VASCULAR NEURO-OPHTALMOLOGICAL EMERGENCIES

Irene Davidescu, Sanda Nica
Neurology Department, "Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania

ABSTRACT

Neuro-ophtalmology is a borderline specialty, as the eye is not an isolated organ but an extension of the brain. A lot of diseases can cause visual troubles and the vascular ones are quite frequent. The precocious evaluation of the visual function is essential for the therapeutical approach and for the recovery, and all this process seldom needs involvment of a medical team.

The anterior ischemic optic neuropathy includes some pathogenic entities which determine the optic nerve ischemia, with a papilar edema present acutely, painless and without any predicting signs. This disease is very often associated with giant cell arteritis, and the non-arteritic one is the most frequent cause of vision loss in patients over 50 years. The main risk factors are hypertension, diabetes and dyslipidemia. The antiagregant therapy remains essential. Retinal artery occlusion is the ocular equivalent of cerebrovascular disease. The occlusion may be central, causing a loss of central vision in the affected eye, or in a branch of the retinal artery, affecting only a part of the retina served by that branch retinal vessel and sparing central visual acuity. The main cause is the carotidian stenosis and less the cardiac embolia. This is a medical emergency which can be approached with intra-arterial trombolysis if the pacient comes within 100 minutes from the beginning. The transient ischemic attack with ocular implications is manifested with a transitor monocular blindness (amaurosis fugax) and represents a medical emergency, as it could predict a definitive stroke. The ethiology differs, depending on the patient's age, in young ones it can be associated with migraine and the antiphospholipid antibodies syndrome, and in the elderly the main mechanism is the atherosclerotic one.

Key words: neuro-ophtalmology, ischemic optic neuropathy, transient ischemic attack, migraine, antiphospholipid antibodies syndrome

CLASSIFICATION

A sudden visual loss can be uni or bilateral, transitory or definitive.

The most frequent causes of transient monocular visual loss are:

- Ischemia
- Carotidian stenosis
- The gigant cells arteritis
- Oclusion of the retinian vein
- Retinian vasospasm or the retinian migraine
- Ischemic optic neuropathy
- Compression of the optic nerve
- Local causes

The causes for transient bilateral visual loss are:

- Vertebro-basilar ischemia
- Microvascular lesions of the optic nerve (as in hypertension or diabetes mellitus)
- Seizures
- Migraine

ANTERIOR ISCHEMIC OPTIC NEUROPATHY

- This disease includes some pathological entities which determine the optic nerve ischemia (the proximal part to the globe), with monocular visual loss, suddenly, and the appearance of a papilar edema, without pain and other alarming signs.
- The causing mechanism is unknown, but it seems that it is related to the presence of hypotension, with the appearance of a watershed infarction between the central artery of the retina and the posterior ciliary arteries. The embolic mechanism is very rare.
- There are two types of anterior ischemic optic neuropathy:
  - Specific arteriolar pathology, especially giant cell arteritis
  - Nonarteritic: associated with the process of atherosclerosis, representing the most frequent cause of monocular visual loss in patients after 50 years. Approximately 30% of these patients will present disorders in the other eye, especially those with hypertension, diabetes and hyperlipidemia.

CHARACTERISTICS

- Predilection for caucasian race
- No sex predilection
- The average age of onset is in the 7th decade
- Smoking can determine appearance in younger ages

- There is no confirmation of association with carotidian stenosis, but there were not done studies on a significant number of patients
- It can be associated with: migraine, nocturnal arterial hypotension, polymialgia reumatica, increased intraocular pressure, cataract surgery, and use of sildenafil (Viagra) and tadalafil (Cialis)
- There are no genetical factors discovered to determine a predisposition to the appearance of this disease
- Studies of external and systemical factors which influence the homocysteine levels showed that the levels of homocysteine are higher among patients with anterior ischemic optic neuropathy, but we must not forget that the homocysteinemia increases with age.

