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CAROTID-CAVERNOUS FISTULA (CCF) - A REVIEW

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ABSTRACT A carotid-cavernous fistula (CCF) is the result of an abnormal vascular connection between the carotid system (ICA/ECA) and the venous channels of the cavernous sinus. CCFs are classified based on the arterial system involved, hemodynamics, and etiology. The presentation can be varied and nonspecific; commonly presents with ophthalmic manifestations due to impairment of venous drainage of the orbit from the cavernous sinus. Early diagnosis and appropriate management is essential to avoid vision and life-threatening complications. Patients with CCF may initially present to an ophthalmologist with decreased vision, conjunctival chemosis, external ophthalmoplegia and proptosis. Ophthalmologist may be the first physician to encounter a patient with clinical manifestations of CCF, and this review article should help in understanding the clinical features of CCF, current diagnostic approach, usefulness of the available imaging modalities, possible modes of treatment and expected outcome.

KEYWORDS : Carotid, Cavernous Sinus, Diagnosis, Fistula, Ophthalmological Findings.

INTRODUCTION:

CCFs are a rare entity that can occur spontaneously or secondary to trauma. Traumatic CCFs account for the majority of CCFs. They can occur following closed head injury, skull base fractures, penetrating head trauma, or after craniotomies, endoscopic trans-sphenoidal or sinus surgery, and endovascular procedures. [1]

The true incidence is not well-documented; however, studies have reported an incidence of 0.2% in patients with traumatic brain injury and up to 4% with skull base fractures. [2, 3] Bilateral CCFs are rare but more commonly reported in traumatic CCFs, occurring in up to 1% of traumatic cases. [4]

Spontaneous CCFs account for up to 30% of all CCFs reported in the literature. [5]

Spontaneous CCFs may result from the rupture of an aneurysm of the cavernous segment of the internal carotid artery (ICA). This can occur in up to 24% of individuals with such aneurysms, dependent on size and morphology. Musculoskeletal and collagen related disorders including Ehler's-Danlos syndrome, pseudoxanthoma elasticum, osteogenesis imperfecta, and fibromuscular dysplasia are also thought to predispose to CCF formation due to presumed arterial wall defects and risk of dissection.[6-8] Associations with hypertension, female gender, and older age have also been reported. An increased incidence is seen in pregnancy and is postulated to be related to a hormonally induced hypercoagulable state or cavernous sinus thrombosis.

CLASSIFICATION:

CCFs are primarily classified into 4 types based on the arterial system involved as described by Barrow et al (Table 1).

Table 1: Barrow Classification [9]

TYPE	DRESCRIPTION
A	Direct connection between the ICA and cavernous sinus
B	Connection between meningeal branches of ICA and cavernous sinus
C	Connection between meningeal branches of ECA and cavernous sinus
D	Connection between meningeal branches of both ICA and ECA and the cavernous sinus

Table 2: CCF Classification [6]

TYPE	CLASSIFICATION
Anatomical	Direct vs Indirect
Hemodynamic	High vs Low Flow
Etiology	Traumatic vs Spontaneous

An updated classification system proposed by Thomas et al. is based on venous drainage and demonstrates a significant correlation with clinical symptoms, treatment planning, and outcome. [10]

Table 3: Classification of CCF by Venous Drainage.

TYPE	DESCRIPTION
1	Posterior/inferior venous drainage only
2	Posterior/inferior and anterior venous drainage
3	Anterior venous drainage only
4	Retrograde cortical venous drainage
5	Direct ICA-cavernous sinus fistulae corresponding to the type A Barrow classification

PATHOPHYSIOLOGY:

All types of CCFs lead to shunting of blood from a high-flow arterial system (ICA or ECA) into a low-flow venous system (cavernous sinus) without an intervening capillary bed i.e arterialization of cavernous sinus. This produces increased vascular pressure and resistance which impedes venous drainage and leads to vascular congestion in draining areas. Impaired drainage of the orbit in an anterior draining CCF leads to the common ophthalmic manifestations as a result of congestion, ischemia, and mass effect.

