Case Report

Coiling and liquid embolization of a carotid-cavernous fistula

ABSTRACT

A carotid-cavernous fistula (CCF) results from an abnormal vascular shunt between the carotid artery and the venous channels of the cavernous sinus. The clinical presentation varies based on the involved neurovascular structures, the anatomy of the shunt, the underlying cause, and the hemodynamics of the CCF. In this case report, we describe a 46-year-old man who presented with recurrent headaches, redness, bulging, and a rapid decline in vision in his right eye, along with diplopia. Brain magnetic resonance imaging revealed dilation of the superior ophthalmic vein. A subsequent digital subtraction angiogram confirmed a Barrow classification Type D (indirect) CCF. The patient underwent endovascular therapy involving combined coil and Onyx™ embolization (a mixture of ethylene-vinyl alcohol copolymer, dimethyl-sulfoxide, and micronized tantalum powder from Medtronic, USA) intervention resulting in an exceptional angiographic and clinical outcome. The patient became entirely symptom-free within 2 weeks following the treatment.

Keywords: Carotid-cavernous fistula, cavernous sinus, coil embolization, digital subtraction angiogram, endovascular therapy, Onyx embolization, transvenous approach

INTRODUCTION

A carotid-cavernous fistula (CCF) arises due to an abnormal vascular connection between the carotid artery and the venous channels within the cavernous sinus (CS).[1] The clinical manifestations vary depending on the specific involvement of neurovascular structures within the sinus.[2] Left untreated, these fistulas can lead to potentially devastating consequences such as vision loss, subarachnoid hemorrhage, intracerebral hematoma, and progressive proptosis.[3] Spontaneous occurrences of CCFs are rare, and their precise incidence remains unknown.[2] Within our report, we highlight a case involving a spontaneous Type D (indirect) CCF. This case was successfully managed through transvenous endovascular therapy (ET).

CASE REPORT

The patient is a 46-year-old male, a known diabetic on regular treatment with good compliance, maintaining a nonsmoking, nondrinking lifestyle. He had no history of prior head or faciomaxillary trauma. Three weeks before seeking medical attention, the patient started experiencing headaches and observed redness in his right eye. Initially, he received eye drops (antibiotics, steroids, and artificial tears) and analgesics from his local practitioner. However, even after 2 weeks of this treatment, there was no noticeable improvement in his symptoms. However, a sudden onset of bulging accompanied by rapidly declining vision in the right eye and double vision

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occurred 1 week before the presentation [Figure 1]. The patient promptly sought care from a nearby ophthalmologist, who conducted a comprehensive ocular examination. Examination of the left eye showed slight chemosis, conjunctival injection, a 3 mm pupil with intact light reflexes, and a visual acuity of 6/24. On examination of the right eye, pronounced conjunctival injection and chemosis, a 3.5 mm pupil with intact direct and indirect light reflexes, and nonaxial proptosis with restricted ocular movements – particularly abduction – were noted. The visual acuity in the right eye was measured at 6/60. Tono-Pen tonometry indicated increased intraocular pressure in the right eye. Fundoscopy revealed papilledema and retinal venous engorgement in the right eye. Considering the clinical presentation and ocular findings, a preliminary diagnosis of a CCF was suspected. Subsequent brain magnetic resonance imaging (MRI) depicted significant enlargement of the right superior ophthalmic vein (SOV) [Figure 2]. Consequently, the patient was transferred to the neurovascular intervention department for further management. A digital subtraction angiogram (DSA) was performed, revealing a Type D CCF with an early filling of the right CS in the arterial phase [Figure 3]. Given the rapidly deteriorating clinical condition, shunt anatomy, and hemodynamics, the decision was made to embolize the CCF.

