Case report

Hyperbaric oxygen for the treatment of the rare combination of central retinal vein occlusion and cilioretinal artery occlusion

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Abstract

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A 43-year-old male presented with sudden onset of painless, blurred vision in his left eye. Dilated fundoscopic examination showed signs consistent with the diagnosis of a combination of central retinal vein occlusion (CRVO) and cilioretinal artery occlusion (CLRAO). He received daily 2-h sessions of hyperbaric oxygen treatment (HBOT), 253 kPa for 14 days. At the end of the HBOT course, the patient's left visual acuity had improved from 20/200 to 20/20. Dilated fundoscopic examination showed that the intra-retinal haemorrhages in the entire retina and the retinal whitening along the course of the CLRA seen at presentation had completely resolved. The combination of CLRAO and CRVO comprises a discrete clinical entity. Even though there are many hypotheses concerning this condition, it is most likely the result of elevated intraluminal pressure in the retinal capillaries due to CRVO that exceeds the pressure in the CLRA. HBOT may be an effective treatment for CRVO-associated CLRAO.

Key words

Vision; sudden blindness; hyperbaric oxygen therapy; case report

Introduction

The cilioretinal artery (CLRA) arises from the short posterior ciliary arteries and can be seen in about 32% of eyes.[1] The number, size and distribution of these arteries vary widely. In approximately 19% of eyes, the CLRA contributes to the macular blood supply.[1] Veno-occlusive retinal disorders are a common visual-impairing condition and central retinal vein occlusion (CRVO) is among the most common primary veno-occlusive disorders of the retina.[2] The frequency of CRVO ranges from 0.2% to 0.8% in population-based studies.[2] At the onset of CRVO some eyes may have associated cilioretinal artery occlusion (CLRAO).

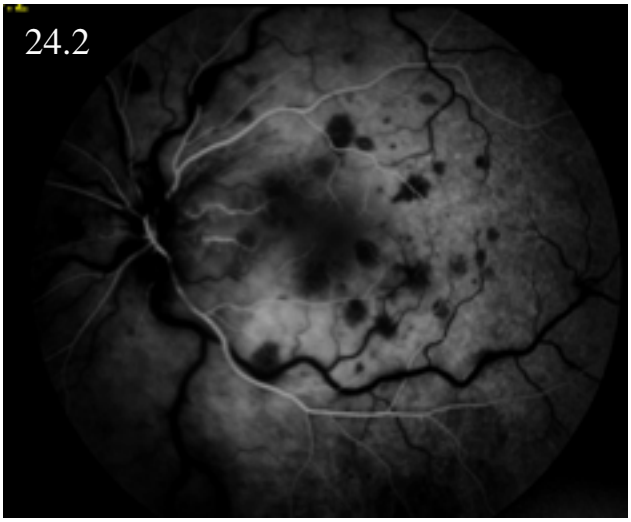
The combination of CRVO and CLRAO was first described in 1968.[3] The pathogenesis of CLRAO in patients with CRVO is not precisely known. Numerous therapies (e.g., surgical embolectomy, Nd:YAG embolysis) have been used to treat retinal arterial occlusive disorders. Supplemental oxygen is administered to maintain retinal function and restore vision via diffusion from the choroidal circulation to the inner layers of the retina.[4] Hyperbaric oxygen treatment (HBOT) administered as early as possible within 24 hours after the diagnosis of occlusive vascular disease of the retina is being used in some centres with some success in selected patients.[5–7] We present a patient with CRVO and CLRAO who was successfully treated with HBOT, and summarise the pathogenesis of this dual entity.

Case report

A 43-year-old male presented with sudden onset of painless, blurred vision in his left eye. He reported having visual obscurations during the week prior to this. The patient's ophthalmic and systemic history was negative prior to this visual complaint. At presentation, visual acuity was 20/20 in the right eye (OD) and 20/200 in the left (OS). Intraocular pressures were 16 mmHg OD and 18 mmHg OS. Slit-lamp examination was unremarkable. Dilated fundoscopic examination of the left eye showed left optic disc swelling, multiple small intra-retinal haemorrhages in the papillomacular bundle, deep blot haemorrhages in the entire posterior pole, as well as increased vessel tortuosity and venous dilatation. Retinal whitening was observed along the course of the CLRA. Examination of the right eye was normal. Visual field testing was unavailable at the time due to technical problems with the equipment.

Fundus fluorescein angiography (FFA) of the left eye showed delayed filling of the central retinal vein and prolonged arteriovenous filling time. The CLRA began to fill as dye appeared in the retinal arteries 24.2 s after injection of the dye. At 33.7 s the retinal veins had just begun to fill proximally with dye, which indicated prolonged retinal arteriovenous transit time (Figures 1 and 2). Based on these findings, the patient was diagnosed as having combined CRVO and CLRAO. Systemic examination showed previously undetected hypertension. A full blood

Figure 1
Fundus fluorescein angiography of the left eye showing slow filling of the CLRA with dye (image taken at 24.2 s)



count, coagulation screen, C-reactive protein, fasting blood glucose, homocysteine level, and liver function were all normal, as were electrocardiography, chest X-ray, carotid ultrasonography and transthoracic echocardiography. The only abnormalities noted were high LDL-cholesterol (157 $mg \cdot dl^{-1}$) and triglyceride (244 $mg \cdot dl^{-1}$) levels. The patient was begun on anti-hyperlipidaemia treatment. His blood pressure was not lowered during the event but later on he was started on antihypertensive medications with beta-blockers on a regular basis.

