

Ophthalmic characteristics of carotid cavernous fistula: a case report

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Dear Editor,

We report a case of traumatic carotid cavernous fistula (CCF) that initially presented in the Ophthalmology Department because of the ocular manifestations. CCF is a clinical syndrome characterized by ocular abnormalities, which is caused by abnormal communication between the cavernous sinus and the cavernous segment or other meningeal branches of the internal carotid artery (ICA) due to traumatic or spontaneous factors. In clinical practice, CCF can be classified into A, B, C, and D types according to the supply artery. Type A is a direct high-flow fistula between the cavernous sinus and the ICA, which is mostly caused by craniocerebral trauma and partial aneurysm rupture with typical clinical manifestations. By contrast, most of types B, C, and D are low-flow fistulas with many blood supply arteries and small fistulas, namely indirect types. About 80% of the patients take ocular symptoms or signs as initial manifestations^[1], such as blepharal or conjunctival congestion, exophthalmos, visual impairment, diplopia, and restricted movement of the involved eye. As a result, many patients with CCF may initially seek help from an ophthalmologist. Due to the diversity and complexity of its clinical manifestations, especially for low-flow fistula of which the initial clinical symptoms are usually mild and the onset is insidious, CCF is prone to be misdiagnosed as

other ophthalmic diseases. Herein, by presenting a CCF case combined with a literature review including its etiology, manifestations, intervention, and outcome, we aim to enhance the understanding of CCF by ophthalmologists.

A 43-year-old female presented in the Ophthalmology Department with a 10-day history of progressive redness, pain, bulging, and blurry vision in her left eye (Figure 1A). The study was conducted in accordance with the principles of the Declaration of Helsinki. The informed consent was obtained from the patient. Computed tomography (CT) imaging of the orbits showed an enlargement in the left extraocular muscles, the dilation of superior ophthalmic vein (SOV; Figure 1C) and inflammation in the bilateral maxillary sinuses. The patient had been treated with an empirical systemic antibiotic at a local clinic and discontinued after the symptoms subsided. On ophthalmic examination, visual acuity and intraocular pressure (IOP) of the right eye were 1.0 and 16 mm Hg. For the left eye, visual acuity and IOP were hand motion (HM)/20 cm and 51 mm Hg respectively; severe proptosis, chemosis, corneal edema, eyelid swelling with mild tenderness and skin temperature rise, and ocular motility restriction presented; the anterior chamber and lens appear unaffected, while the fundus was not clear due to the corneal edema. We noted anisocoria with a mydriasis and relative afferent pupillary defect in the left eye. Abnormally elevated laboratory results showed as follows: leukocyte $9.53 \times 10^9/L$, neutrophil 79.20%, whole blood high sensitivity C-reactive protein 62.91 mg/L. Thyroid function test results were negative. A presumptive diagnosis of orbital cellulitis was made.

The patient reported a history of surgical removal of subdural hematoma resulted from head trauma 1mo ago, and was considered to have fully recovered. This raised our suspicion of CCF. Further inquiries revealed a crepitus on the left side after surgery, which became obvious during sleep and disappeared recently. There was no eye pulsation, but we noted a vascular bruit consistently with arterial pulsation. Thus, cranial magnetic resonance angiography (MRA) was performed timely and the results highly suspected CCF. As a result, the patient was referred to the Neurosurgery Department. The digital subtraction angiography (DSA) confirmed the diagnosis of left CCF (type A, Figure 1D) and the patient accepted an

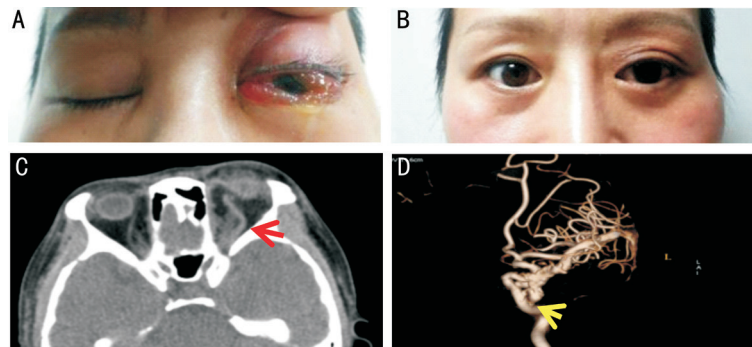


Figure 1 The female patient A: Extraocular appearance pre-treatment; B: 1-week post-treatment; C: Orbital CT images revealed the dilation of SOV; D: 3-D DSA images showed enlargement of the left cavernous sinus. SOV: Superior ophthalmic vein.

embolization surgery. One week after surgery, the corrected visual acuity in left eye was improved to 0.8, and the IOP was reduced to 16 mm Hg. The appearance of the globe almost returned to normal (Figure 1B). The cornea and pupils and ocular motility also returned to normal. Fundus view was clear and retinal linear hemorrhage was noted.

