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Carotid cavernous fistula: A rare but treatable cause of ophthalmoplegia - A case report

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Abstract:

Carotid cavernous fistulas (CCFs) are a rare but debilitating entity that may present with orbital or cerebral venous hypertension. CCFs may pose diagnostic and management pitfalls for clinicians as they can initially be misdiagnosed as primary orbital pathology or nonarteriovenous shunting-related cavernous sinus pathology. Furthermore, the resolution of pulsatile tinnitus could be an ominous sign in patients with untreated dural arteriovenous fistula. We describe a case of a 56-year-old male who presented with progressive right eye proptosis, congestion, decreased visual acuity, limited duction, exophthalmos, and pulsatile tinnitus. The patient had poor response to antibiotics and steroids. Magnetic resonance imaging brain showed significant inflammation involving the right orbit and atypical enhancement of the basal frontal lobe adjacent to the orbit. Cerebral angiography revealed an indirect right CCF and right sigmoid sinus thrombosis with stenosis of the right internal jugular vein. No clear predisposing factor was identified. Given the rapidly progressive nature of the condition, the patient successfully underwent endovascular treatment with transvenous approach to preserve flow in the internal carotid artery while ensuring occlusion of the fistula. A triad of proptosis, eye congestion, and signs of turbulent flow such as tinnitus or orbital bruit should raise suspicion for CCF. An interesting feature in this patient is that CCF may have occurred secondary to sigmoid sinus thrombosis with accompanying small cortical vein drainage. Our case highlights the importance of early recognition and timely intervention to ensure the resolution of orbital hypertension-related symptoms in rare cases of CCFs.

Keywords:

Carotid cavernous fistula, embolization, ophthalmoplegia

Introduction

Carotid cavernous fistulas (CCFs) are rare vascular shunts from the carotid artery to the cavernous sinus. Patients with a CCF may have a rapidly progressive presentation with unique challenges and necessitate evaluation with neurovascular imaging for classification and diagnosis. Clinicians should maintain high vigilance for a triad of proptosis, eye congestion, and signs of turbulent flow to diagnose this entity. Modern endovascular techniques offer a viable option for obliteration of CCFs with low morbidity and mortality. Navigation is typically achieved through the inferior petrosal sinus or facial and superior ophthalmic vein. Here, we describe a challenging case of carotid cavernous fistula that was successfully treated through occluded right inferior petrosal sinus access using coil and Onyx embolic material.

Case Report

A 56-year-old male with no past medical history presented to the hospital with right eye proptosis. He experienced right-sided pulse-synchronous pulsatile tinnitus a few weeks prior which was followed

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by congestion of the right eye. His eye symptoms progressively worsened with mild decrease in acuity of vision (20/40) in setting of elevated intraocular pressure, limited duction, and exophthalmos which correlated with the resolution of pulsatile tinnitus. There was no optic disc swelling. On initial outpatient ophthalmology evaluation, he received antibiotics and steroids without significant improvement. During hospitalization, systemic infection and recent trauma were ruled out. There were no laboratory or coagulation abnormalities. Magnetic resonance imaging (MRI) demonstrated significant inflammation involving the right orbit and atypical enhancement of the basal frontal lobe adjacent to the orbit [Figure 1]. Magnetic resonance angiography (MRA) with time-of-flight demonstrated an enlarged superior ophthalmic vein, cavernous sinus, and enlarged frontal vein [Figures 2 and 3]. At this point, the differential diagnosis was broad and encompassed orbital infection and cavernous sinus thrombosis.

Considering the involvement of the cavernous sinus and subtle flow voids noted in the posterosuperior pocket of the right cavernous sinus, a conventional cerebral angiogram was performed. Evaluation with angiography revealed an indirect right carotid cavernous fistula (CCF) and right sigmoid sinus thrombosis with stenosis of the right internal jugular vein. Figure 4 illustrates the basal frontal vein, superior ophthalmic vein, and occluded inferior petrosal sinus on digital subtraction angiography before embolization. The fistula was successfully treated with transvenous approach through the right inferior petrosal sinus using a coil and Onyx liquid embolic agent [Figure 5]. After treatment, the fistula was angiographically cured and the patient had remarkable improvement in his symptoms. He had no evidence of right eye congestion, exophthalmos, elevated intraocular pressure, or vision changes at this point. MRI since his discharge noted improvement in edema involving

