OCULAR ISCHEMIC SYNDROME: TWO CASE REPORTS OF BILATERAL INVOLVEMENT

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SUMMARY

The ocular ischemic syndrome is characterised by ocular symptoms and signs secondary to severe carotid artery obstruction. In this paper, two cases of bilateral ocular involvement are presented. The first case was caused by severe bilateral carotid stenosis and the second by bilateral occlusion of the carotid artery.

SAMENVATTING

Oculair ischemisch syndroom wordt gekenmerkt door een reeks oculaire symptomen en tekens die secundair aan een ernstige arteria carotis obstructie optreden. In dit artikel worden twee gevallen van bilaterale aantasting voorgesteld. In de eerste casus was een ernstige bilaterale carotisstenose de oorzaak, en het tweede geval werd veroorzaakt door een bilaterale totale carotis interna occlusie.

RESUME

Le syndrome oculaire ischémique se présente par une série de symptomes et signes oculaires secondaires à une obstruction sévère de la carotide interne. Nous présentons deux cas d'atteinte bilatérale. Dans le premier cas une sténose bilatérale était la cause et le deuxième cas était dû à une occlusion bilatérale de la carotide interne.

KEY WORDS

Ocular ischemic syndrome, carotid artery disease.

MOTS CLES

Syndrome oculaire ischémique, athérosclérose de la carotide.

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DEFINITION

Ocular ischemic syndrome refers to the constellation of ocular symptoms and signs secondary to the chronic hypoperfusion of the entire arterial supply to the eye.

Case 1

A 56 year old man was referred to our department for visual loss in both eyes. His visual acuity was 0.25 in the right eye and counting fingers in the left eye. In his medical history we retain diabetes mellitus since several years. Slitlamp examination showed marked rubeosis iridis in both eyes. Fundus examination showed venous dilatation, venous beading and cotton wool spots. On fluoangiography the choroidal filling was extremely delayed. There was pronounced neovascularisation elsewhere with diffuse leakage from the vessels. Both eyes showed pronounced macular edema.

The patient was referred for a doppler ultrasonography of the carotid arteries. A bilateral total occlusion of the common carotid artery was found. In the right internal carotid artery a minimal flow was registered probably due to retrograde pulsations from the vertebral circulation. We proposed an arteriographic examination to confirm the occlusions and to evaluate the vascular status of the vertebral circulation and to evaluate the possibilities of revascularisation. Unfortunately, the patient was lost for further follow-up.

Case 2

A 63 year old man with a history of hypercholesterolemia presented with a floater in the right eye. His visual acuity was 1.0 in both eyes. Slitlamp examination was normal. Funduscopy showed peripheral occlusions in both eyes, neovascularisation elsewhere (NVE) and a vitreous hemorrhage in the right eye. Fluoangiography showed marked ischemia in the peripheral fundus. Laboratory findings were normal. Doppler ultrasonography revealed a stenosis in the right common carotid artery but the flow in the ophthalmic artery was normal. The patient received laser treatment in the ischemic regions. During the next 4 years the patient had 2 new vitreous hemorrhages and additional laser coagulation was performed. After resorption of the hemorrhages his visual acuity was 0.8 in both eyes. Fluoangiographic evaluation showed new regions of NVE.

Carotid duplex and arteriographic evaluation demonstrated a high grade stenosis on the right side and a moderate stenosis on the left side. A carotid endarterectomy was performed. Visual acuity remained stable and no new regions of retinal ischemia were found.

ETIOLOGY

Ocular hypoperfusion is most frequently due to severe carotid artery stenosis or occlusion. Other possible causes (1) are the Takayasu syndrome, ophthalmic artery stenosis, vasospasm (9) and giant cell arteritis (2). In these cases the vascular obstruction can occur anywhere proximal to the point where the central retinal and ciliary arteries branch from the ophthalmic artery.

PREVALENCE

Patients with ocular ischemic syndrome are 65 years on average. Men are more often affected than women (6). It is estimated that approximately 5 - 15 % of patients with marked carotid stenosis present with ocular ischemic syndrome and that the syndrome is probably underdiagnosed (7,8). The true incidence is estimated to be approximately 7,5/1.000.000 (6). The involvement is bilateral in 20 % of the cases.

SYMPTOMS

More than 90 % of patients present with loss of vision, which generally develops gradually. The pathogenesis of visual loss is chronic ischemia of the posterior segment, macular edema and cataract secondary to longstanding anterior segment ischemia and inflammation. In those cases where the visual loss occurs suddenly, a cherry red spot is seen. Amaurosis fugax is the presentation in 5 % of cases. Prolonged dark adaptation has been reported (1).

Figure 1. Red-free image of the left eye of patient 1. Marked non-tortuous venous dilatation, retinal arterial narrowing and midperipheral dot hemorrhages are present.

Fourty % of the patients complain of a dull ache over the eye and brow that sometimes radiates to the temple. This 'ocular angina' is believed to be caused by ischemia to the globe, neovascular glaucoma or ipsilateral dural ischemia.