After the diagnosis was established, he was transferred to a hyperbaric facility elsewhere for daily 2-h HBOT at 253 kPa, which was continued for 14 consecutive days without incident. After the final HBOT, his visual acuity had increased to 20/20 OS. Dilated fundoscopic examination of the left eye showed that the intra-retinal haemorrhages in the entire retina and the retinal whitening along the course of the CLRA had completely resolved. There were small refractile yellow-white iridescent crystals noted at the end of the CLRA. Fundus fluorescein angiography showed a normal dye arteriovenous transit time of 16.6 s and a lack of abnormal florescence (Figure 3).

Discussion

Cilioretinal artery occlusion has been reported in association with embolism, CRVO and a variety of medical conditions as well as with pregnancy.[1] Combined CRVO and CLRAO was first described in 1968, and subsequently reported to account for 40% of all CLRA obstructions.[3,7] There are three forms of CLRAO: isolated non-arteritic; associated with giant cell arteritis, and associated with CRVO.[8] CLRAO associated with CRVO is a clinical entity thought to be due to transient haemodynamic blockage of the CLRA caused by a sudden sharp increase in intraluminal pressure in the retinal capillary bed to a level higher than that in the CLRA.

Figure 2
Fundus fluorescein angiography of the left eye showing delayed filling of the central retinal vein and prolonged arteriovenous filling time in the left eye; dilatation and tortuosity of the retinal vessels was also noted in all four quadrants (image taken at 33.7 s)

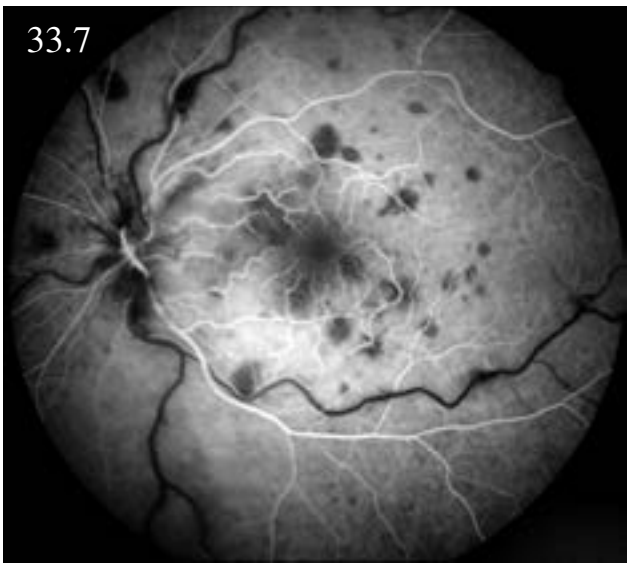
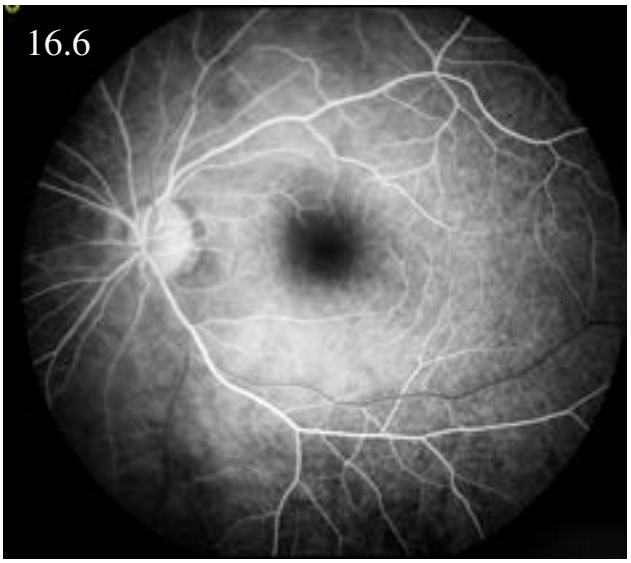


Figure 3
Fundus fluorescein angiography of the left eye following 14 HBO treatment showing a normal dye transit time and lack of abnormal florescence (image taken at 16.6 s)