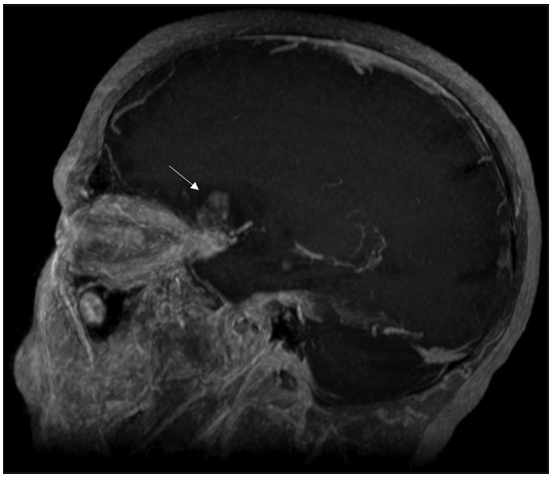

Figure 1: Magnetic resonance imaging demonstrating frontal basal lobe edema (arrow) and congestion

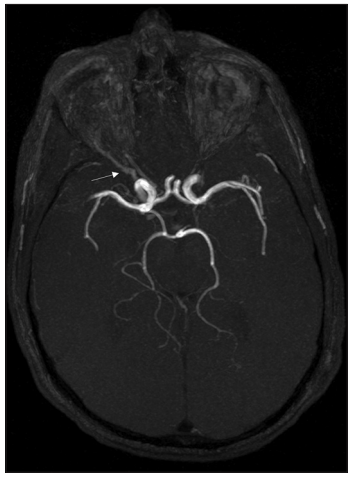

Figure 2: Time-of-flight magnetic resonance angiography with carotid cavernous fistula demonstrating enlarged superior ophthalmic vein (arrow), right internal carotid artery, and cavernous sinus

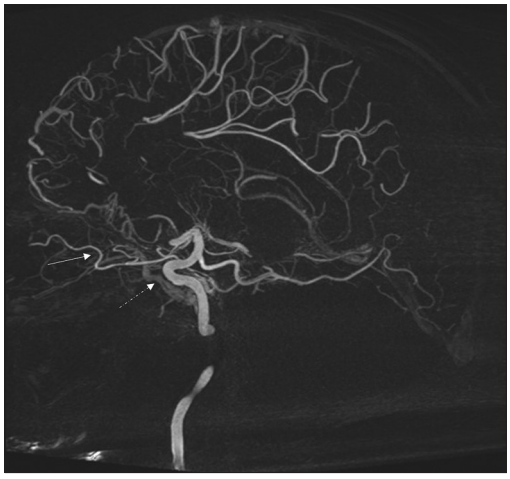

Figure 3: Magnetic resonance angiography with carotid–cavernous fistula, superior ophthalmic vein (solid arrow), cavernous sinus (dashed arrow), and enlarged frontal vein corresponding to the area of cortical vein on MRI. MRI: Magnetic resonance imaging

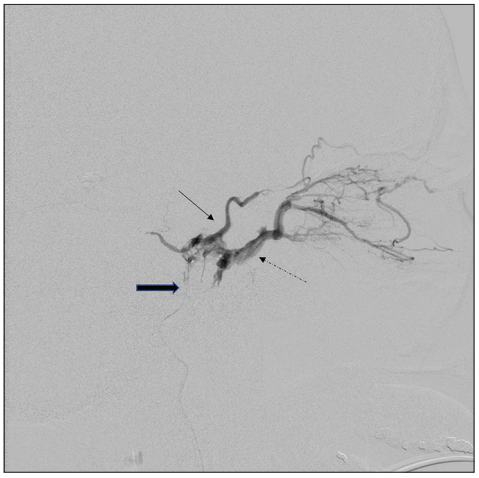

Figure 4: Digital subtraction angiography before embolization demonstrating basal frontal vein (solid arrow), superior ophthalmic vein (dashed arrow), and inferior petrosal sinus (block arrow) with no drainage due to occlusion

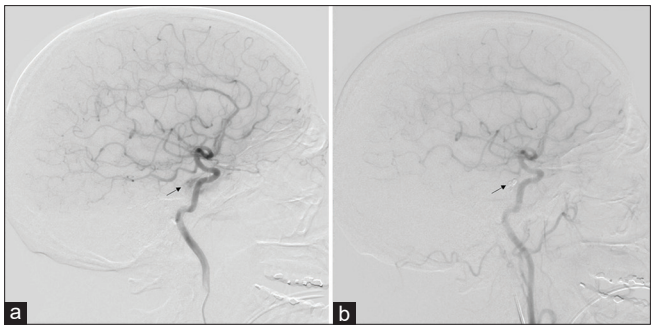

Figure 5: Digital subtraction angiography (a) lateral view of the right internal carotid artery run with early opacification of the right cavernous sinus. (b) lateral view of the right internal carotid artery runs post-Onyx and coil