Procedure

The patient was placed supine on the angiographic table. The patient was intubated, and the procedure was performed under general anesthesia following a strict aseptic protocol. A simultaneous right transfemoral venous and left transfemoral arterial access was taken. A 5F diagnostic catheter from the left groin puncture was placed at the common carotid artery. A 5F diagnostic catheter was navigated from the right transvenous groin access through the inferior vena cava, right atrium, superior vena cava, and jugular vein. At the level of the jugular sigmoid junction, a microcatheter was maneuvered into the inferior petrosal sinus (IPS), leading to the CS. After cannulating the CS, an angiogram was performed to confirm the position of the microcatheter. Embolization was done with Onyx on the left side and coils on the right side of the CS [Figure 4]. The final angiogram demonstrated occlusion of the CCF with arterial branches within the normal limits [Figure 5]. The procedure was completed and the patient was extubated without any new neurological deficit. A total of five coils and 2 ml of Onyx were used to achieve complete embolization.

There was a significant improvement in the vision, diplopia, and redness in both eyes within 48 h postprocedure [Figure 6]. The patient was discharged on the 3rd day postoperatively. At his 2nd week follow-up, the patient's ocular redness, proptosis, headaches, and visual disturbances have completely resolved.

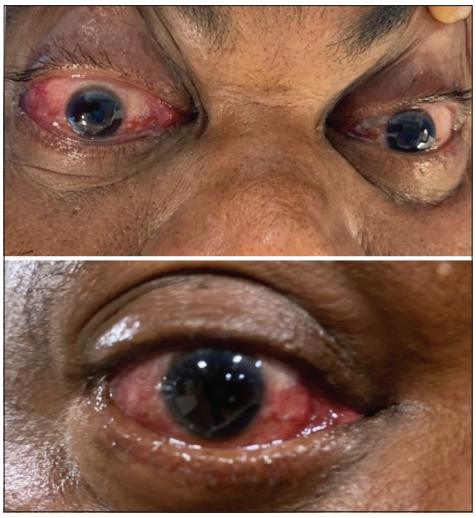

Figure 1: Clinical image showing chemosis, conjunctival injection, and slight proptosis of the right eye

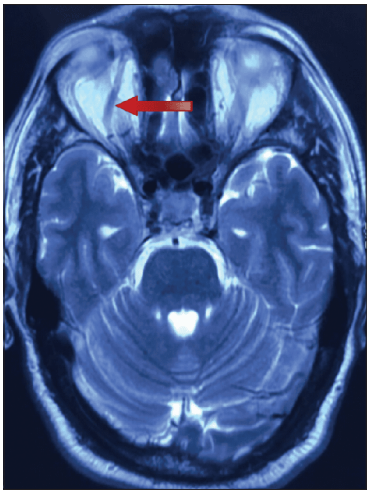

Figure 2: Brain magnetic resonance imaging, axial view, demonstrating a dilated superior ophthalmic vein (red arrow)

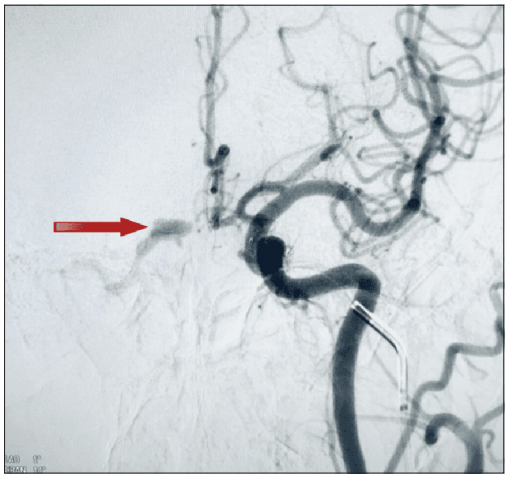

Figure 3: Digital subtraction cerebral angiogram revealing a carotid-cavernous fistula with an early filling of the right cavernous sinus in the arterial phase. The red arrow indicates the early filling of the cavernous sinus in the arterial phase (due to the carotid-cavernous fistula)

DISCUSSION

CCFs can be categorized according to several factors: shunt anatomy (direct vs. indirect), etiology (traumatic vs. spontaneous), and hemodynamics (high flow vs. low flow).[2] Barrow *et al*. established a classification system for CCFs based on the involved arterial system [Table 1]. Type A CCFs represent direct fistulas, while types B, C, and D are categorized as indirect.[4] Indirect CCFs are considered low-flow variants because they originate from dural branches rather than the high-flow internal carotid artery (ICA).[2] In 2015, Thomas *et al*. proposed an updated classification system based on venous drainage [Table 2].[5]