This patient presented with severe proptosis, eyelid swelling, chemosis, corneal edema, restricted eye movement, increased IOP, and linear retinal hemorrhage. All these manifestations can be explained by the pathophysiology of CCF.

Cavernous sinus receives the drainages from SOV, inferior ophthalmic vein (IOV), superior petrosal sinus, inferior petrosal sinus, sphenoidal sinus, and the basal venous plexus. SOV and IOV collect the venous reflux of orbital tissue and eyeball. Increased intracavernous sinus pressure leads to impeded venous return and increased intraorbital venous pressure, followed by ocular symptoms and signs. It should be noted that conjunctival congestion is characterized by tortuous dilation of the blood vessels and corkscrew hyperemia due to the pathophysiology of episcleral and conjunctival arterIALIZATIONS. This patient did not exhibit typical conjunctival congestion, probably because it was accompanied by severe edema. What is worse, the raised episcleral venous pressure can cause retinopathy and optic neuropathy^[2], leading to visual impairment. A few patients may undergo neovascular glaucoma due to long-term retinal ischemia and serious choroidal congestion, or even choroidal detachment^[3]. Optical coherence tomography angiography of the anterior segment is a non-invasive method to reveal the abnormal episcleral venous plexus secondary to CCF^[4]. Inam *et al*^[5] found that the thickness of the choroidal membrane under the macular and the choroidal blood flow index of CCF significantly increased. As the intraorbital pressure decreased and the corneal edema subsided postoperatively, we observed linear retinal hemorrhage in this case, which is evidence of retinal venous reflux.

Furthermore, there are oculomotor nerve (III), trochlear nerve (IV), trigeminal nerve (V), and abductor nerve (VI) passing

through the cavernous sinus. Increased pressure within the cavernous sinus can lead to nerve compression. Among them, abductor nerve paralysis is the most common type because it is located in the cavernous sinus adjacent to the ICA. This patient presented with ptosis and restricted eye movement owing to oculomotor nerve palsy to a great extent, while pupil dilated due to increased IOP and pupillary sphincter paralysis. In addition to ocular symptoms, CCF can also cause neurological symptoms such as headache, diplopia, confusion, aphasia, which were not presented in this case. Given the definite history of craniocerebral trauma with typical symptoms and signs in this case, we classified it as CCF type A.

In this case, orbital CT showed an enlargement of left extraocular muscles and dilation of SOV. MRA demonstrated a dilation of the left cavernous sinus and SOV. DSA exhibited the specific location and size of the fistula, the arterial blood supply and the venous drainage. Although DSA is an invasive examination, it remains the gold standard for diagnosis and classification of CCF at present and can be used as a therapeutic method as well. There is no significant difference in the diagnostic sensitivity on CCF between computed tomographic angiography (CTA) and DSA, while MRA was less sensitive. These differences depend on the segmental location of the fistula. Clinical evidence showed that some patients were finally diagnosed with CCF by DSA, while no positive results were found in early orbital ultrasonography, CTA or MRA^[6]. Thus, CCF cannot be excluded even with normal CTA and MRA findings, especially in case of low-flow and indirect types. When CCF is highly suspected, DSA examination should be performed.

The location of fistula and the pressure in sinus determine the diversity and complexity of clinical manifestations of CCF. Therefore, it is easy to cause missed diagnosis or misdiagnosis. In this case the patient was initially misdiagnosed as orbital cellulitis due to markedly reddened and swollen eyelid, elevated skin temperature, conjunctival congestion, effective antibiotic treatment, and the abnormal results from a routine

full blood count and C-reactive protein test. And beyond that, CCF may also be mistakenly identified as other eye problems due to its atypical manifestations, such as conjunctivitis, scleritis, thyroid-associated ophthalmopathy (TAO), painful ophthalmoplegia, inflammatory pseudotumor^[6]. Some patients have multiple comorbidities, which makes diagnosis more difficult. Previous case studies^[7-8] reported that traumatic CCF induced brainstem edema or hemorrhage, suggesting that delayed diagnosis could lead to serious consequences.

The therapeutic principle of CCF is to close the fistula. The methods are various and complex, including endovascular intervention, manual carotid jugular compression, radiosurgery and open surgery^[9]. However, endovascular intervention has remained the first-line treatment for most CCFs, and more than 90% of patients can be cured in this way^[10]. In this case the patient underwent embolization surgery and her ocular symptoms relieved obviously. Some low-flow fistulas often resolve spontaneously, however, it is noteworthy that individuals with a significantly closed fistula after conservative treatment should be closely monitored, to avoid worsening ocular complications. For example, Thinda *et al*^[11] reported a case of choroidal detachment and angle-closure glaucoma worsened within two months subsequent to fistulous closure.

Despite the disease involves the interdisciplinary fields of neurology and ophthalmology, in most cases, patients with CCF were primarily presented to ophthalmologists. It is important for ophthalmologists to identify this condition, by virtue of combining its characteristic symptoms and signs with medical history and imagological examination results. In this case, the patient's medical history and clinical manifestations were typical, and the data collected were complete, which can provide reference for ophthalmologists.

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