Acute Exacerbation on Stable Longstanding Right Carotid-Cavernous Fistula: A Case Report

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ABSTRACT
A 32-year old woman presented with worsened headache for the past 2 weeks accompanied by an audible tapping sound in her right ear. She experienced significant head trauma about 10 years before. Clinical examination showed a mild protrusion of right eye with a small conjunctival chemosis. Computed Tomography Angiography (CTA) indicated giant fusiform mass at cavernous segment of right internal carotid artery. Cerebral Digital Subtraction Angiography revealed a right carotid-cavernous fistula. The patient underwent surgery to entrap the aneurysm portion from internal carotis artery (IC) after the bifurcation, to proximal ophthalmic segment of IC. After surgery, visual acuity of right eye is preserved, and follow up CTA showed disappearance of CCF. The protrusion improves after one year follow up.

Keywords: carotid-cavernous fistula, internal carotid artery, protrusion, audible tapping sound, headache

INTRODUCTION
Carotid-cavernous fistula (CCF) is an abnormal communication between internal carotid artery and cavernous sinus. CCFs are classified into direct (type A) and indirect (type B, C, D) types. A direct CCF is a rare life-threatening disorder where a direct shunt arises between the internal carotid artery (ICA) and the cavernous sinus. The classical triad of pulsating exophthalmos, conjunctival chemosis, and pulsatile-tinnitus are well established clinical symptoms of the disease but are not usually present in the majority of patients as early indication.

CASE REPORT
A 32 years old female worker presented with severe right-sided headache for the past 2 weeks. The headache worsened in the last few days. Pain medication doesn’t help. An audible tapping-like sound was also heard when the headache occurred. Her right eye protruded but her visual acuity is intact for the past 10 years.

She had an accident ten years ago; she was thrown off her motorcycle and was unconscious. Three months after the accident, she went to a neurologist due to severe headache. In the following few months, she realized her right eye became protruded, accompanied by occasional soft clicking noises on the right

Figure 1 The MSCT-Angiography demonstrated a sac aneurysm mass in distal right ICA
On clinical examination the right orbital is protruded with mild conjunctival chemosis. Visual acuity and pupillary reflex were normal. Extra ocular movements are full. A mild bruit was audible over the right palpebra.

Computed Tomography Angiography (CTA) demonstrated a giant fusiform aneurysm on the cavernous segment of right internal carotid artery, 29 mm in diameter, with thrombosed wall. Transfemoral Digital Subtraction Angiogram (DSA) revealed the carotid cavernous fistula (CCF) with high flow to the jugular vein.

The operation was done by trapping the right ICA, from cervical segment to the proximal ophthalmic segment to preserve the ophthalmic artery.

Four months after surgery, CTA confirmed no fistula. Clinically she is improved, no bruit and no additional deficits.

**DISCUSSION**

The sinus essentially functions as a dural venous structure, receiving blood supply from superior and inferior ophthalmic veins. The internal carotid artery (ICA) functions as the blood supplier to the anterior brain. The ICA enters the skull and passes through cavernous sinus on its medial aspect but still bound by dural attachments, especially at its entrance and exit. The ICA leaves the cavernous sinus underneath the anterior clinoid process of the sphenoid bone and it is fixed to the surrounding dura matter. Other clinical significant structures within the cavernous sinus are cranial nerves III, IV, V1, V2, and VI. This anatomical configuration is very important to evaluate CCFs patients.

Abnormal communications between the cavernous sinus and carotid artery can be classified by etiology (traumatic, spontaneous; iatrogenic), by flow dynamics (low flow; high flow) and by anatomy (direct versus dural; ICA versus external carotid artery (ECA) versus both). Other classification by angiography depends on blood flow velocity across the shunt and the anatomical origin of the arteries supplying the cavernous sinus fistula. The famous classification was made by Barrow et al: type A fistulas are high flow direct fistula between ICA and cavernous sinus, usually traumatic. Type B, C, and D CCFs are low indirect fistula where multiple microfistulas are located within the wall of cavernous sinus and drain into it (table 1).

