



Carotid cavernous fistula secondary to ruptured carotid cavernous aneurysm causing orbital compartment syndrome

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ARTICLE INFO

Keywords:

Carotid cavernous aneurysm/fistula
Orbital compartment syndrome
Proptosis
Canthotomy/cantholysis
Ophthalmoplegia
Elevated IOP

ABSTRACT

Purpose: To report the rare presentation of orbital compartment syndrome secondary to formation of carotid cavernous fistula in the absence of trauma or surgery and demonstrate the role of emergent canthotomy/cantholysis as a vision saving procedure.

Observations: A 28-year-old African American female with obesity and migraines presented to the emergency department (ED) with the worst headache of her life after a severe episode of vomiting and dry heaving. Initial CT scan was negative for subarachnoid hemorrhage and patient was discharged. She re-presented to ED several days later with worsening proptosis and left eyelid edema and was discharged on oral antibiotics for presumed preseptal cellulitis after repeat CT scan. When seen by Ophthalmology in clinic, she was noted to have decreased vision, proptosis, decreased color perception, and double vision. She was noted to have orbital compartment syndrome requiring emergent canthotomy/cantholysis in the minor OR. She was sent back to ED for work-up, ultimately revealing a carotid cavernous fistula secondary to rupture of a carotid cavernous aneurysm. She was treated with several coil embolization procedures and ultimately her visual acuity rebounded to 20/20.

Conclusions and Importance: Orbital compartment syndrome may be a rare presentation of carotid cavernous fistula secondary to ruptured carotid cavernous aneurysm in the absence of trauma or surgery. Our case demonstrates the importance of properly assessing the cause of a red, edematous, proptosed eye in clinical setting and initiating an appropriate workup and treatment plan without delay.

1. Introduction

Carotid cavernous aneurysm (CCA) rupture is rare. However, it can result in various complications such as epistaxis, spontaneous thrombosis, subarachnoid hemorrhage, and namely carotid cavernous fistula (CCF).¹ A carotid cavernous fistula (CCF) is a rare and potentially sight-threatening abnormal communication between the cavernous sinus and the carotid arterial system.^{2–4} We present the case of a ruptured CCA causing a fistula and subsequent spontaneous orbital compartment syndrome in the absence of trauma or post-surgical procedure requiring emergent canthotomy/cantholysis to preserve vision. Given that most cases of elevated intraocular pressure for a CCF is secondary to increased episcleral venous pressure and not from a secondary compartment syndrome, we present this case to show the importance of lateral canthotomy/cantholysis in absence of trauma to decompress the globe and orbit.

2. Case report

A 28-year-old African American female with obesity and migraines presented to the emergency department (ED) with the worst headache of her life after a severe episode of vomiting and dry heaving. A CT head without contrast for suspected intracranial hemorrhage revealed no evidence of subarachnoid hemorrhage. She was given a migraine cocktail and discharged home. Seven days later, she returned to the ED with proptosis and left eyelid edema. CT orbits with contrast was ordered for suspected orbital cellulitis, and stated the following: No evidence of post-septal orbital inflammation. Surrounding preseptal periorbital soft tissues appear slightly prominent/edematous. After a presumptuous diagnosis of preseptal cellulitis was made, she was discharged on doxycycline/cephalexin and referred to the outpatient ophthalmology clinic. Nine days after initial presentation, she presented to ophthalmology with a red, painful left eye with proptosis, ophthalmoplegia, and chemosis in the setting of acute headache. She described worsening visual acuity (VA), proptosis, decreased color

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<https://doi.org/10.1016/j.ajoc.2022.101310>

Received 25 January 2021; Received in revised form 28 April 2021; Accepted 18 January 2022

Available online 21 January 2022

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perception, and double vision. Her eye was extremely firm to palpation. Her VA was 20/20 on the right and 20/60 on the left, pinholing to 20/40; there was also a notable left relative afferent pupillary deficit with reduced Ishihara color plates as well. Intraocular pressure (IOP) was 26 mmHg on the right and 60 mmHg on the left. Her extraocular movements (EOMs) were full on the right, but with -3 supraduction, -1 adduction, -4 abduction, and -2 infraduction deficit on the left. Anterior segment exam of the right eye revealed vascular congestion and minimal subconjunctival hemorrhage, while the left eye revealed proptosis with 3+ hyperemia and 360-degree corkscrew vascular congestion with notable optic nerve pallor on funduscopy exam (Fig. 1).

Given high suspicion for orbital compartment syndrome (OCS), an emergent canthotomy/cantholysis was performed in the minor OR, which decreased her IOP to 25 mmHg. The patient was then sent to the University of North Carolina (UNC) hospital to evaluate for potential arteriovenous fistulas, cavernous sinus thrombi, retrobulbar hemorrhage, subarachnoid hemorrhage, or other orbital processes. During thorough hospital work-up, endocrinology was consulted for labs remarkable for panhypopituitarism. After a positive MRI/MRA/MRV, ultimately, a cerebral arteriogram revealed a right sided carotid cavernous fistula (CCF) with drainage into bilateral orbital veins,

with significant venous congestion of the left orbital structures and a left sided intracavernous aneurysm (Fig. 2). This suggested that a ruptured contralateral aneurysm may have been the etiology for the CCF. It was presumed that this mass effect had also caused chronic ischemia to her pituitary gland as noted by the endocrinology service. She subsequently underwent coil embolization procedures with neurosurgery and neuro-radiology. Nine months post treatment, the patient's VA was 20/20 with minimal residual EOM deficits.