Direct or Type A CCFs are the most common type and form a direct connection between the cavernous segment of the ICA and the cavernous sinus. Type B, C, and D CCFs are dural, low-flow fistulae that result from an indirect connection between meningeal branches of the internal carotid artery (ICA) and/or external carotid artery (ECA) and the cavernous sinus. Indirect CCFs are considered low-flow shunts. The majority of indirect CCFs occur spontaneously and are thought to be caused by a dural rupture in the arterial wall; however, the pathophysiology is poorly understood.

The diagnosis of CCF is based on the combination of clinical presentation, physical and ophthalmic examination, and diagnostic procedures including neuroimaging (MR Angio + MR Venogram (TRICKS sequence) /CT Angio brain/ DSA –Cerebral Angio).

CLINICAL PRESENTATION:

Direct CCFs tend to have a more severe, acute onset presentation while indirect CCFs are associated with a more gradual onset and chronic course.

Direct CCFs often present with a classic triad of pulsatile exophthalmos, orbital bruit, and chemosis. Patients may report diplopia, ocular redness, orbital/retro-orbital pain, swelling, swishing or buzzing sounds, headache, or vision loss. Patients can also have pulsatile tinnitus. Symptoms are typically ipsilateral to the fistula but can occur bilaterally depending on the severity and chronicity of venous congestion. (Figure 1 and 2).

Figure 1.a. B/L chemosis, proptosis.

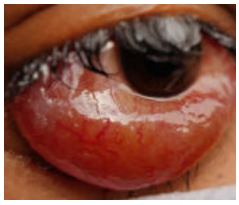

Figure 1.b. Proptosis with chemosis with corkscrew conjunctival vessels.

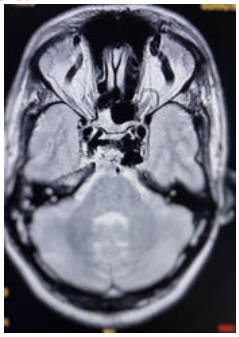

Figure 1.c. MRI BRAIN T2 sequence showing flow voids of B/L ICA communicating with cavernous sinus with prominent B/L superior ophthalmic veins suggestive of Type A CCF.

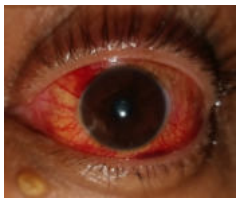

Figure 2. a. Unilateral prominent corckscrew conjunctival vessels

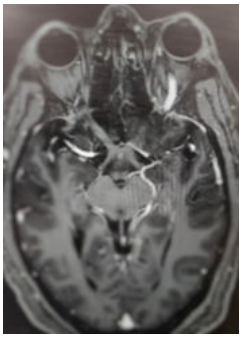

Figure 2.b. CEMRI Brain showing bulky left cavernous sinus with prominent left superior ophthalmic vein.

Ophthalmic signs that can be seen in both direct and indirect CCFs include proptosis, eyelid edema, ptosis, conjunctival arterialization, elevated intraocular pressure, pulsatile exophthalmos, pupillary abnormalities, ocular misalignment, papilledema, dilated retinal veins, ischemic optic neuropathy, central retinal vein occlusion, or choroidal detachment. The conjunctival hyperemia associated with CCFs is a result of the arterialization of conjunctival and episcleral veins, consisting of distinct tortuous corkscrew blood vessels that converge at the limbus. [11,12]

In contrast to direct high-flow CCFs, indirect low-flow CCFs may be asymptomatic and insidious in onset. The most common presentation is conjunctival injection which may be initially treated as conjunctivitis causing a delay in diagnosis. Cranial nerve deficits from ischemia to cranial nerves III, IV, and/or VI can occur and change over time.

DIAGNOSTIC IMAGING:

The goal of diagnostic procedures is for initial diagnosis of the CCF and then to evaluate the anatomy and physiology of the CCF as it relates to potential treatment options. Most patients initially undergo noninvasive imaging with computed tomography (CT), magnetic resonance imaging (MRI), CT angiography (CTA), or magnetic resonance angiography (MRA).