**Table 1** Anatomic-angiographic classification of CCFs

<table>
<thead>
<tr>
<th>Category</th>
<th>Definition</th>
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<tr>
<td>Type A</td>
<td>Direct high flow fistulas resulting from a tear between the ICA and the cavernous sinus</td>
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<tr>
<td>Type B</td>
<td>Dural shunts between meningeal branches of the ICA and the cavernous sinus; spontaneous</td>
</tr>
<tr>
<td>Type C</td>
<td>Dural shunts between meningeal branches of the ECA and the cavernous sinus; spontaneous</td>
</tr>
<tr>
<td>Type D</td>
<td>Dural shunts between meningeal branches of both the ICA and ECA and the cavernous sinus; spontaneous</td>
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Regardless of the direction of drainage, CCFs often present with ocular symptoms due to its anatomical relation. Exophthalmus may present in 72% of patients but its extent is highly variable (from 3 to 16 mm) (11, 43). Ocular pulsation is caused by pulsatile wave from the ICA level or the ophthalmic artery through dilated ophthalmic veins. Bruit can be heard by the patient and objectively by physicians. The subjective bruits are result of bone conduction from posterior drainage. Chemosis and arterialisations of the episcleral veins are present in 55% of patients. In this case report, the bruit can be heard by the patient and mildly in examination which demonstrated the posterior drainage and in accordance with the DSA imaging result.
Cranial nerves palsy may occur due to nerves compression. Diplopia may present in 60-70% of cases and abducens palsy occurs in 49-85% of patients, the oculomotor and trochlear nerve are less common damaged due to their localization in the lateral wall of cavernous sinus. Visual impairment has been reported in some degree particularly in untreated cases where the fistula is not closed.

Ocular and head pain is more often a minor component of the presentation, although the later is presented as the chief complaint in the patient of this report. The most likely cause of the ocular and head pain is stretching of the richly innervated dural membrane in CCF including the cavernous sinus, venous thrombosis, venous hemorrhage, or pathologic involvement of the V cranial nerve, either by direct pressure in the cavernous sinus or pulsatile pressure from abnormal arterial flow into the sinus. The presence of tinnitus and proptosis are helpful features.

If untreated, CCFs may have severe effects and can lead to death. Halbach et al have identified high-risk features in 155 cases of CCF including 127 direct fistulas, important risk factors responsible for acute deterioration includes increased intracranial pressure (8.7%), decreased visual acuity (32.3%), rapidly progressive proptosis (1.6%), and cerebral ischemia and hemorrhages (11%).

CT and MRI studies as initial investigation help to detect orbital involvements, but the diagnosis of CCF can be confirmed by more invasive diagnostics; cerebral angiography is the ‘gold standard’ for diagnosing CCF. Contrast-enhanced CT, CT-angiography and MR-angiography imaging show enlarged tortuous superior ophthalmic veins (the “hockey stick sign”). Selective DSA is essential for confirming the diagnosis and delineating the exact venous drainage patterns for the evaluation of the fistula prior to treatment.

Treatment of direct CCF is mandatory for immediate orbital symptom relief and to prevent the development of intracranial venous hypertension. The goal of treatment is the occlusion of the fistula and maintaining carotid artery patency which is mandatory when the collateral blood flow is insufficient.

Historically, angiography is in use for the treatment of intracranial vascular pathologies since at least the 1960s and followed by endovascular treatment. There has been continuous innovation in angiographic equipments and catheter technologies. Balloon embolization, coil embolization, combination of balloon-coil embolization, stents, and use of liquid embolization agents perform transarterial and transvenous approach. Very rarely, if all endovascular approach fails, ptetional craniotomy with a Dolen approach is pursued.

In this reported patient, the careful decision to perform trapping of ICA made with social consideration after ensuring that contralateral ICA is sufficient.

After successful occlusion of the CCF, the symptoms especially orbital will gradually disappear in days to several months. Large studies with a long follow-up time (> 10 years) have not been performed, but after occlusion of the CCF, most patients remain asymptomatic. Recurrence of the fistula is possible, Luo et al report that the recurrence rate is highest in patients in which the initial treatment was delayed and with large fistula.

In conclusion, the suspicion of CCF in this patient is directly made because there are positive triad symptoms (exophthalmus, pulsatile-tinnitus or audible bruit sounds, and conjunctival chemosis), although the first dominant complaint was severe headache which is not often presented in CCF. Diagnosis of traumatic CCF was confirmed by DSA showing a picture of direct fistula formation between cavernous sinus and ICA. It was supported by history of trauma; trauma is the most frequent etiology of direct CCF type. Treatment of choice for CCF should be endovascular embolization but in this case, trapping of ICA was the choice. The outcomes are usually asymptomatic yet post operative investigation should be performed to confirm the result of therapy.

REFERENCES: