Sudden-onset Blindness from a Spontaneous Carotid-cavernous Fistula with Secondary Central Retinal Artery Occlusion and Posterior Ischemic Optic Neuropathy

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ABSTRACT

Our case describes a patient diagnosed with a carotid-cavernous fistula (CCF) secondary to a spontaneously ruptured cavernous carotid aneurysm, presenting with sudden vision loss, and a concomitant central retinal artery occlusion as visualized by a cherry-red spot in the macula and posterior ischemic optic neuropathy. Computed tomography of the brain and orbits showed mild hydrocephalus, orbital fat haziness, and proptosis with concern for fluid in the basal cisterns. Cerebral angiography confirmed the suspected diagnosis of CCF. After angiography, a Magnetic resonance imaging of the brain demonstrated abnormal diffusion restriction in the posterior right optic nerve confirmed on the apparent diffusion coefficient map, consistent with ischemia of the optic nerve in this location. Two weeks after discharge, outpatient fundus photography showed resolution of her cherry-red spot, and optical coherence tomography showed thinning of the entire retinal nerve fiber layer as compared to the contralateral eye. In CCFs, congestive symptoms of proptosis, pain, and even central retinal vein occlusion findings are frequently described. However, our patient's no light perception vision and imaging findings suggest associated central retinal artery occlusion and ischemic optic neuropathy. These findings underscore the multitude of serious visual effects of high flow CCFs.

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High-flow carotid-cavernous fistulas (CCFs) typically present acutely with subjective bruit, blurry vision, headache, diplopia, and pain in the setting of trauma;¹ however, a ruptured cavernous carotid artery aneurysm may also present in this manner.² We describe a case of spontaneous high-flow CCF presenting as sudden vision loss, with venous congestion and concomitant central retinal artery occlusion (CRAO) and posterior ischemic optic neuropathy (P-ION).

Case presentation

A 37-year-old woman with a history of poorly controlled hypertension presented with right eye swelling, throbbing retro-orbital pain, and complete loss of right eye vision for two days. She denied recent trauma. She did have accompanying symptoms of mild surface irritation but pain was described to be more periorbital and of an achy

quality. She also noted a right temporal headache that was pulsating in quality in addition to an unusual whooshing sound in her right ear. She noted that three days prior, she had no complaints and was seeing well with both eyes. She denied fevers, chills, and weight loss. After a one-day delay in presentation, she went to an outside hospital where a preliminary diagnosis of orbital cellulitis was made and she was transferred to our institution. On presentation to us, she was found to have a blood pressure in the 220s/100s. She was no light perception (NLP) in the right eye with a fixed, dilated, amaurotic pupil. Her extraocular muscle movements (EOM) were restricted in all directions of gaze, and her eyelids were very tight and difficult to open (Figure 1a,b). There was significant injection and chemosis, mostly inferiorly, that was apparent even through a small palpebral fissure. The eye itself was not pulsatile. Her Hertel measurements were 22mm in the right and 18mm in the left

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Figure 1. a: The patient presented with notable proptosis, chemosis, and marked restriction of her extraocular muscle movements. b: The patient's eyelids were swollen shut making it difficult to lift the upper eyelid.

eye. Her cornea was diffusely oedematous and her retina was diffusely pale with a cherry-red spot in the macula but without optic disc pallor or oedema. Her intraocular pressure (IOP) in the right eye was as high as 86mmHg. Computed tomography (CT) scan of the brain demonstrated concern for subarachnoid hemorrhage in the basal cisterns, lateral ventricle enlargement concerning for hydrocephalus, and retro-orbital fat haziness (Figure 2a–c). Despite the elevated IOP, canthotomy, and cantholysis were deferred by the primary team due to the concerning CT findings showing subarachnoid hemorrhage and need for emergent angiography. For similar reasons, additional ophthalmic imaging was not possible at this time.

Neurosurgery placed an external ventricular drain and a cerebral angiogram showed a right complex CCF and right cavernous internal carotid

aneurysm, which were successfully embolized (Figure 3a–d). Notably, the ophthalmic artery appeared patent on the cerebral angiogram (Figure 3, red arrows). A three-dimensional reconstruction following coiling is shown (Figure 4). Her IOP normalized immediately following the embolization of the CCF. Magnetic resonance imaging (MRI) was pursued after embolization demonstrating restricted diffusion in the posterior right optic nerve confirmed on the apparent diffusion coefficient (ADC) map (Figure 5a–b). The patient was discharged after passing her external ventricular drain trial, but her vision remained NLP. Two weeks later, she remained NLP. We obtained fundus photography showing the normalization of the cherry-red spot which was initially observed (Figure 6a). On optical coherence tomography (OCT), the retinal nerve fiber layer

Figure 2. a–c: Axial CT brain and orbits demonstrating subarachnoid hemorrhage in the basal cisterns, lateral ventricle enlargement concerning for hydrocephalus, and retro-orbital fat haziness. Red arrow: Enlargement of the lateral ventricles concerning for hydrocephalus. Blue arrow: A 1.6x 0.6cm area of low attenuation in the right lentiform nucleus that may represent either encephalomalacia or dilated perivascular space. Green arrow: Soft tissue swelling in the right orbit with proptosis.

Figure 3. a–d: Time sequence of a cerebral angiogram showing a right complex CCF and right cavernous internal carotid aneurysm (blue arrow). A patent ophthalmic artery is also seen (red arrows).

and inner retinal layers were thin (Figures 6b, 7). One month later, she remained NLP.

Discussion

Low-flow dural sinus fistulas are known to occur spontaneously without trauma followed by spontaneous resolution. However, high-flow CCFs are usually traumatic in origin with gradual vision loss and EOM movements over weeks.³ CCFs have most often been classified according to the Barrow carotidocavernous fistula classification based on the arterial supply⁴: Type A is the most common, and is a direct connection between the intracavernous internal carotid artery and the cavernous sinus. Type B is an indirect dural shunt connection between the meningeal branches of the intracavernous internal carotid artery and the cavernous sinus. Type C is an indirect dural shunt connection between meningeal branches of the external carotid artery and the cavernous

sinus. Type D is a combination of Types B and C. Our patient presented with a type A CCF but did not have a history of trauma. Her findings were consistent with a CCF including CT imaging with subarachnoid hemorrhage, proptosis, whooshing sound in the ears, and prominent proptosis. She also had a history of hypertension that was untreated and poorly controlled. Our patient had a ruptured cavernous carotid aneurysm that led to the formation of a high-flow CCF rather than a ruptured CCF. The CT findings of subarachnoid hemorrhage are consistent with a type A CCF. Secondly, the CT findings of hydrocephalus are likely from obstruction secondary to the unruptured aneurysm, specifically the blockage of ventricles leading to the accumulation of cerebrospinal fluid.

There are also reported cases in the literature of ruptured cavernous carotid aneurysms as the cause of high-flow CCFs,^{5–7} but many others have involved other pathology or iatrogenic trauma.

Figure 4. A three-dimensional reconstruction of the cerebral angiogram showing embolization coils (grey). Patent right ophthalmic artery shown (blue arrows).

Figure 5. a: Magnetic resonance imaging performed after embolization of the CCF shows high signal intensity in the posterior right optic nerve on diffusion-weighted imaging (blue arrow). b: An apparent diffusion coefficient (ADC) map shows corresponding a hypointense posterior right optic nerve (red arrow).

