA)

Rapid shunting from ICA to CS

Enlarged draining veins

Retrograde flow from CS, most commonly into the ophthalmic veins

see Heuber maneuver

see Mehringer Hieshima maneuver.

Ultrasound

Complications

Subarachnoid hemorrhage is low. Major risk is to vision.

Outcome

50 % spontaneous thrombosis in low flow carotid cavernous fistula.

Treatment

see Carotid cavernous fistula treatment.

Case series

Ten out of a total of 31 direct carotid cavernous fistulas (DCCFs) were treated with Willis covered stents (WCSs) (Microport, Shanghai China) at West China Hospital from January 2015 to December 2016. The indications for treatment, perioperative findings, and postoperative and follow-up results were collected and analyzed.

All ten patients had successful deployment of WCSs. Complete exclusion of the fistula was achieved in 6 patients immediately after deploying one stent. Endoleak was observed in 4 patients (cases 2, 4, 5 and 9); thus, redilation of the stent with higher pressure was performed, which resolved the endoleak in 2 patients (cases 2 and 9). The other two patients' endoleak persisted after redilation of the balloon; hence, a second stent was deployed in these 2 patients (cases 4 and 5), which eliminated the endoleak in one patient (case 4), and the other patient (case 5) continued to have minimal endoleak. Nine patients had fistulas that were successfully occluded by WCSs during follow-up. One patient had recurrence of a DCCF at the 10-day follow-up; we chose coil embolization to address this DCCF. No stenosis of the internal carotid artery (ICA) or DCCF recurrence, except that in the abovementioned patient, was observed.

WCS was proven to be an alternative treatment method for complex DCCFs through reconstruction and preservation of the ICA. The study also confirmed the safety, efficacy, and midterm durability of WCSs for complex DCCFs without any serious delayed complications [3)].

A total of 55 patients with 56 tCCFs (1 bilateral tCCF) were included. Thirty-nine patients (40 tCCFs) were treated successfully in single session of a procedure, while 16 patients (16 tCCFs) experienced a recurrence of tCCF. In multivariate analysis, Lin et al. found that the involvement of C2 or C4 segments (Debrun classification) of intra-cavernous internal carotid artery is an independent risk factor (HR: 2.95, 95% CI: 1.34 - 6.52; $p < 0.01$) for the recurrence of tCCFs. Endovascular coil embolization demonstrated superior efficacy in successful interventions of tCCFs when compared with detachable balloons (HR: 2.63, 95% CI: 1.06 - 6.57; $p < 0.05$) and other modalities (HR: 3.06, 95% CI: 1.27 - 7.37; $p < 0.05$).

A detachable coil is a favorable approach in the management of tCCFs when considering the rate of recurrence. In addition, the involvement of C2 or C4 segments (Debrun classification) served as an independent risk factor of the recurrence of tCCFs. [4)].

[1)]
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