Internal carotid cavernous fistula: One case report and literature review

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ABSTRACT

Spontaneous carotid cavernous fistula is usually caused by lesions in the internal carotid cavernous sinus and its branches. We report a young male patient with no history of trauma, in whom the fistula was located at the cavernous sinus of the internal carotid artery. The post-operative angiography found a pseudoaneurysm at the origin of the cavernous sinus of internal carotid artery, which suggested that cavernous sinus fistulas were caused by bleeding from rupture of thin-walled vessels due to the local abnormalities of the vascular wall.

Key words: headache, internal carotid artery, internal carotid cavernous fistula, male, ophthalmoplegia, pulsating exophthalmos, spontaneous

INTRODUCTION

Internal carotid cavernous fistula (CCF) can be divided into spontaneous CCF and traumatic CCF according to the etiology. Traumatic CCFs account for 75-85%, with causes including basal skull fracture and laceration, bony fragment puncture, penetrating injury by foreign body and fire arm injury. About 25% of the cases are spontaneous CCFs, which are usually caused by lesions in the internal carotid cavernous sinus and its branches, with slow progression. Spontaneous CCFs are relatively rare in the clinic, including CCFs induced by ruptured aneurysm, arteritis, atherosclerosis and spontaneous CCF during pregnancy. In this article, we report a case with spontaneous CCF and review the related literatures.

CASE REPORT

A 25-year-old male patient was admitted to our hospital on March 26, 2010. The chief complaint was congestion of the left eye for 2.5 months and left eye proptosis complicated by left ear tinnitus for 2 months. The patient suffered left eye congestion without obvious incentives for 2.5 months, with absence of eye itching, foreign

body sensation, blurred vision or ocular motility disorder, and was diagnosed as left eye conjunctivitis and treated at a local hospital; however, no improvement was found after treatment. Symptoms developed 2 months ago with left eye proptosis complicated by persistent blow-drying tinnitus in the left ear, which was considered as cerebrovascular disease at the local hospital; then, the patient came to our hospital for further diagnosis and treatment. Physical examination showed left eye proptosis, conjunctival congestion [Figure 1], vascular murmur in the eye and orbital region, clear and light-red boundary of papilla optica in the fundus of the left eye, A:V = 2:5 and normal results from all the other examinations of the nerve system. A magnetic resonance imaging (MRI) of the orbit showed asymmetrical eyeballs, left eye proptosis, expansion of the left cavernous sinus with an internal flow void and significantly thickened and tortuous dilated veins in the left eye with a flow void [Figure 2]. Digital subtraction angiography (DSA) showed left internal CCF. Results showed obscure artery imaging in the left brain and no imaging was found in the left anterior brain; thickened and tortuous veins were presented by early imaging of the eyes

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[Figure 3a] and thickened draining veins were shown by early imaging of the cavernous sinus; for patency of the communicating artery after left vertebral artery imaging [Figure 3b], artery imaging in the left brain was visible, and was even clearer after compression of the left internal carotid artery. The patient was diagnosed as left internal CCF (spontaneous). Balloon occlusion surgery was performed and the post-operative angiography showed a clear imaging of the left internal carotid artery and no deletion of distal vessels [Figure 3c], a cystic protrusion with smooth and sharp edges was observed at the origin of the left internal carotid cavernous sinus with a size of about 6 mm × 8 mm. The right internal carotid artery and the left vertebral artery imaging showed compensation of the anterior communicating artery, and the posterior

communicating artery disappeared [Figure 3d]. Symptoms were improved after surgery, including the disappearance of tinnitus and vascular murmur in the orbital region, significant alleviation of ocular and conjunctival congestion and retinal arteriovenous ratio returning to 2:3. The patient had transient headache

Figure 1: Left eye congestion and slight left eye proptosis

Figure 2: Thickened left ophthalmic vein shown by magnetic resonance imaging

Figure 3: a) Thickened and tortuous ophthalmic veins were observed before surgery, with obscure imaging of the internal carotid artery and distal vessels; b) Compensatory hyperplasia of the left vertebral artery could be seen before surgery, which provided blood supply for the left middle cerebral artery; c) Thickened draining vein disappeared after surgery, with clear imaging of the internal carotid artery and distal vessels; d) Compensatory blood flow disappeared after surgery

after surgery, which disappeared after symptomatic treatment.

DISCUSSION

The classification of CCF is based on anatomy and hemodynamics. Barrow *et al.*^[1] divided CCF into four types according to the fistula location and blood flow: Type A is internal CCF, and most cases have one high-flow fistula, no blood supply from the external carotid arteries; type B is cavernous fistula in the meningeal branch of the internal carotid artery, with several low-flow fistulas in most cases; type C is cavernous fistula in the meningeal branch of the external carotid artery; type D is a cavernous fistula in the meningeal branch of the external carotid artery with several low-flow fistulas. The case of this patient was spontaneous CCF with no history of trauma, and the fistula was located at the cavernous sinus of the internal carotid artery. Post-operative angiography found a pseudoaneurysm at the origin of the cavernous sinus of the internal carotid artery, which suggested that the cavernous sinus fistulas were caused by bleeding from rupture of thin-walled vessels due to the local abnormalities of the vascular wall.

The anatomy of the cavernous sinus and its relationship with draining veins are the anatomical basis of the clinical manifestations. Cavernous sinus mainly has three venous lacunae known as internal lacuna, anterior inferior lacuna and posterior superior lacuna, which are named according to their relationship with the internal carotid artery. The three lacunae are separated by fibers and connected by split holes. Of the three lacunae in the cavernous sinus, the anterior inferior lacuna is usually unobstructed. The main draining vein of CCF is the ophthalmic vein, especially the superior ophthalmic vein. In normal conditions, the cavernous sinus receives blood return via the superior and inferior ophthalmic veins (mainly the superior ophthalmic veins, as the inferior ophthalmic veins are quite small), sphenoid sinus, lateral fissure veins and basilar venous plexus, and drains into the inferior and superior petrosal sinuses, basal veins and converging via dural veins in the pterygoid plexus. Clinical manifestations of CCF are mainly correlated with the increased pressure in cavernous sinus and the direction of venous return. In these cases, most patients first visit the ophthalmology department because of the eye symptoms in addition to headache, and the symptoms are as follows:

1. Pulsating exophthalmos, chemosis, tortuosity and thickening of the blood vessels in the sclera. Arterial blood that fills into the cavernous sinus leads to the increase of pressure in the sinus, resulting in the reverse drainage in one direction or multiple directions, which is the most significant in the superior ophthalmic

vein of the anterior cavernous sinus. Blood backflow finally leads to tortuous expansion of the ophthalmic veins, tissue edema, increased intraorbital pressure and exophthalmus;

2. Nystagmus, murmur from head and orbital region. It originates from the impact on the periosteum by influx of arterial blood into the cavernous sinus. Patients are conscious of sustained intracranial murmur and murmurs could be heard by ocular and periorbital auscultation, which are consistent with pulse, as well as a concomitant nystagmus;
3. Ocular motility disorder and diplopia. As a result of the increased pressure in the cavernous sinus, various degrees of damage could be caused due to the disturbance of blood supply and axoplasmic transport to the intracavernous cranial nerves, which leads to the ophthalmoparesis; it may also be induced by extraocular muscle congestion;
4. Secondary glaucoma, which is induced by the increased intraocular pressure due to increased venous pressure in the sclera;
5. Hypopsia, which could be visual acuity decrease resulting from insufficient retinal arterial infusion, occlusion and venous stasis, or secondary to corneal involvement, optic nerve involvement or glaucoma;
6. Life-threatening epistaxis, a rare manifestation, which may be associated with pseudoaneurysm. Jiamsripong *et al.*^[2] reported one elderly male patient with pulsatile headache in the right orbit and frontal region and diplopia for 10 weeks as well as concomitant murmur in the head. A sudden emergence of epistaxis was shown with no local abnormalities found by the Department of Otorhinolaryngology. The patient then developed refractory epistaxis again and finally died due to hemorrhagic shock. The MRI test found large amounts of blood in the sphenoid sinus and maxillary sinus, with thickening of the ophthalmic veins, and was diagnosed as CCF; and
7. Other symptoms. Because of the communication between the cavernous sinuses on both sides, lesions of one side could result in symptoms in both eyes. In accidental cases, blood was diverted into the contralateral sinus for venous drainage because of venous thrombosis on one side, leading to symptoms in the contralateral eye. Cases of telangiectasia on the optic disc secondary to CCF could also be found clinically. In cases with low-flow fistulas, clinical symptoms are usually not serious, with an occult onset and presentations of mild orbital congestion and exophthalmus, with or without murmurs. The patient in this report had spontaneous CCF, no previous history of trauma, and the fistula was found located at the internal carotid cavernous sinus. Arterial blood

filled into the anterior inferior lacuna of the cavernous sinus via vessel wall rupture and increased pressure in the sinus, leading to the obstruction of ophthalmic venous return and expansion of the superior and inferior ophthalmic veins, especially the significant expansion of the superior ophthalmic veins and blood backflow. Clinical manifestations included pulsating exophthalmos, conjunctival congestion and murmurs in the orbital and frontal regions. Because of the short duration of only 2.5 months and the low-flow fistulas, symptoms including ectropion of eye lid, ocular motility disorder and visual impairment had not yet developed; exophthalmus as well as congestion was rapidly improved after surgery.

Clinical manifestations of CCF are also related to the vascular compensatory response of the patients. Blood from the internal carotid artery of the affected side is diverted via the sinus shunt and involved in the blood supply to the distal vessels of the affected side by the contralateral internal carotid artery and vertebral artery via the anterior and posterior communicating arteries. Symptoms of cerebral ischemia could be developed in cases with poor compensation, while no neurological deficit would be found in adequate compensation cases even if the internal carotid artery of the affected side is occluded during embolotherapy. By imaging of the vertebral artery and internal carotid artery of the uninjured side by compression of the internal carotid artery of the affected side during DSA imaging, knowledge of compensatory blood supply of cerebral circulation in the cerebral hemisphere of the affected side could be acquired and therapies could be chosen according to compensatory blood supply of collateral circulation. In this case, patency of the anterior and posterior communicating artery was shown by pre-operative angiography. Blood flow of the left anterior cerebral artery was compensated by the right internal carotid artery and blood flow of the left middle cerebral artery was supplied by the left vertebral artery via the posterior communicating artery, with adequate compensation. Imaging of the distal vessels, including the left middle cerebral artery and the left anterior cerebral artery, was clearly shown by post-operative angiography, with disappearance of compensatory blood flow. Cerebral ischemia was not developed after the successful surgery, and symptoms including exophthalmos and conjunctival congestion also disappeared 5 days after the operation.

Diagnosis of CCF mainly relies on the typical ophthalmic symptoms, and diagnosis of arteriovenous fistula is based on nystagmus and murmur in the patient's head. Information about location and size of the fistulas, steal

phenomenon, venous return and blood supply of the internal and external carotid arteries can be acquired from DSA examination, which is the gold standard in defining CCF. Exophthalmos, enhanced imaging of the cavernous sinus or thickened ophthalmic veins could be found by cranial computed tomography and MRI, which are frequently used to assist diagnosis. CCF has a low incidence and is mostly traumatic; it rarely occurs spontaneously. With the rare cases and lack of knowledge, ophthalmic symptoms due to the orbital congestion, chemosis and exophthalmos induced by injuries of the venous system have been the most common and first-onset symptoms, which frequently lead to missed diagnosis or misdiagnosis of CCF. CCF needs to be differentially diagnosed from diseases including Graves' disease, infectious conjunctivitis, vascular injury and tumors such as arteriovenous malformations, cavernous sinus thrombosis, cavernous sinus tumor, orbital apex tumor and skull base tumor.^[3] The characteristic clinical manifestations and imaging examination of CCF could be used as a diagnostic basis to distinguish from other diseases.

The clinical symptoms of CCF are usually aggravated progressively, which may further lead to the development of severe lesions including blindness, more serious neurological deficit and intracranial hemorrhage; thus, early diagnosis and treatment is of great importance to the prognosis. For the treatment of CCF, effective measures must be taken to block the arterial blood flow into the cavernous sinus. Different approaches of treatment are needed for different patients. For elderly patients or those with small fistulas, conservative treatment could be the first choice with the aim of decreasing intraocular pressure. Drugs including acetazolamide and β -receptor blocker could be selected, and arteriovenous compression therapy could be another alternative, which is short and repeated manual compression of the carotid and internal jugular veins, to realize stable conditions by promoting the formation of fistula thrombosis. Close attention should be paid to contraindications including carotid atherosclerosis or hypercoagulable state at the beginning of compression.^[4] Conservative treatment will be considered as ineffective if symptoms aggravate or visual acuity decreases progressively, and fistula blockage is needed as soon as possible. For partial blockage of fistulas in CCF treatment, endovascular treatment is often performed due to the large wounds and poor prognosis resulting from craniotomy. DSA technique, which includes endovascular embolization and endovascular stent implantation, could accurately identify the location of vascular lesions in CCF and enable the endovascular treatment.^[5,6] Embolization could be performed via arteries and veins using mainly detachable balloons and coils as embolic materials, and

fistulas are blocked with the maintenance of carotid artery patency, which makes it an effective therapy. In this case, fistulas were blocked by detachable balloons via the artery and normal blood flow was restored after surgery, with disappearance of compensatory blood supply. However, incomplete blockage or balloon rupture can be found in some patients and stent implantation could be an effective alternative in such cases.^[7]

In summary, herein we reported a CCF case and analyzed the related literatures and arrived at the conclusion that spontaneous CCF is quite rare, of which most are type A according to Barrow's classification, with pulsating exophthalmos, intracranial vascular murmur, conjunctival congestion and edema as the characteristic manifestations. Most patients with CCF often first visit the Department of Ophthalmology, and diagnosis of CCF is based on MRI and DSA examination. Early diagnosis and treatment could reduce the incidence of related complications such as cranial nerve palsy and decreased visual acuity, and improve the prognosis.

REFERENCES

1. Barrow DL, Spector RH, Braun IF, Landman JA, Tindall SC, Tindall GT. Classification and treatment of spontaneous carotid-cavernous sinus fistulas. *J Neurosurg* 1985;62:248-56.
2. Jiamsripong P, Mookadam M, Mookadam F. An uncommon cause of Epistaxis: Carotid Cavernous Fistula. *Emerg Med J* 2007;24:e28.
3. Chaudhry IA, Elkhamry SM, Al-Rashed W, Bosley TM. Carotid cavernous fistula: Ophthalmological implications. *Middle East Afr J Ophthalmol* 2009;16:57-63.
4. Howlett DC, Watson GM, Jeffree MA, Garlick D. A red eye. *Br J Radiol* 2006;79:1007-8.
5. Yu JS, Lei T, Chen JC, He Y, Chen J, Li L. Diagnosis and endovascular treatment of spontaneous direct carotid-cavernous fistula. *Chin Med J* 2008;121:1558-62.
6. Gomez F, Escobar W, Gomez AM, Gomez JF, Anaya CA. *et al.* Treatment of carotid cavernous fistulas using covered stents: Midterm results in seven patients. *AJNR Am J Neuroradiol* 2007; 28:1762-8.
7. Wang C, Xie X, You C, Zhang C, Cheng M, He M, *et al.* Placement of covered stents for the treatment of direct carotid cavernous fistulas. *AJNR Am J Neuroradiol* 2009;30:1342-6.

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