The pathogenic mechanism of CLRAO combined with CRVO remains unknown. One hypothesis is that CLRAO develops secondary to elevated capillary pressure caused by CRVO.[9] Another is that a primary reduction in the perfusion pressure in the cilioretinal and retinal arteries causes a

decrease in retinal circulation, and subsequent venous stasis and thrombosis.[10] In eyes with a cilioretinal supply, the probability that cilioretinal infarction will complicate retinal vein occlusion is thought to increase as the severity of venous obstruction increases and as the origin of CLRA increases distally from the posterior ciliary artery tree.[11] Indicators of the degree of venous obstruction that may be necessary to instigate cilioretinal infarction include: a very prolonged (defined as more than 30 seconds) dye transit time in the central circulation, increases in venous cyanosis and tortuosity, perivenous cotton wool sentinels and macular perivenular whitening.[11]

Another hypothesis is that a primary reduction in central retinal arterial and CLRA perfusion, or arterial vasospasm, produces secondary venous hypoperfusion and stasis, promoting thrombosis.[10,12] A further hypothesis is based on the fact that arterial perfusion pressure must overcome venous pressure to maintain circulation, and that a marked increase in intraluminal retinal capillary bed pressure due to venous blockage exceeds intraluminal CLRA perfusion pressure, resulting in occlusion.[13] Experimental studies have also shown that arterial constriction following venous obstruction is attributable to a decrease in local levels of nitric oxide, which might contribute to reduced CLRA perfusion.[14] The central retinal artery has sufficient auto-regulatory capacity to maintain perfusion, in contrast to the CLRA arising from the choroidal vascular bed.[13] Furthermore, perfusion pressure in the choroidal vascular bed is lower than that in the central retinal artery.[15]

Fluorescein fundus angiography provides useful information in eyes with CLRAO. Normally, the CLRA begins to fill immediately before the central retinal artery at the optic disc, although in some eyes the cilioretinal and the central retinal arteries begin to fill at the same time.[13] However, in eyes with CRVO and CLRAO, the CLRA filling time depends upon the time between the onset of visual symptoms and fluorescein angiography. Observation of eyes within a couple of hours of the onset of visual symptoms reveals a classical oscillating blood column in the CLRA (i.e., the artery fills for a variable distance from the optic disc during diastole), whereas in eyes observed two to three days after the onset of symptoms, the CLRA begins to fill earlier. The shorter the time interval between the onset of visual symptoms and signs (i.e., the greater the retinal venous stasis), the longer it takes for the artery to fill.[13] The time it takes for the CLRA to fill depends on: the severity of retinal venous stasis; the speed with which the venous collaterals develop in the optic nerve, and the time between the onset of visual symptoms and angiography.[1,9]

Patients with CRVO and CLRAO generally present with a history of an episode(s) of transient visual blurring before the onset of persistent blurred vision, which is first experienced upon waking from sleep or in the morning when the need for fine central vision first arises. In the eyes of patients with non-ischaemic CRVO without foveal involvement from CLRAO, marked improvement in visual acuity may occur; however, when the retinal infarct involves the foveal zone, central scotoma is irreversible. A detailed discussion of the primary differences between ischaemic and non-ischaemic CRVO is to be found elsewhere.[13] In the presented case, because visual obscurations were reported prior to sudden visual loss, it was thought that there was partial occlusion in the central retinal vein and that the CLRAO was secondary to this.

There is no treatment proven to be effective for CLRAO associated with CRVO.[1] The challenge is to administer supplemental oxygen soon enough after the onset of visual loss to prevent irreversible retinal damage. Supplemental oxygen as a treatment option in retinal arterial occlusions showed promising visual results. In experimental models of complete central retinal arterial occlusion the ischaemic time window before permanent retinal damage occurs is 91 mins; in clinical settings in which occlusion may be incomplete vision may be restored even after 8–24 h.[5,6] In patients with retinal arterial occlusion that present within 24 h of visual loss, supplemental oxygen should be started immediately. Recent studies have suggested that HBOT is a safe, easily administered, low-cost and effective treatment in patients with non-arteritic CLRAO.[17] If the patient responds to HBOT, follow-up treatment with supplemental oxygen should be customized to maintain retinal viability until the obstructed retinal artery is re-canalized, which typically occurs within 72 h.[5] HBOT was reported as a safe and effective treatment for a case of cystoid macular edema secondary to retinal vein occlusion.[19] There are no clear recommendations with regard to the number or frequency of HBOT in this clinical situation, though guidelines are available for acute central retinal artery obstruction (CRAO).[5]

The major parameters for visual prognosis are the time from onset of symptoms to the beginning of HBOT, and the time until retinal reperfusion begins.[4] HBOT not only increases oxygenation and perfusion pressure, but also probably reduces intraocular and episcleral venous pressure, which moves a thrombus to a more distal site.[1,17,18]

In conclusion, the combination of CLRAO and CRVO comprises a discrete clinical entity. Even though there are many hypotheses concerning this condition, it is most likely the result of elevated intraluminal pressure in the retinal capillaries due to CRVO that exceeds the pressure in the CLRA. Prospective controlled trials are needed to investigate more fully the role of HBOT as a treatment of choice for CRVO-associated CLRAO and for CRAO.

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