One case series of spontaneous CCFs illustrated presentation with gradual onset EOM restriction, proptosis, or just injection of the conjunctiva that can occur in a variety of ages.³ Notably, our patient

did have a history of uncontrolled hypertension which can be associated with dural sinus fistulas.⁸ However, more uniquely, our patient also presented with a pale retina with a cherry-red spot despite an

Figure 6. a: Two weeks after presentation, the patient had resolution of her cherry-red spot in the macula, resulting in a normal appearing fundus. b: Optical coherence tomography showing thinning of the inner retinal layers two weeks after presentation.

angiographically patent ophthalmic artery. While our patient had congestive symptoms of pain and proptosis with elevated IOP, her retinal findings suggested a simultaneous CRAO likely from prolonged elevated intraocular and intraorbital pressure that normalized after embolizing treatment.^{1,9} Elevated IOP in particular is known to cause CRAO.¹⁰ The normalization of the cherry-red spot indicates the return of perfusion following the embolization of the CCF during angiography, however, the OCT showed persistent thinning of the retinal nerve fiber layer and the inner retinal layers two weeks post-presentation. Fluorescein angiography was unable to be pursued at the time of presentation given the need for acute intervention for the CCF, and was later deferred given the return of normal macular colour observed on fundus photography alone.

In addition to the CRAO, our patient likely had a P-ION based on the amaurotic pupil and corroborated on MRI following embolization. The patient also did not have any optic nerve head

oedema, which supports a P-ION. MRI brain was ordered; however, images obtained from the standard brain MRI sequences passed through the optic nerve. DWI and ADC sequences were requested on the MRI brain. DWI and ADC maps on MRI showed changes consistent with ischemia of the posterior right optic nerve, which has been described for ischemic optic neuropathies of other etiologies.^{11,12} If it weren't for the findings on MRI, it could have been conceivable that the APD and RNFL loss could be explained by a large CRAO alone. A large CRAO can cause a reverse APD and thinning on OCT could have been due to Wallerian degeneration from the large CRAO causing RNFL loss. Furthermore, MRI of the optic nerve in CRAO has not been studied and while P-ION is strongly suspected based on the compelling MRI findings, it is not certain.

One case report of P-ION caused by CCF has been described.¹³ In this case, it was postulated to be related to arterial steal in the setting of a meningeal-ophthalmic artery anastomosis and a posterior draining CCF.¹³ Aside from arterial steal, one can consider ischemic due to compression with orbital congestion as a mechanism for P-ION caused by CCF. Another consideration is the low flow through the internal carotid artery causing decreased transient flow to the optic nerve, perhaps initially during rupture or later on during the expansion of the fistula. And while the literature described elevated IOP as a risk factor for anterior ischemic optic neuropathy, it has not been definitely demonstrated for P-ION. In summary, the findings of CRAO and P-ION from an underlying CCF explain her persistent NLP vision.

Few reports of CCFs associated with CRAO have been reported in the literature.^{9,14,15} However, to our knowledge, ours is the first case to associate a non-traumatic etiology (malignant hypertension) with simultaneous P-ION. The association of CCF with CRAO has been intriguing and open to speculation in prior case reports. In our patient, although the ophthalmic artery on the cerebral angiogram was patent, the cherry-red spot seen clinically in the macula suggested a central retinal artery occlusion had occurred. The central retinal artery is distal to the ophthalmic artery and such a finding could be plausible. One can also postulate that an ischemic low-flow state with maintained

Figure 7. Two weeks after presentation, retinal nerve fiber layer thickness analysis quantitatively shows diffuse thinning in all quadrants after the posterior ischemic optic neuropathy and central retinal artery occlusion.

vascular patency, can lead to shunting away from the ophthalmic artery. In addition, the ophthalmic or central retinal artery may have undergone temporary vasospasm during the time of the angiogram.⁹ Another possibility described in the literature is a phenomenon similar to Terson's syndrome where a subarachnoid hemorrhage, as in our patient, can increase the venous pressure which can raise IOP,⁹ or that the venous backflow pressure or episcleral venous pressure caused by the CCF

communication was great enough to block retinal perfusion leading to a similar presentation of a central retinal artery occlusion. Interestingly, in our patient, there were no ocular congestion signs such as that seen in a central retinal venous occlusion including tortuous or dilated venules in the retina.

This case report serves to illustrate the diversity of clinical sequelae that can occur with CCFs and that careful history and examination must be

performed in conjunction with advanced imaging modalities to identify all contributing factors for vision loss in these complex cases.

Declaration of Interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the article.

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