Illustrative Teaching Case

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Carotid-Cavernous Fistula
A Rare but Treatable Cause of Rapidly Progressive Vision Loss

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Case Description

An 89-year-old woman with hypertension, hyperlipidemia, stroke, deep venous thrombosis, and atrial fibrillation presented with 4 days of right eye (OD) redness and swelling and intermittent diplopia. An ophthalmologic evaluation revealed visual acuity 20/50 OD. There was limited abduction OD, right upper and lower lid erythema and edema, and conjunctival injection. Funduscopic examination was unremarkable. Erythrocyte sedimentation rate and C-reactive protein were normal. Follow-up ophthalmologic examination 2 weeks later showed visual acuity decreased to 20/70 OD with increased right orbital congestion, exophthalmos (25 mm OD and 21 mm OS [left eye]), and worsening limitation of ductions OD. A magnetic resonance imaging/magnetic resonance angiography of her head demonstrated fusiform dilatation of the cavernous segment of the right internal carotid artery (ICA) with asymmetrical enhancement of the right cavernous sinus and an enlarged right superior ophthalmic vein (SOV) consistent with arterialized flow from a carotid-cavernous fistula (CCF).

The patient was urgently referred to neuro-interventional radiology for diagnostic angiography and embolization of the CCF. The cerebral angiogram showed a right direct (Barrow type A) CCF, with rapid enhancement of the right cavernous sinus and right SOV. A 7-mm fusiform aneurysm of the right cavernous ICA was found to be the likely cause of the CCF (Figure 1A). Transarterial access of the fistula for coil embolization was unsuccessful.

Repeat ophthalmologic examination demonstrated continued worsening of her visual acuity to 20/400 OD, elevated intraocular pressure in the right eye (24 mmHg), a relative afferent papillary defect, and complete limitation of ductions OD. External examination revealed worsening right upper and lower lid edema and erythema, proptosis, and conjunctival injection with corkscrew vessel tortuosity (Figure 2A). Goldmann visual field testing revealed 360° significant peripapillary constriction. Four days after her initial angiography, the patient underwent coil embolization of her right direct CCF via direct access of the right SOV provided by the Ophthalmic Plastic Surgery Service (Figure 1B and 1C). Complete closure of the CCF was demonstrated without residual arteriovenous shunting (Figure 1D).

Postoperatively, her visual acuity, intraocular pressure, congestion, chemosis, lid edema, and extraocular eye movements have improved substantially (Figure 2B).

Discussion

CCFs arise from abnormal connections between the carotid arteries and the cavernous sinus.1 These acquired vascular malformations disrupt the cavernous sinus, an intricate anatomic site of cerebral venous drainage, through which the ICA and cranial nerves III, IV, V1, V2, and VI traverse.

CCFs are classified based on hemodynamics (high flow or low flow), pathogenesis (spontaneous or traumatic), and angiographic anatomy (direct or indirect).1–3 The angiographic classification by Barrow et al. divides CCFs into 4 types (A, B, C, and D). Type A CCFs are direct connections between the cavernous ICA and the cavernous sinus. Types B, C, and D CCFs are indirect connections between the cavernous sinus and branches of the ICA (type B), branches of the external carotid artery (type C), and branches of the ICA and external carotid artery (type D).1

Type A, or direct CCFs, are the most frequent type of CCF.4 Trauma is the most common cause of direct CCFs and accounts for the higher prevalence in young men.1–4 Traumatic CCFs are usually unilateral but can occur bilaterally in 2% of patients.1–4 Less frequently, spontaneous direct CCFs occur because of rupture of a cavernous carotid aneurysm (as in our patient) or rupture of a weakened carotid wall in patients with connective tissue diseases such as fibromuscular dysplasia or Ehlers–Danlos syndrome.3,4 Iatrogenic causes of type A CCFs include transsphenoidal surgery and endovascular procedures.1 The majority of type A CCFs are high-flow lesions, with acute onset of symptoms and low likelihood of resolving spontaneously.1,4

