



Case Report

Indirect Left Carotidocavernous Fistula

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Background

The carotidocavernous fistula (CCF) is an arteriovenous fistula and it is divided in general to direct fistula and indirect types. Indirect fistulas are due to communication by multiple branches between the internal and/or external carotid arteries and the cavernous sinus. The most frequent with is where the meningeal branches of the external carotid forming the fistula [1]. The indirect type came with important signs and symptoms as mentioned in the case report below. 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 iatrogenically after craniotomies, endoscopic transsphenoidal or sinus surgery, and endovascular procedures [2,3]. These CCFs typically correspond to Type A CCFs (see Classification section). 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 [4]. Bilateral CCFs are rare but more commonly reported in traumatic CCFs, occurring in up to 1% of traumatic cases [5].

Spontaneous CCFs account for up to 30% of all CCFs reported in the literature and is more common in the postmenopausal patients [6,7]. 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 [6]. 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 [8-10]. CCF from an cavernous ICA aneurysm rupture or cavernous ICA dissection would commonly result in a Type A CCF (see Classification section).

Abbreviations: IOP: Intraocular Pressure; CCF: Carotidocavernous Fistula; OCT: Ocular CT; MDT: Multi-disciplinary Team; ICA: Internal Carotid Artery; ECA: External Carotid Artery; CS: Cavernous Sinus; CN: Cranial Nerve; SOF: Superior Orbital Fissure; SOV: Superior Ophthalmic Vein; IOV: Inferior Ophthalmic Vein; CTA: CT Angiography

Direct CCFs [11-15]

Direct CCFs are unlikely to close spontaneously, and upfront treatment is indicated due to the risk of neurological deficits and worsening of venous congestion. Signs for urgent treatment of direct CCFs include visual impairment, progressive paresis of extraocular muscles, intractable orbital pain, bruit, and progressive exophthalmos. Because direct CCFs originate from a defect in the cavernous ICA either from an aneurysm rupture or dissection, the goal of treatment is to cease flow into the CS and reconstruct the cavernous ICA. There are numerous endovascular techniques utilized to achieve this end including coil embolization, 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.

Indirect CCFs [11-15]



Figure 1: Cerebral angiogram, ICA.

Indirect CCFs, this occurs when there is an irregular connection between the carotid artery and the cavernous sinus. That fails conservative therapy or demonstrates progression of symptoms can be considered for endovascular treatment. Symptoms may include decreased vision, diplopia, intractable headache or bruit, or worsening proptosis with exposure keratitis can. Further, neurological deficits, intradural hemorrhage, and venous thrombosis indicate a need for more urgent treatment. While endovascular treatment is the mainstay of therapy, manual compression therapy via contralateral hand compresses has been reported to occlude indirect CCFs in up to 30% of cases. Indirect CCFs may also spontaneously thrombose in up to 60% cases.

Endovascular treatment of indirect CCFs is primarily via a Tran's venous route unlike treatment of direct CCF. Tran's arterial embolization can be attempted if the primary fistulous connection is from branches of the ECA but fistulous connection from Dural branches the ICA limits the Trans arterial approach due to the risk of stroke and arterial dissection with treatment. Transvenous embolization occurs via catheterization of the cavernous sinus commonly via the petrosal sinuses and pterygoid plexus and less commonly via a transorbital approach. Embolization proceeds with detachable coils or liquid embolics (Onyx, n-BCA) to obliterate the fistula, restore normal orbital venous drainage and preserve intradural arterial flow. During transvenous embolization, the arterial circulation is monitored with intermittent angiography

via the ipsilateral ICA to ensure the patency of normal cerebral vasculature.

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.

Anatomy [16-19]

The cavernous sinus (CS) is a paired dural venous sinus centered around the sella turcica. It contains multiple venous channels extending from the endosteal dural layer of the sphenoid bone inferiorly and medially to the meningeal dural layer of the floor of the middle cranial fossa. The CS extends from the superior orbital fissure (SOF) anteriorly to the petrous apex posteriorly, between the dorsum sellae medially and Meckel's cave laterally. The CS serves as a confluence of venous systems draining the orbit (via superior and inferior ophthalmic veins, SOV, IOV), Sylvian fissure, anterior and middle cranial fossae (via sphenoparietal sinus) and posterior cranial fossa (via the basilar plexus and superior and inferior petrosal sinuses), and they are connected by intercavernous sinuses existing within the sella. The CS also drains inferiorly via emissary veins through the foramen ovale into the pterygoid venous plexus. The venous drainage patterns of the cavernous sinus are pertinent for the pathophysiology of CCFs.

The cavernous segment of the ICA courses through the cavernous sinus. This segment has also been referred to as both the intercavernous and intracavernous ICA. The ICA enters the cavernous sinus as it passes the petrolingual ligament and exits at the proximal dural ring becoming the clinoidal segment. Although the course of the cavernous ICA is variable and dependent of patient specific factors, the typical segments include the proximal ascending, posterior genu, horizontal and anterior genu. The anterior genu is at the posterior border of the optic strut, a region of bone between the anterior clinoid process and body of the sphenoid between the optic canal and SOF. Branches of the cavernous ICA include the meningo-hypophyseal trunk (MHT), inferolateral trunk (ILT) and capsular arteries of McConnell.

The oculomotor (CN III), trochlear (CN IV), trigeminal (CN V) and abducens (CN VI) nerves are associated with the CS. From superior to inferior, CNs III, IV, V1 (ophthalmic division) and V2 (maxillary division) course within the lateral wall of the CS. CNs III (superior and inferior divisions), IV, V1 (lacrimal nerve, frontal nerve, nasociliary branch) and VI exit the CS via the SOF along with the SOV and IOV.

Classifications

CCFs are primarily classified into 4 types based on the arterial system involved as described by Barrow et al [15] (Table 1). Type A are direct fistulas, whereas types B, C, and D are indirect

fistulas. CCFs are further classified into traumatic or spontaneous etiology and high or low flow based on hemodynamic properties (Table 2). The flow rate is determined by angiography and largely influences clinical presentation (Table 3).

Type	Description
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 1: Barrow Classification.

Type	Classification
Anatomical	Direct vs Indirect
Hemodynamic	High vs Low Flow
Etiology	Traumatic vs Spontaneous

Table 2: CCF Classification.

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

Table 3: Classification of CCF by Venous Drainage [20].

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. This produces increased vascular pressure and resistance which impedes venous drainage and leads to vascular congestion in areas that are drained by the cavernous sinus. Impaired drainage of the orbit in an anterior draining CCF is what leads to the common ophthalmic manifestations as a result of congestion, ischemia, and mass effect [21].

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. This high-flow variant of CCFs causes retrograde blood flow from the cavernous sinus into the superior ophthalmic vein (SOV) leading to dilation of the SOV and ophthalmic clinical manifestations. The majority of direct CCFs is caused by trauma, frequently involving a tear in the muscular wall of the ICA, a laceration of one of its branches, or completes transection of the ICA by shearing forces. The cavernous segment of the ICA is particularly susceptible to injury in basilar skull fractures. In a review of 91 cases with a direct CCF, Gupta et al. identified 85 trauma-related causes, while 6 were secondary to aneurysm rupture. Less commonly, direct CCFs can occur spontaneously; particularly in patients with predisposing muscular and collagen disorders or cavernous ICA aneurysms.

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 because they originate from these dural arterial branches rather than the high-flow ICA. The majority of indirect CCFs occurs spontaneously and is thought to be caused by a dural rupture in the arterial wall; however, the pathophysiology is poorly understood. Associations with hypertension, female gender, and older age have been reported. An increased incidence is seen in pregnancy and is postulated to be related to a hormonally induced hypercoagulable state [22,23]. Indirect CCFs can also be seen in patients with sinusitis, recent trauma or surgery, hypercoagulable states, and cavernous sinus thrombosis.

Case Study

A 59 years old woman with a background history of thyroidectomy on levothyroxine presented to the ophthalmology outpatient clinic with a one week history of bilateral blurred vision, no redness or tearing. Her eye examination showed bilateral mild cataract and her investigation showed that the HbA1C was 9.5. At that point, metformin 500mg once a day was commenced and lifestyle changed has been advised. Gradually, the visual acuity became better. The cataract just needed follow up at that point without any intervention was needed.

Two weeks later, she waked up in the morning with mild proptosis in the left eye. There was no blurred vision, redness, pain or any eye other eye symptoms. No eye pain or signs of thyroid ophthalmoplegia. Because her hypothyroidism history and being on levothyroxine 150 microgram, she went to the endocrine outpatient clinic which showed the thyroid function test within normal limit and this proptosis is not related to the thyroid gland. Gradually, the proptosis became more obvious with gradually blurring of vision in the left eye. She went to the ophthalmology clinic; she underwent a full eye examination including retinal examination by the slit lamp and an ocular CT (OCT) which did

not show any abnormal finding in the optic nerve.

Investigations

Thyroid function test: normal

HbA1C: 9.5

Her intraocular pressure (IOP) was gradually increasing from 14 then 16 then jumped to 25

Treatment

Because of the increasing in the intraocular pressure, the patient was started on timolol drops to reduce the IOP. Also she was, on hypermellose eye drops.

Progression of the Condition

Despite being on the timolol eye drops, her intraocular pressure was still running high ranging from 22-26. The proptosis was gradually increasing for 4 weeks duration and the visual acuity was also decreasing in the left eye and became blurry. The right eye was alright without any symptoms. She had another eye examination which did not show any findings. She became very anxious about the shape of the left eye and she was thinking about retro orbital tumor. She had a CT angiography which showed there was a left indirect coraticocavernous (arteriovenous) fistula. The ophthalmologist did a referral to the neurosurgeon to take his opinion. There was a multi-disciplinary team (MDT) meeting included the ophthalmologist, a neurosurgeon and the interventional radiologist and the outcome was the best option is to do an angiography and angioplasty (embolism) urgently to close the fistula, control the IOP pressure and to prevent any damage to the optic nerve.

The Diagnosis

Indirect coraticocavernous fistula

Post Operation Care and Management

The patient was not able to close her left eye fully before the time of the operation, the visual acuity was blurry and the IOP was 24. The operation (coil embolization) went urgently to save the optic nerve from damage by increasing the pressure inside the anterior chamber. The patient had the procedure through the left carotid artery under general anesthesia. The procedure lasted around 5 hours. The operation went well without any complication and the patient was admitted in the intensive care unit for monitoring for 24 hours' time.

The patient was admitted in the ophthalmology ward for another 48 hours. There was no complication and the patient has been discharged on timolol, enoxaparin prophylactic dose and analgesia.

The visual acuity of the left eye returned back to normal and the proptosis has been resolved. The patient still using the timolol and the IOP has been reduced to 16. She is English teacher and happily returned back to her career.

Take Home Message

The indirect coraticocavernous fistula is one of the important differential diagnosis in causing eye proptosis (exophthalmos) and increase the IOP. We should act urgently by doing the CT angiography (CTA) in the carotid artery to locate the fistula and to try to do an embolization. The most important this is to monitor the IOP and to save the optic nerve from damage (amblyopia).

NB: I have gained consent from the patient.

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