3. Discussion/Conclusions

A progressive orbital compartment syndrome (OCS), without doubt, is a vision-threatening entity. When our patient was seen in Ophthalmologic clinic, there was certainly high suspicion for carotid cavernous fistula based on her ocular exam including proptosis, corkscrew vessels, elevated IOP, ophthalmoplegia, and history of headache. This case is unique in demonstrating spontaneous evolution of compartment syndrome from ruptured carotid cavernous aneurysm (CCA) in the absence of trauma or surgery; it also highlights the effectiveness of OCS decompression with canthotomy/cantholysis as evidenced by reduction of IOP from 60 to 25 and reperfusion of the optic nerve head.

Aneurysms of the cavernous tract of the carotid artery are a rare occurrence, with a reported prevalence varying from 0.3 to 1.4% of all intracranial aneurysms.⁵ These are often more common in women and can be secondary to trauma, infections, collagen-vascular disease, or idiopathic.⁵ Most cavernous carotid aneurysms (CCAs) have long been considered benign lesions, often asymptomatic, with a low risk of life-threatening complications.⁶ There are, however, several conditions in which treatment of these aneurysms is warranted including size, rate of growth, mass effect, or rupture.⁶ Ruptured lesions have a propensity to cause a subarachnoid hemorrhage or form a carotid cavernous fistula (CCF), a rare and potentially sight-threatening abnormal communication between the cavernous sinus and the carotid arterial system.²⁻⁵ Even when a CCA does not rupture, it can cause several complications including acute thrombosis within the cavernous tract or progressive compression of cranial nerves in the cavernous sinus.⁵ Symptoms that should be monitored in patients with CCA include diplopia, due to involvement of cranial nerve III, IV, or VI independently or in conjunction with miosis from compression of the sympathetic nerves in the cavernous sinus, trigeminal dysesthesias, and non-specific pain or unilateral headache.⁷ This case highlights the presentation of a CCA that ruptured, creating a CCF, ultimately resulting in orbital compartment syndrome requiring canthotomy/cantholysis.

In hindsight, our patient's previous complaints of chronic fatigue and migraines were likely related to the undetected CCA. Her laboratory workup in the ED revealed panhypopituitarism which was likely secondary to a CCA causing a compressive, secondary pituitary infarction over several months to years; this mass effect could also have been the direct cause of her previously endorsed "migraine" headaches. The CCA likely ruptured the night she experienced dry heaving and vomiting, thereby precipitating the 'worst headache of her life.' Subsequently this rupture event led to the formation of a CCF resulting in orbital compartment syndrome (OCS). Due to the patient's lack of a primary care provider and appropriate follow-up, a number of unproductive ED visits ensued before proper assessment and management of her condition was initiated. Ultimately, several coil embolization procedures were done in order to restore physiologically normal hemodynamic flow across the cavernous sinus and orbit; this resulted in reduction of proptosis, optic nerve pallor, and ophthalmoplegia.

In patients with clinical manifestation of OCS without history of predisposing risk factors, diagnosis of ruptured cavernous sinus aneurysm and resulting direct CCF should be considered. It is imperative to take a close look at the superior orbital vein (SOV) for any signs of congestion on imaging studies in patients of concern. General practitioners should have a low threshold of suspicion for CCA/CCF in patients with headache, proptosis, chemosis, tortuous and dilated (i.e.