Type B, C, and D CCFs are indirect or dural fistulas. Indirect CCFs are more often spontaneous and infrequently traumatic.5–5 They are more common in elderly women.1–5 The precise spontaneous mechanism is unknown but may be related to venous thrombosis in the cavernous sinus or weakening of the arterial wall from predisposing factors such as
hypertension, atherosclerosis, or connective tissue diseases.\textsuperscript{3–5} Indirect CCFs are low flow lesions, with insidious symptom onset and high likelihood of spontaneous resolution.\textsuperscript{3–6}

In CCFs, arterialized blood is shunted into the venous system leading to venous hypertension.\textsuperscript{3,4} The clinical symptoms depend in large part on the venous drainage route and available collaterals.\textsuperscript{3–4} Patients with direct CCFs classically present with sudden onset of the clinical triad of chemosis, proptosis, and bruit.\textsuperscript{3,4} Other common clinical findings include headache, diplopia, ophthalmoplegia, and decreased visual acuity.\textsuperscript{2–4,6} Less common findings include intracerebral hemorrhage, subarachnoid hemorrhage, and external hemorrhage, such as epistaxis.\textsuperscript{2–6} Patients with indirect CCFs have a more insidious onset of symptoms, and the clinical course is often relapsing remitting, making diagnosis difficult.\textsuperscript{2–4} Chronic conjunctival injection, proptosis, glaucoma, and chemosis are common.\textsuperscript{2–4,6}

The diagnosis of CCF is made by neuroanatomic and neurovascular imaging. First-line modalities include computed tomography and computed tomography angiography or magnetic resonance imaging and magnetic resonance angiography of the brain. Signs of CCF detected with noninvasive imaging include proptosis, dilatation of the SOV, extraocular muscle enlargement, ipsilateral cavernous sinus enlargement, and skull fracture.\textsuperscript{2–4,6} Digital subtraction angiography is the gold standard in the diagnosis of CCF and must be performed before any potential intervention.\textsuperscript{3,4}

The main goal in the treatment of CCFs is to preserve flow in the ICA while occluding the fistula.\textsuperscript{2,6} Clinical indications for emergent treatment include progressive ptosis, visual decline, hemorrhage (intracranial or external), and increased intracranial pressure.\textsuperscript{7} Angiographic signs for emergent treatment include cavernous sinus varix, pseudoaneurysm, cortical venous drainage, and thrombosis of distal venous pathways.\textsuperscript{7}

Conservative management is an option for some low risk, low flow, indirect CCFs. External manual compression of the ipsilateral carotid artery and contralateral jugular vein, several times per day over several weeks, achieves cure in ≈30% of patients.\textsuperscript{5,8} Surgical management is limited to cases when endovascular approaches are not possible or not effective because of the increased morbidity associated with the open management.\textsuperscript{2–4}

Endovascular management is the current treatment modality of choice for CCFs.\textsuperscript{3–4} Transarterial embolization is the preferred access method for direct CCFs and transvenous embolization is preferred for indirect CCFs.\textsuperscript{4} In the transarterial approach, a microcatheter is passed through the fistula into the cavernous sinus and coils and/or embolic agents, such as Onyx (Covidien, Ireland), are placed in the cavernous sinus.\textsuperscript{4} In the transvenous approach, the cavernous sinus is most commonly accessed through the inferior petrosal sinus.\textsuperscript{4}
Direct cannulation of the SOV is occasionally needed when more standard approaches are not possible. Cure rates after endovascular repair of CCFs approximate 80%. In summary, CCFs are a rare but treatable cause of orbital injury and vision loss. Endovascular embolization of CCFs with coiling or liquid agents is the treatment modality of choice. With appropriate treatment, progressive resolution of symptoms is expected in most patients.

**TAKE-HOME POINTS**

- Carotid cavernous fistulas (CCFs) are a rare but potentially devastating cause of orbital symptoms, visual loss, and periocular disfigurement.
- Carotid cavernous fistula patients typically present with proptosis, elevated intraocular pressure, prominent tortuous conjunctival vessels, and sometimes headache.
- Endovascular treatment is the modality of choice for carotid cavernous fistulas. Prompt treatment can prevent further complications of carotid cavernous fistulas and lead to complete resolution of symptoms.

**Disclosures**

None.

**References**


**Key Words:** aneurysm ■ atrial fibrillation ■ carotid artery ■ fistula ■ stroke
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