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Original Study • DOI: 10.2478/ahem-2024-0001 • AHEM • Vol. 78 Issue 1/2024 • 1–4

AHEM

Advances in Hygiene and Experimental Medicine
Postępy Higieny i Medycyny Doświadczalnej

Ocular ischemic syndrome – case report and literature analysis

Original Study

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Received 21 June, 2023, Accepted 5 January, 2024

Abstract

Introduction. Ocular ischemic syndrome (OIS) is a rare condition associated with ocular hypoperfusion caused by carotid artery occlusive disease. This article describes the case of a patient who was admitted to the Department of Ophthalmology at UCK in Katowice, Poland, due to significant loss of visual acuity and pain in the right eye.
Materials and Methods. Retrosopective case report.
Results. Patient showed up in the ophthalmological emergency department complaining of significant loss of visual acuity and pain in the right eye persisting for 2 weeks. The patient had a history of type II diabetes mellitus, hypertension, status post ACS and PCI, and long-term nicotinism. Examination of the right eye revealed ocular irritation, mixed conjunctival injection, hyphema in the anterior chamber, iris neovascularization, and discrete central retinal fading. Additional examinations were performed, including an angioCT, which showed a long-standing obstruction of the right internal carotid artery, a ballooning thrombus in the lumen of the brachiocephalic trunk, and suspected critical stenosis of the left vertebral artery.
A neurologist and vascular surgeon were consulted, and treatment was instituted to limit further complications from the underlying disease. The patient remains under close supervision of the vascular surgeon and neurologist.
Conslusions. OIS is a rare but potentially blinding condition and should be considered in the differential diagnosis of transient visual acuity reduction or vision loss.

Keywords

ocular ischemic syndrome • sudden vision loss • eye pain

1. Introduction

Ocular ischemic syndrome (OIS) is a rare condition resulting from ocular hypoperfusion, which occurs due to stenosis of the common or internal carotid artery [1,2]. Most patients who develop OIS have a history of hypertension, diabetes or peripheral vascular disease [2]. Symptoms of OIS include eye pain, gradual or sudden reduction in visual acuity, and even complete loss of vision [2]. These symptoms are not specific, thus being an additional complication in the diagnosis of OIS. The aim of this study is to present the case of a patient who was admitted to the Department of Ophthalmology at UCK in Katowice, Poland, due to significant loss of visual acuity and pain in the right eye.

2. Aim of the study

This article describes the case of a patient who was admitted to the Department of Ophthalmology at UCK in Katowice, Poland, due to significant loss of visual acuity and pain in the right eye.

3. Materials and methods

A 70-year-old man with hypertension and diabetes mellitus and a history of status post-acute coronary syndrome (ACS) and percutaneous coronary intervention (PCI), ischemic stroke and long-term nicotinism presented with a two-week history of intermittent orbital pain and progressive vision loss in his right eye, deteriorating to hand motions. The intraocular pressure (IOP) was in the treshold range at 21 mm Hg. Biomicroscopy showed prominent iris neovasculature, hyphema, iris asymmetry with no response to light, posterior synechiae, cataract, eye was significantly injected and hyperemic (Figure 1A, 1B, and 1C).

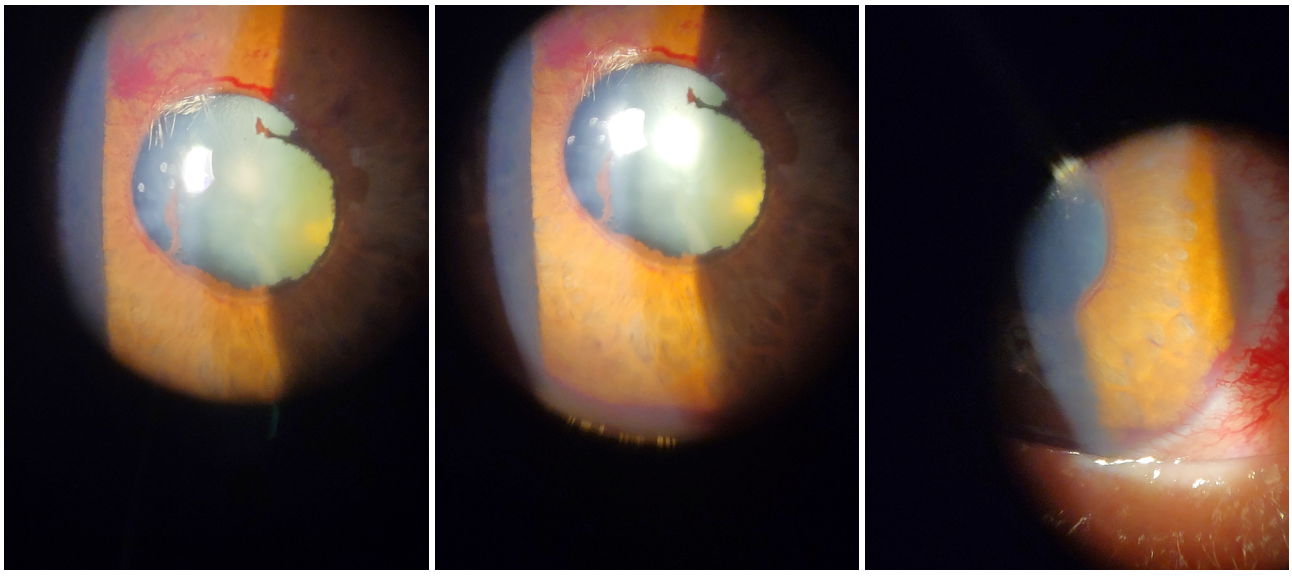
In gonioskopy of the right eye, there was neovascularization of the angle with partial occlusion of the angle. Best corrected visual acuity (BCVA) of the fellow eye was 5/5, IOP was 19 mm Hg. Patient had no history of ophthalmic disease.