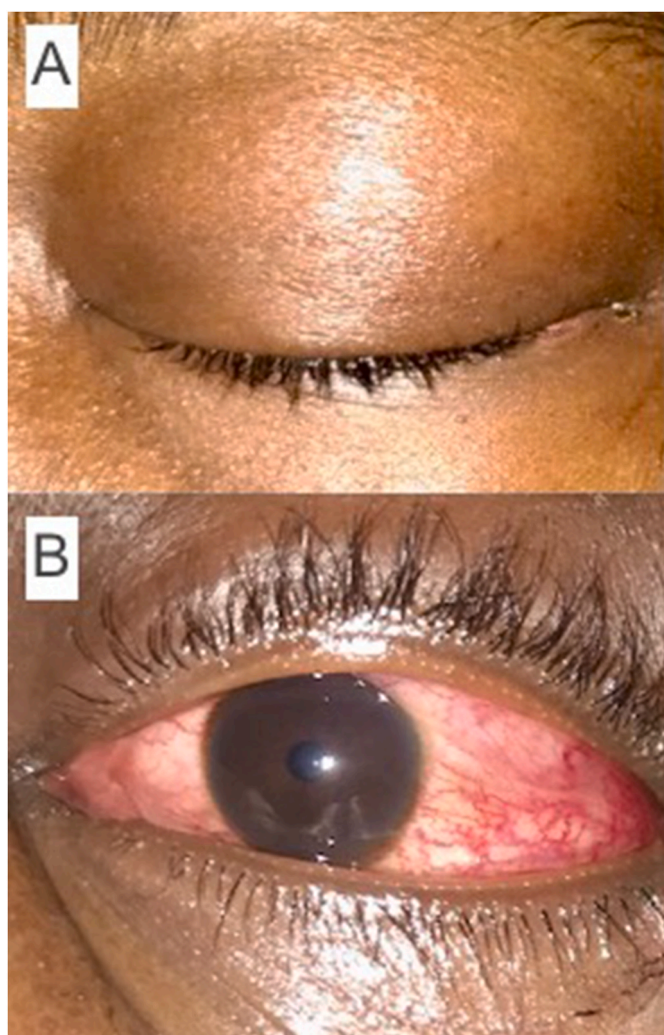


Fig. 1. Clinical Features of Orbital Compartment Syndrome and Increased Episcleral Venous Pressure **A**, Color photograph of patient's left eye showing prominent upper eyelid edema and ptosis **B**, Color photograph of patient's left eye showing chemosis and injected episcleral and scleral vessels. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

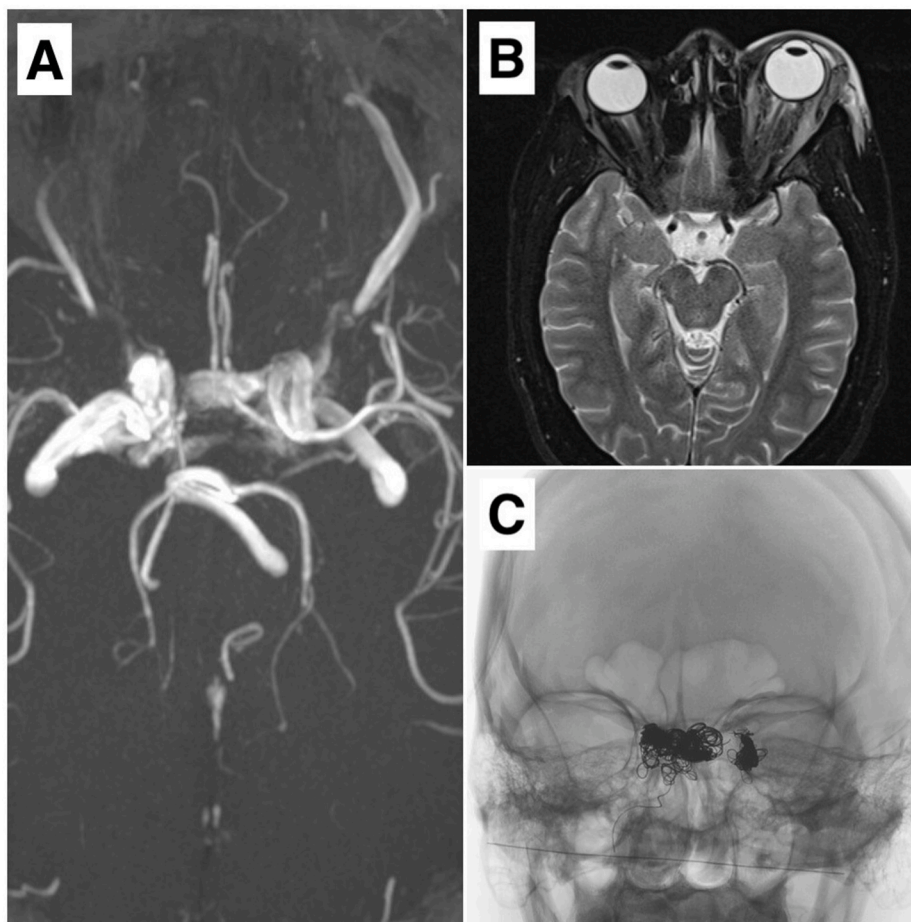


Fig. 2. Radiologic Features of Carotid Cavernous Aneurysm/Fistula A, Magnetic resonance angiography scan demonstrating carotid cavernous fistula on the right. B, Magnetic resonance imaging scan demonstrating left sided proptosis and vascular congestion. C, Interventional cerebral arteriogram demonstrating complete obliteration of right sided carotid cavernous fistula and successful coiling of left sided intracavernous aneurysm.

orkscrew) conjunctival vessels, orbital bruit, and ophthalmoplegia in order to provide timely, effective, and vision saving management.^{2,3,8} Ophthalmologists should not attribute elevated IOP to increased episcleral venous pressure alone in the setting of CCF and should consider emergent canthotomy/cantholysis when indicated to avoid a secondary optic neuropathy and irreversible vision loss.¹

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

Funding

No funding or grant support.

Conflicts of interest

Basil Mathews “none”; O’Rese J. Knight “none”.

Authorship

All authors attest that they meet the current ICMJE criteria for authorship”

Acknowledgments

None.

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