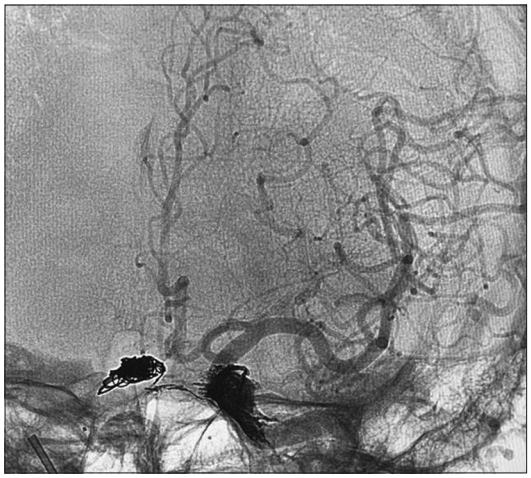

Figure 4: Digital subtraction angiogram cerebral angiogram showing the placement of coils on the right side and Onyx embolization on the left side

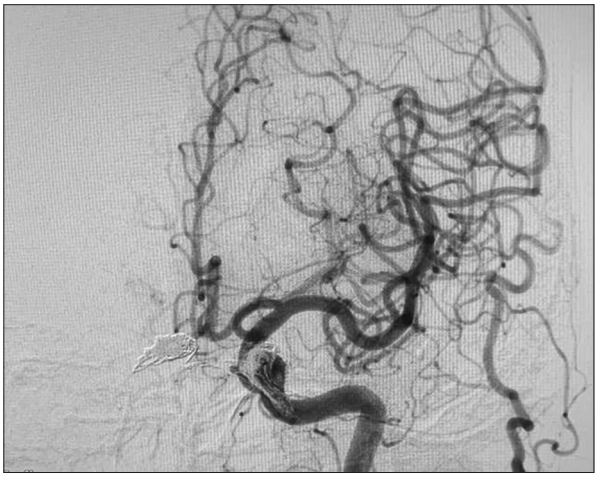

Figure 5: The final check angiogram demonstrating occlusion of the carotid-cavernous fistula with normal arterial flow

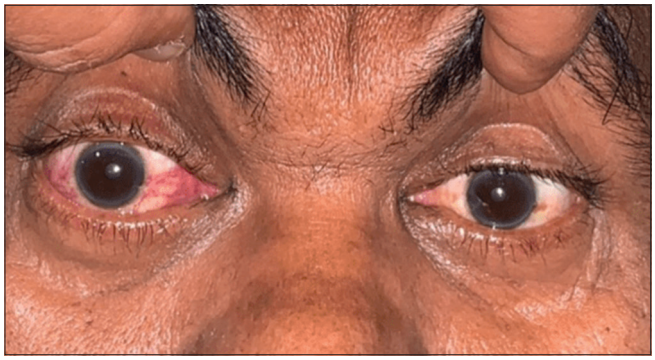

Figure 6: Clinical picture 48 h postoperatively showing a noticeable reduction of the right eye redness within 48 h postprocedure

CCFs occur when high-pressure blood from the arterial system (ICA or external carotid artery) is shunted into a low-pressure venous system, specifically the CS, without an intervening capillary bed to reduce the pressure. This pressure disparity and vascular resistance obstruct venous drainage, causing congestion in the areas drained by the CS.[6] Ophthalmic manifestations, commonly observed, may take several days to weeks to develop as venous hypertension needs to reach critical levels before becoming apparent.[7]

The classical clinical triad consists of chemosis, proptosis (pulsatile exophthalmos), and ocular bruit.[8] They are less frequent in indirect CCFs. Other observable features include headache, pain (ocular/periorbital), proptosis, cephalic bruit, pulsatile tinnitus, loss of visual acuity, raised intraocular pressure, secondary glaucoma, ophthalmoplegia, trigeminal nerve dysfunction, anhydrosis, otorrhagia, and epistaxis.[7,9,10] In our patient, his initial atypical and seemingly mild presentation might have led to the erroneous diagnosis and treatment earlier on. Conjunctivitis, nonspecific orbital inflammation, orbital/retrobulbar hemorrhage, orbital infection, Grave's ophthalmopathy, orbital tumor, tumor with CS involvement, orbital vasculitis, orbital apex syndrome, CS thrombosis, and