Finally, symptoms of cerebrovascular disease, such as transient focal neurological deficits can be associated with the ocular ischemic syndrome.

SIGNS

The classic triad of signs includes midperipheral dot and blot hemorrhages, dilated non tortuous venes and iris neovascularisation.

Various anterior segment signs may accompany the classic triad among which corneal edema, Descemets' folds, rubeosis iridis, neovascular glaucoma, anterior segment flare and cells, keratic precipitates and lens opacities are the most common.

Fundus signs that are variably present are retinal arterial narrowing and venous dilatation, retinal midperipheral hemorrhages, microaneurysmata, neovascularisation of the optic disc and the retina, vitreous hemorrhage, cotton-wool spots, pulsation of retinal arteries and the presence of a cherry red spot.

Signs of ischemic optic neuropathy may also be present. Signs of cerebrovascular disease, such as a diminished or absent carotid pulse are frequent.

INVESTIGATIONS

Fluo-angiography reflects the chronic hypoperfusion of the retinal and choroidal circulations, as well as ischemic damage to the retina and retinal vessels. Prolonged arm-to-choroid and arm-to-retina circulation times of over 20 seconds are common. The most frequently observed fluorescein angiographic sign is an increased arteriovenous transit time. The choroidal filling is delayed and/or patchy in 60 % of eyes. Staining of the retinal vessels, macular edema and microaneurysms are other fluoangiographic findings.

A

B

Figure 2. Fluorescein angiogram of the left (A) and right (B) eye of patient 1.
A. Patchy and extremely delayed choroidal filling at 35 seconds post contrast injection.
B. At 40 seconds post-injection, diffuse leakage of the arterial vessels is observed.

Orbital color doppler imaging can demonstrate reduced peak systolic velocities of the central retinal and/or of the posterior ciliary arteries. Color doppler imaging data suggest that posterior ciliary artery hypoperfusion and therefore secondary ischemia of the optic nerve, choroid, retinal pigment epithelium and outer segments of the photoreceptors correlates with visual loss (3). Reversal of ophthalmic artery blood flow is seen in 75% of cases. The flow reversal represents collateralisation through the external carotid artery system.

In ocular ischemic syndrome the electroretinography shows a decreased amplitude of both the a- and b-waves, contrary to a solely central retinal artery obstruction in which only the b-wave is affected. This can be explained by the fact that the b-wave is generally believed to correspond to the function of the inner retinal layers, while the a-wave correlates with photoreceptor function (4). In the ocular ischemic syndrome the central retinal arterial supply as well as the choroidal perfusion is compromised.

Carotid duplex scanning and angiography are excellent imaging modalities that visualise ulcerative vascular lesions, grade of stenosis or occlusion and formation of a mural thrombus. They can also demonstrate collateral circulatory patterns.

TREATMENT

The management of ocular ischemic syndrome consists of the restoration of ocular perfusion as well as local treatment of ischemic damage. Laser photocoagulation or cyclocryotherapy however do not seem to improve the vision in eyes with ocular ischemic syndrome but they appear to reduce the incidence of neovascular glaucoma (7).

Treatment of the carotid stenosis by endarterectomy or bypass surgery may be the most important factor in maintaining or improving vision in eyes with ocular ischemic syndrome. However, despite these efforts, the visual results are discouraging. Whether successful carotid endarterectomy will improve diminished posterior ciliary artery perfusion is not yet known. In many cases the reversal of the carotid stenosis will fail to improve or stabilise vision because of damage caused by neovascular glaucoma, vascular occlusive disease, retinal capillary drop-out and hypoxic damage to the retina and retinal pigment epithelium. Postoperatively, a marked rise in intra-ocular pressure may occur because of an improved ciliary body perfusion.

Treatment of atherosclerotic disease with systemic anticoagulation is controversial because of the risk of bleeding complications. Therefore, systemic anti-platelet therapy and anticoagulation alone is reserved for patients who have inoperable carotid disease. Aspirin may be helpful in preventing further TIA's or strokes in symptomatic patients.

PROGNOSIS

In general, the prognosis is poor. The presence of rubeosis iridis seems to be the best prognostic indicator for visual acuity. The presence of iris neovascularisation most likely implies a greater degree of ischemia to the globe and increased ocular damage.

CONCLUSION

We presented two cases of bilateral ocular ischemic syndrome, a severe ocular complication of systemic atherosclerotic disease. Ocular ischemic syndrome can present in a variety of anterior and posterior segment signs and symptoms. The disease is probably underdiagnosed and gives diagnostic and therapeutic difficulties.

Color Doppler imaging studies suggest the importance of the posterior ciliary artery perfusion in the pathogenesis of ocular ischemic syndrome (3). Other studies (5) report that no capillary drop-out was found in patients with ocular ischemic syndrome. They suggest that uveal ischemia alone can be responsible for the neovascularisation seen in ocular ischemic syn-

drome. Therefore there was no rationale for panretinal photocoagulation.

Prognosis remains quite bad even after surgical reversal of the stenosis.

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