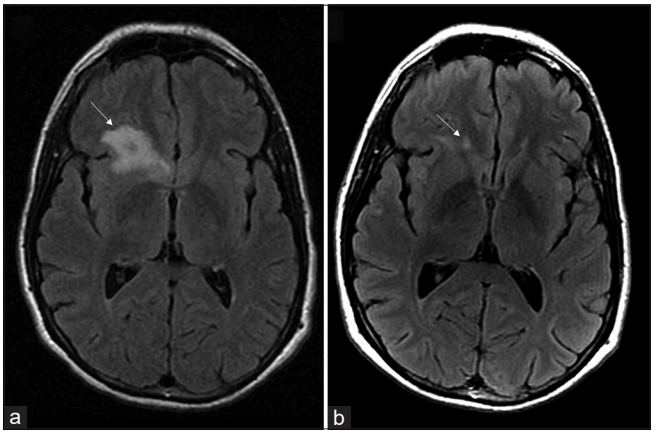

Figure 6: Magnetic resonance imaging FLAIR images showing (a) edema from venous congestion and (b) significant improvement after treatment. FLAIR: Fluid-attenuated inversion recovery

the right basal frontal lobe with complete resolution of orbital findings [Figure 6]. There was also interval improvement of thrombus burden involving the right sigmoid sinus which was treated with dabigatran.

Discussion

We discuss a complex case of CCF that presented a diagnostic challenge as the patient presented with not only orbital signs and ophthalmoplegia but also had an enhancing lesion involving the right basal frontal lobe. Orbital venous hypertension is not uncommon; however, a small cortical vein directly draining the right basal frontal lobe contributed to the appearance of an enhancing lesion, thus posing an initial diagnostic challenge. Our patient had concurrent occlusion of the inferior petrosal sinus and thrombus in the sigmoid sinus which likely contributed to the reversal of fistulous flow toward the superior ophthalmic vein and basal frontal lobe. The interesting feature, in this case, is that CCF may have occurred secondary to sigmoid sinus thrombosis. The enhancement is likely from abnormal dilatation of the venous system causing venous congestion from cortical reflux. Since the resolution of symptoms is expected with treatment, early recognition, and timely intervention is paramount in this population yet pose a challenge given the uncommon and rapidly progressive nature of the condition.

Classification and pathogenesis

A CCF is an abnormal communication between arteries and veins within the cavernous sinus. Barrow *et al*. classified CCFs into four subtypes: direct fistulas (Type A) and indirect or dural fistulas (Types B, C, and D).[1] Type A fistulas are direct high-flow shunts between the internal carotid artery (ICA) and cavernous sinus. Low-flow fistulas include indirect connections between the cavernous sinus and meningeal branches of the ICA (Type B), meningeal branches of the external carotid artery (Type C), and meningeal branches of both internal and external carotid arteries (Type D).[2] Etiology includes spontaneous or secondary causes such as trauma, carotid–cavernous aneurysm, venous thrombosis, genetic conditions with arterial wall defects, and iatrogenic interventions such as transsphenoidal surgery and endovascular procedures.[3,4] Indirect fistulas are typically insidious in onset and more common among elderly women. The exact pathogenesis of dural CCFs is unknown but postulated to involve primary venous thrombosis in the cavernous sinus or arterial wall weakening from risk factors such as hypertension, atherosclerosis, and connective tissue diseases such as Ehlers–Danlos syndrome or fibromuscular dysplasia.[4,5]

Clinical features

As arterialized blood is shunted to the venous system, venous hypertension develops, and clinical findings manifest based on the venous drainage route and collateral circulation.[3,4] The most common presenting symptoms of CCFs include proptosis (72%–98%), chemosis (55%–100%), orbital bruit (71%–80%), and headache (25%–84%).[2] Patients also report visual disturbances such as diplopia (88%), blurry vision, orbital pain, ophthalmoplegia (23%–63%), and cranial nerve deficits (17%–44%).[2,6] Less common clinical findings include epistaxis, subarachnoid hemorrhage, and intracerebral hemorrhage in 5% of patients.[6] Our patient demonstrated proptosis, pulsatile tinnitus, orbital pain, visual changes, and cranial neuropathy. Past cases report pulsatile tinnitus as an unusual clinical finding in conjunction with normal otoscopic examination which indicates the necessity of further workup in this population.[7]