Table 1: Barrow classification

Type	Description
A	Direct connection between the ICA and CS
B	Connection between meningeal branches of ICA and CS
C	Connection between meningeal branches of ECA and CS
D	Connection between meningeal branches of both ICA and ECA and the CS

CS: Cavernous sinus, ECA: External carotid artery, ICA: Internal carotid artery

Table 2: Thomas classification of carotid-cavernous fistula 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-CS fistulae corresponding to the type of barrow classification

CS: Cavernous sinus, ICA: Internal carotid artery

superior orbital fissure syndrome are some of the probable differential diagnoses for a case of CCF.[9]

Ocular Doppler ultrasound can be a helpful screening tool. It can identify the dilation and flow velocities within the SOV, as well as the enlargement of the extraocular muscles.[11] A plain computed tomography (CT) is more helpful in CCF cases with a history of trauma as it is more sensitive in detecting basilar skull fractures. CT with contrast can reveal SOV dilation and proptosis. An MRI is superior to CT in delineating abnormal flow voids or orbital edema.[12] CT angiogram and magnetic resonance angiogram (MRA) are the first-line noninvasive imaging modalities in evaluating CCFs, particularly those with visual symptoms.[1,13] However, in our case, an MRA was not performed and we directly opted for a DSA. Although invasive, DSA is the gold standard imaging modality. It can identify the location, arterial supply, venous drainage, and flow rate to classify the CCF and help plan for potential ET strategies.[11,14]

Endovascular intervention is widely considered the first-line treatment of CCFs. Recent technological advances have increased the number of safe and viable treatment options, with a cure rate of well over 80%.[2] Surgical intervention is the most invasive option available. Success rates range from 31% to 79% and depend on the method, approach, and skill of the operator.[1] Surgery is associated with higher perioperative risks, residual fistulous communications, and complications like cranial nerve palsies. Nowadays, it is primarily indicated as salvage for failed ET.[7] Awaiting spontaneous closure, compression treatment or stereotactic radiosurgery are other viable alternatives for select cases with low-flow indirect fistulas.[1,2,7,15] However, these treatments are associated with a considerable time lag for the resolution of symptoms, which excludes them as an option for emergencies with rapid deterioration, as with our patients.

Transvenous procedures are the preferred treatment approach for indirect fistulas because of their simplicity, higher success rates, lower ischemic risk, and capability to cure the fistula in a single session. In the transvenous approach, the abnormal CS is super selectively catheterized and the fistula is occluded without rerouting venous drainage to the cortical structures.[16] IPS is the most commonly used venous pathway for cannulation of the CS. Less commonly, in some technically inaccessible cases, the anterior approach through the SOV through the facial vein lateral pterygoid plexus, superior petrosal sinus, cortical veins, the inferior ophthalmic vein, or the contralateral IPS or SOV with access into the ipsilateral CS through the circular sinus.[7,16]

After successful cannulation of the CS, embolization can be achieved with materials such as coils, n-butyl 2-cyanoacrylate, and ethylene-vinyl alcohol copolymer (EVOH), either alone or in combination. We used a combination of coils and EVOH. Coils are radiopaque, easy to use, and can be redeployed or removed if the initial placement is not optimal. However, adequate volumetric packing or complete occlusion is not always achievable. Concomitant use of EVOH has the ability of mechanical occlusion without vessel wall adhesion. Its nonadhesive nature decreases the risk of microcatheter retention and allows a slow single injection of the embolic agent with concomitant angiogram checks.[16,17]

Successful closure of the CCF results in rapid elimination of the vascular pressure head and alleviates the ocular manifestations produced by the venous congestion.

CONCLUSION

CCF is a rare vascular phenomenon which is capable of atypical presentations, compounding the complexity of its management. A high index of suspicion is necessary for prompt diagnosis and appropriate therapy, which is vital for achieving the best possible visual and neurological outcomes.

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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Conflicts of interest

There are no conflicts of interest.

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