Computed tomography angiography (CTA) showed suspected critical stenosis of *left vertebral artery* (*LVA*), ballooning thrombus in the brachiocephalic trunk, and long-standing right internal carotid *artery* (RICA) obstruction.

The Reti-Port electrophysiological device from Roland Consult (Germany) was used to test flash visual evoked potentials (FVEP) in compliance with the guidelines set forth by the International Society for Clinical Electrophysiology of Vision [3]. Skin gold-cup electrodes (active electrodes were placed at O_1 and O_2 reference one at Fz) and standard flashes with frequency of 1.4 Hz were used in the Ganzfeld stimulator: amplitude and latency of P2 wave were measured.

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(A) (B) (C)

Figure 1. Biomicroscopy examination of the right eye showing prominent iris neovasculature (1A), hyphema (1B), and conjunctival injection (1C)

Right eye and left eye Flash VEP results had delayed P2 latency to about 140–145 ms (reference values 120–130ms), and amplitude about 4–5 uV (right eye) and 8–10 uV (left eye; reference values 9–15uV).

Patient was consulted by a neurologist and vascular surgeon, and he was not qualified for surgical treatment and was advised for anticoagulant treatment with enoksaparin. Ophthalmologist included treatment to the right eye: brinzolamid 3 times a day, tropicamidum 2 times a day, dexamethasone 2 times a day, nepafenacum 2 times a day and pentoxifylline 400 mg per day.

After the following two weeks he had elevation of IOP in the right eye to 33 mmHg with no eye pain. Ophthalmologist modified treatment to timolol + dorzolamid 3 times a day to the right eye.

Patient remains under constant control of an ophthalmologist, neurologist, and cardiologist.

4. Results

OIS may be the first sign of carotid artery stenosis and caused chronic hypoperfusion to the eye [4]. OIS occurs with a frequency of 7.5 cases per million every year [5]. In 40 % of cases, it is accompanied with eye pain [5]. Symptoms of OIS are often unilateral [6] and include asymmetric cataract, iris atrophy, iris neovascularization and secondary neovascular glaucoma, iridocyclitis, and a slow response to light [1,7]. Fundus examination showed narrowed retinal arteries, perifoveal telangiectasias, dilated retinal veins, mid-peripheral retinal hemorrhages, microaneurysms, neovascularization at the optic disk and in the retina, a cherry-red spot, cotton-wool spots, vitreous hemorrhage [1,6,8]. Hypoxia of the eye or neovascular glaucoma can caused eye pain in OIS [5].

In 67% of patients, visual loss occurs gradually over a few weeks or months, in 12%, it occurs over a period of days, and in another 12%, the loss is sudden over a period of minutes or seconds [2]. BCVA of the eye with OIS is counting fingers or worse in 37% [9]. Neovascular glaucoma complication can result in light perception loss [8]. Visual loss which lasts from a few seconds to a few minutes occurs in almost 10% of patients with OIS [10] but most patients with transient visual loss do not have OIS.

In OIS we can observed both elevated and decreased IOP. Low IOP can be caused by ischemia of the ciliary body and reduced production of aqueous humor [10]. Ischemia can also cause atrophy of the sphincter muscle of the pupil, and as a result the pupil is fixed and semi-dilated and also has a sluggish reaction to light [5,8]. A symptom that also occurred in the presented case of the patient is dilatation of conjunctival and episcleral vessels [11].

The first sign which may be noticed in fundus examination are diffuse macular capillary telangiectasias and microaneurysms, which may cause macular edema [12]. A cherry-red spot is observed in 12 % of patients with OIS [10].

Patients with OIS may develop normal-tension glaucoma caused by reduction of the retrobulbar blood flow, additionally ischemia of the optic disk may lead to nerve atrophy [13].

Posterior synechiae may develop after clinically silent iridocyclitis and aqueous humor opalescence also may be observed [10].