Diagnostic approach

The investigation may begin with first-line imaging modalities such as computed tomography (CT) and CT angiography or MRI or MRA of the brain. Signs of CCF

on neuroanatomic and neurovascular imaging include proptosis, expansion of cavernous sinus and superior ophthalmic vein, extraocular muscle enlargement, associated skull fractures, and abnormal cavernous sinus flow void on MRI.[8] Our patient also had enhancement of the basal frontal lobe adjacent to the orbit which is an atypical finding for CCF. Based on the anatomical correlation, it was likely related to venous infarction. Transcranial Doppler ultrasonography typically demonstrates increased blood flow velocity and decreased pulsatility index in the carotid siphon of CCF patients.[9] Ultimately, conventional digital subtraction angiography is the gold standard test for CCF diagnosis and guides intervention.

This is an example of a complex case that initially had a broad differential diagnosis which included orbital infection and cavernous sinus thrombosis. Due to collaboration within our multidisciplinary team involving neurology, neurosurgery, radiology, infectious disease, ophthalmology, and interventional neuroradiology, we were able to reach an accurate diagnosis.

Management

The optimal approach is early recognition and timely intervention to ensure occlusion of the fistula. Typically, direct CCFs are unlikely to close spontaneously and require closure, whereas indirect CCFs may resolve spontaneously. Indications for emergent treatment include decline in visual acuity, progressive ptosis, ophthalmoplegia, intracranial or external hemorrhage, and elevated intracranial and intraocular pressure.[10] For low-risk indirect CCFs, conservative management and close follow-up with serial examinations may be considered. Ultimately, endovascular obliteration is the preferred modality. Transarterial approach is employed for direct CCFs, whereas transvenous embolization is used for indirect CCFs. The rationale for the transvenous approach is a higher occlusion rate and lower recurrence compared to the transarterial route. If endovascular management is not possible or ineffective, surgical treatment may be pursued. Manual vascular compression of the ipsilateral carotid artery and contralateral jugular vein has also demonstrated some utility in closing dural CCFs.[6]

In this case, our patient successfully underwent embolization of the right CCF with transvenous approach. Typically, navigation is achieved through the inferior petrosal sinus or facial and superior ophthalmic vein.[2] A microcatheter was advanced to the origin of the intercavernous sinus but was not able to cross due to exceptionally small channels connecting both cavernous sinuses. The right internal jugular vein was subsequently identified and contrast injection demonstrated occlusion of the right sigmoid sinus due to near complete occlusive thrombus in addition to occlusion of the major channels of the inferior petrosal sinus. To navigate this occluded sinus, the microcatheter was blindly advanced into the inferior petrosal sinus while maintaining the symmetry from contralateral inferior petrosal sinus trajectory. We were able to reach the fistulous point in the posterior medial aspect of the right cavernous sinus when coils were subsequently deployed and the fistula was noted to be occluded. Onyx was injected through the microcatheter to avoid recanalization of the fistula.

Review of transvenous approach

Texakalidis *et al*. conducted a meta-analysis of patients that underwent treatment of CCF with various modalities.[11] With the transvenous approach for indirect CCFs, there was complete obliteration in the majority (86.03%) of cases with 87.54% occlusion rate.[11] Various routes of transvenous access may be employed. In cases of thrombosed sinus, transvenous access may be achieved by crossing or recanalization of a closed venous segment.[12] For instance, it is possible to traverse an ipsilateral occluded inferior petrosal sinus to access a trapped fistulous pouch of the cavernous sinus or traverse an occluded sigmoid sinus to approach an isolated transverse sinus.[13,14] However, the risk of catheter or wire perforation of the thrombosed sinus must be carefully managed.[14]

Prognosis

The prognosis of CCF is variable and follows a spectrum of outcomes based on clinical deficits. After complete closure of CCF, symptoms such as proptosis, chemosis, and increased intraocular pressure generally resolve within hours to days, whereas cranial nerve palsies may take several weeks to resolve.[2] Visual recovery is primarily dependent on pathogenesis, severity, and duration before the procedure.[2] While recanalization is uncommon and the majority experience no recurrences after the intervention, patients can be treated by repeat embolization.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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