Dural Carotid Cavernous Fistula Presenting as Chronic Conjunctival Congestion

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Abstract

Carotid cavernous fistula is a vascular malformation connecting carotid artery and cavernous sinus. A 45-year old male presented with a chronic history of pain and redness in the left eye, without any history of orbital trauma. On examination there was mild proptosis of left eye, with dilated conjunctival vessels. Extraocular movements were full and there were no visual filed defects. On auscultation no bruit was heard over the eyeball. Magnetic resonance imaging of the brain showed swollen extraocular muscles and dilated superior ophthalmic vein on the left side. Magnetic resonance angiography demonstrated a tuft of vessels over the left temporal dura mater. Digital subtraction angiography demonstrated a dural carotid cavernous fistula over the greater wing of sphenoid. The arterial feeders originated from external carotid artery via middle meningeal and accessory meningeal arteries. Venous drainage was seen to the left superior ophthalmic vein, the cavernous sinus, the superior sagittal sinus via dilated cortical veins and to the straight sinus via basal vein of Rosenthal. The patient underwent transvenous endovascular embolization and his symptoms subsided.

Keywords: Carotid Cavernous Sinus Fistula; Magnetic Resonance Imaging; Digital Subtraction Angiography; Therapeutic Embolization

Introduction

Carotid cavernous fistula is a vascular malformation connecting carotid sinus and cavernous sinus, either directly within the cavernous sinus or indirectly through dural connections (Dural carotid cavernous fistula). When it presents as proptosis and ophthalmoplegia, it is easily diagnosed. Here we report a case of dural carotid cavernous fistula which presented as chronic non-healing conjunctivitis, which posed a diagnostic challenge, but responded promptly to endovascular embolization.

Case Report

A 45-year old male presented with a chronic history of pain and redness in the left eye. There was no history of trauma to the eye. On examination there was mild proptosis of left eye, with dilated conjunctival vessels (Figure 1, Panels A and B). Extraocular movements were full and there were no visual filed defects. On auscultation no bruit was heard over the eyeball. Magnetic resonance imaging of the brain showed swollen extraocular muscles and dilated superior ophthalmic vein on the left side (Figure 2, Panel A). Magnetic resonance angiography demonstrated a tuft of vessels over the left temporal dura mater (Figure 2, Panel B). Digital subtraction angiography demonstrated a dural carotid cavernous fistula over the greater wing of sphenoid (Figure 3, Panel A). The arterial feeders originated from external carotid artery via middle meningeal and accessory meningeal arteries. Venous drainage was seen to the left superior ophthalmic vein, the cavernous sinus, the superior sagittal sinus via dilated cortical veins and to the straight sinus via basal vein of Rosenthal (Figure 3, Panel B). The patient underwent transvenous endovascular embolization and his symptoms subsided.

Abbreviations

CCF: Carotid Cavernous Fistula; MRI: Magnetic Resonance Imaging

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Figure 3: (A) Digital subtraction angiography showing carotid cavernous fistula (arrow). (B) Digital subtraction angiography showing the fistula draining to the superior sagittal sinus, the superior ophthalmic vein and the straight sinus (arrows).

Discussion

Carotid cavernous fistula (CCF) is a vascular malformation connecting carotid artery and cavernous sinus [1]. It may be a high-flow or low-flow fistula [2]. In high-flow CCF there is direct connection between the intracavernous carotid artery and the surrounding cavernous sinus due to a luminal tear. This variety mostly occurs following trauma and head injury or in conditions with a fragile arterial wall like fibromuscular dysplasia, Ehlers Danlos syndrome and pseudoxanthoma elasticum. Patients present with rapidly progressive visual loss, eye pain, chemosis, pulsatile exophthalmos, ophthalmoplegia and a bruit is usually heard over the eye. Low-flow CCF results from indirect connection between carotid arterial system and cavernous sinus. This occurs through dural connections and is also called dural carotid cavernous fistula.

Dural CCF which drains anteriorly into superior ophthalmic vein presents with neuro-ophthalmological features like conjunctival injection, proptosis, retro-orbital pain and ophthalmoplegia due to orbital venous congestion [1]. If the fistula drains posteriorly into superior and inferior petrosal sinuses, orbital symptoms are often absent. Dural CCF may also drain into cortical veins and these are at risk of venous infarction and intracerebral hemorrhage. Clinical features of CCF overlap with infiltrative lesions of cavernous sinus, Tolosa-Hunt syndrome and orbital pseudotumor [3]. CCF can hence present with multiple cranial nerve palsies, most often a variable combination of third, fourth and sixth cranial nerves with sensory disturbance in the distribution of ophthalmic division of trigeminal nerve, mimicking a superior orbital fissure or cavernous sinus thrombosis, from which it can be easily distinguished with imaging. The cranial nerve palsies are due to the abnormally high arterial pressure within the cavernous sinus due to the fistula.

MRI is often the first imaging modality done and it may show dilated superior ophthalmic vein and cavernous sinus, edema of extracranial muscles and abnormal cavernous sinus flow void, indirectly demonstrating the CCF [4]. However, the best imaging modality is catheter angiography. High-flow fistulas should be promptly treated with transarterial embolization. Many low-flow fistulas close spontaneously. Those with progressive symptoms are treated with transvenous embolization [5]. Radio-embolization carries risks like distant site infarction due to migration of emboli across the fistula, transient worsening of ophthalmoplegia and intracerebral hemorrhage, but the success rate is high. Those who fail endovascular treatment should undergo neurosurgical clipping of the fistula. Ocular symptoms start to resolve within hours of successful treatment of CCF but recanalization of successfully closed CCF may occur in a minority.

Conclusion

Dural CCF should be considered in any patient with proptosis and dilated conjunctival vessels, when orbital cellulitis, cavernous sinus thrombosis and keratoconjunctivitis are excluded. Many cases can be easily mistaken form chronic non-viral, non-rheumatic treatment resistant conjunctivitis. It is important to be aware of the varied spectrum of its clinical presentation, because it is a treatable cause.

Bibliography