The most important diagnostic tests in OIS are the minimally invasive CTA/magnetic resonance angiography and Doppler ultrasound of the carotid arteries [8]. Fluorescein angiography of the fundus is a test commonly used in the diagnosis of OIS, the most specific (but not the most sensitive) fluorescein angiography sign is retinal filling time—in the affected eyes it

may be 1 minute or longer (in a normal eye it is approximately 5 seconds) [1,2,5,10]. In 17% of eyes with OIS, macular edema may occur and is often accompanied by hyperfluorescence of the optic disk [7,14] caused by leakage from disk capillaries [15].

Electrophysiology tests such as visual-evoked potentials (VEP), electroretinography (ERG) and ophthalmotonometry are not often used to establish the diagnosis of OIS [8,16].

Two of the most vital conditions that should be included in differential diagnosis of OIS are moderately advanced central retinal vein occlusion (CRVO) and diabetic retinopathy [14]. Key clinical features that enable differentiating the above conditions include comparatively few, usually midperipheral retinal hemorrhages and typically dilated, non-tortuous veins. Although cotton-wool spots, a common sign of diabetic retinopathy, may also be observed in OIS, the presence of hard exudates indicates diabetes-related alterations to the fundus. Furthermore, retinal arteria perfusion pressure is decreased. Delayed choroidal filling in fluorescein angiography is characteristic of OIS [5]. Hemorrhaging will often be different; OIS will be more mid-peripheral, random, and intraretinal, whereas CRVO will be more in the posterior pole, directional, and flame-shaped. Diabetic retinopathy can occur together with OIS [1]. Around 20% of diabetic patients have hemodynamically significant carotid artery stenosis. For this reason, such patients with unilateral retinopathy or marked asymmetry of retinopathy should be examined for possible carotid occlusive disease [5,17]. Patients should have a Carotid Duplex Ultrasound.

Other conditions that should be considered during differential diagnosis are uveitis and hyperviscosity syndromes, dry eye, and anterior pole ischemia syndrome. Particularly in unilateral cases that do not improve with conventional therapies [5,18].

Therapeutic management is based mainly on treatment and prevention of complications of OIS. Some of these complications are irreversible due to ischemic optic neuropathy. One of the treatment options is intravenous and the infusion of vasodilatating and anticoagulant drugs. Neovascularization in the iris, retina, and optic disk, as well as at the irido-corneal angle in the anterior segment, is one of the conditions that characterizes OIS. Following OIS, neovascular secondary glaucoma develops in the eye. To reduce intraocular pressure, topical β-adrenergic blockers or α-2-agonists can be administered in conjunction with topical or oral carbonic anhydrase inhibitors; prostaglandin analogues should be avoided due to their potential pro-inflammatory action [5]. Pilocarpine should not be used because it may promote the growth of posterior synechiae and secondary miosis, both of which increase the closure of the irido-corneal angle and elevate intraocular pressure. It is preferable to employ a diode laser or cryosurgery for partial cycloablation in patients who have glaucoma and ocular discomfort [1].

The most effective ocular hypotensive medications are prostaglandin analogues, which are topical treatments used to lower IOP in patients of normal-tension glaucoma [19].

Carotid artery stenosis is treated surgically using carotid artery endarterectomy (CEA) in systemic treatment [19]. It is effective in symptomatic carotid artery stenosis of 70–90% and in asymptomatic stenosis of at least 60%. As a result, all patients with OIS and significant carotid artery stenosis should take this therapeutic option into consideration [20].

Around 40 % of OIS patients die within 5 years from myocardial or cerebral infarctions. That's why OIS should be treated by a multidisciplinary team of specialists, including an ophthalmologist, cardiologist, neurologist, and general practitioner [1]. Antiplatelet medications, the management of hypertension, atherosclerosis, diabetes mellitus, and coronary heart disease are examples of systemic treatment. A healthy diet, cassation of smoking, and engaging in physical activity are crucial.

5. Conclusions

Due to the nonspecific symptoms, the key to making a correct diagnosis is not only taking a detailed history, and slit lamp examination, but most importantly taking imaging studies of the carotid and vertebral arteries. Despite its low incidence, OIS should be considered in the differential diagnosis of transient visual acuity reduction or vision loss.

Abbreviations

OIS - Ocular ischemic syndrome
BCVA - Best corrected visual acuity
CTA - Computed tomography angiography
LVA - *Left vertebral artery*
RICA - Right internal carotid *artery*
FVEP - Flash visual evoked potentials

Authors' contribution

Aleksandra Górska – Writing—Original Draft Preparation
Marek Sosnowski – Research concept and design
Sebastian Sirek – Literature review, writing—review and editing
Rafał Leszczyński – Supervising the project
Mikołaj Kuźniak – Acquisition of data
Ewa Mrukwa – Kominek – Final proofreading and approval of the version for publication

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Conflict of interest

The authors have no potential conflicts of interest to declare.

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