If there is a high degree of clinical suspicion or suggestion of a CCF on noninvasive imaging, diagnostic cerebral angiography (digital subtraction angiography, DSA) can confirm the diagnosis and guide treatment.

CTA and MRA will identify the majority of arterial dissections and cavernous ICA aneurysms causing a CCF as well as enlargement of the affected CS. These modalities can also detect abnormal flow voids, proptosis, enlargement of the SOV/IOV and enlargement of extraocular muscles. [14]

CT without contrast is a useful screening modality to detect skull base fractures in the setting of trauma.

MRI can better demonstrate fat stranding reflecting orbital edema and abnormal flow voids.

DSA remains the gold standard imaging modality in the diagnosis of direct and indirect CCFs. DSA is necessary to identify CCF location, arterial supply, flow rate, and venous drainage to classify the CCF and help plan for potential endovascular treatment strategies. [14]

DIFFERENTIAL DIAGNOSIS:

Cavernous sinus thrombosis
Tolosa hunt syndrome
Pseudotumor orbitalis- IGG4 disease
Superior orbital fissure syndrome
Orbital apex syndrome
Invasive mycoses
Retrobulbar haemorrhage

TREATMENT:

Patients with asymptomatic, incidentally discovered indirect CCFs can often be safely observed with routine follow-up to assess for ophthalmologic symptoms and exam changes. Patients with mild ocular symptoms can be treated with appropriate topical medications and observed for visual acuity, intraocular pressure, or ophthalmoscopic changes. Indirect CCFs may also spontaneously resolve in some cases. Direct CCFs are rarely asymptomatic and need urgent treatment. [15]

DIRECT CCFs:

Direct CCFs are unlikely to close spontaneously, and upfront treatment is indicated due to the risk of neurological deficits. There are numerous endovascular techniques utilized including coil embolization with balloon remodeling of the ICA and coil embolization with stent assistance. Combined transvenous and transarterial approaches can be used to

preserve the ICA and pack the CS with coils to stop flow. Liquid embolics such as Onyx (ethylene vinyl alcohol copolymer, Medtronic, USA) and n-butyl cyanoacrylate glue (n-BCA, Trufill, Cerenovus, USA) are less commonly used for direct CCF due to the risk of distal embolization into cerebral arteries and stroke. A large cavernous ICA defect such as that from a traumatic transection may require ICA sacrifice as a life-sustaining treatment. Parent vessel sacrifice (endovascular occlusion) may also be option in cases of recurrence if the patient passes a balloon test occlusion. Less commonly used approaches include covered stents, packing the cavernous sinus via open microsurgery, and cavernous ICA trapping with bypass. [16-18]

INDIRECT CCFs:

Indirect CCFs that fail conservative therapy or demonstrate progression of symptoms can be considered for endovascular treatment.

Endovascular treatment (similar to that of direct CCFs) of indirect CCFs is primarily via a transvenous route unlike treatment of direct CCF. [19]

Radiosurgery may be considered in patients with low-flow, indirect CCFs who cannot tolerate endovascular treatment or as salvage therapy for recurrent CCF with limited endovascular options.

PROGNOSIS:

The results of embolization for direct CCFs and indirect CCFs are favorable with successful angiographic closure in up to 93% and 92%, respectively. [20, 21] Direct or indirect CCFs carry a favorable visual prognosis unless there is evidence of retinal or optic nerve ischemia prior to treatment.

Improvement of proptosis, chemosis, and extraocular muscle enlargement is typically seen within weeks of treatment. A transient "paradoxical worsening" after treatment due to an extension of cavernous sinus thrombosis to the SOV may occur and can be mitigated with corticosteroids prior to spontaneous resolution.

ABBREVIATIONS:

ICA-Internal Carotid artery; ECA- external carotid artery;CCF-Carotid-cavernous fistula; SOV- Superior ophthalmic vein; IOV- Inferior ophthalmic vein.

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