Clinical characteristics

- Visual acuity is decreased and varies from 20/20 vision to complete blindness
- Color vision is affected proportionally with the visual acuity decreasing, as opposed to optic neuritis, in which color vision typically is reduced drastically
- The visual field we observe an inferior altitudinal defect, but we can see all the types of deficits, including central scotoma
- We will see an edematous optic nerve head, and hemorrhage on the disc is also very frequently present
- Biological inflammatory signs, especially in giant cell arteritis
- The clinical outcome is generally favorable, only a small number of patients will have a severe diminution of visual acuity in the first two weeks from the onset

TREATMENT

- Addressed to the vascular risk factors enunciated
- The antiagregants are essential, especially by preventing the reccurence in the other eye
- For giant cell arteritis, the corticotherapy in high doses and for at least 6 months is important in reducing the risk of reccurence at the other eye.

RETINAL ARTERY OCCLUSION

- The retinal vascularisation (the central retinal artery and the ciliary arteries) comes from the oftalmic artery, the first intracranial branch of the internal carotid artery.
- The retinal artery occlusion is one of the most dramatic ophtalmological emergencies with acute onset.
- The occlusion of the central retinal artery represents over 50% of the occlusions of the retinal vessels, and the gravity of the event is depending of the vascular retinal type.
- The occlusion of the retinal artery by platelet/fibrin or calcium emboli determines the sudden loss of the vision and represents the ocular equivalent of cerebrovascular disease. The occlusion can be total, affecting the central vision in the affected eye or on one branch of the artery, causing a partial visual loss, with the integrity of the central acuity.
- The mechanism of appearance is by determining the retinal tissue ischemia, resulting in acute retinal edema and death of retinal ganglion cells.
- The prognostic is poor, in spite a lot of studies and therapeutical approaches.
- Ethiology:

1) Embolia:
– The arterio-arterial emboli (after carotidian stenosis-20 to 45% of the patients have hemodynamically significant stenosis, >60%), and rarely cardiac emboli.
– In younger ages, under 30 years, it is frequently associated with: posttraumatic carotid dissection, hypercoagulable states, migrena, vasospasm
– The carotidian emboli are due to atheromatous ulcerations at the bifurcation, associated with stenosis; the emboli can be formed of cholesterol, platelet/fibrin (causing TIAs-amaurosis fugax) or calcium (the most dangerous because they can cause irreversibile obstruction)
– The cardiac emboli can be formed of calcium (like in valvular calcifications), infectious (endocarditis), platelet/fibrin (after cardiac infarction) or mixomatous.

2) Trombosis:
– Atherosclerosis
– Systemic vasculitis with periarteritis lesions
– Hemathological states: antiphospholipidic syndrome, Protein S and C deficiency, antithrombin lll deficiency)
– Retinian migrena (an exclusion diagnosis)

- Associated risk factors:

– Atherosclerosis
– Arterial hypertension

- Clinical characteristics:
– Most frequent in the 7th decade of life
– No sex or race predilection
– The visual deficit is irreversible
– The diagnosis is confirmed by angio-MRI
– The visual acuity is affected in central vision and partial if one of the branches is occluded
– The visual field examination shows different deficits, proportional with the extension of the ischemia
– At the fundus examination we can observe in 20% of the cases, emboli.
- Treatment:
– The aim of the therapy is to reduce the intraocular pression by topical or systemical agents, or by performing an anterior paracentesis
– It can be used: intra-arterial thrombolysis (if the patients comes in less than 100 minutes from the onset) or hyperbar oxigen (in the first 2-12 hours)

OCCLUSION OF RETINAL ARTERY BRANCHES

- Almost 40% of the retinal occlusion are affecting only branches of the retinal artery
- The most frequent arteries implied are the temporal ones
- The outcome is better: the partial recovery of the vision up to 80% of the patients
- The therapeutical approach is similar with the one in the total occlusion
- The neovascularisation risk is low but increases when the patients have diabetes

OPHTALMIC ARTERY OCCLUSION

Ethiology:
- Similar factors with those producing the retinal artery occlusion: atrial fibrillation, atrial mixoma, carotidian atherosclerosis with stenosis
- Local factors: retrobulbar anesthesia, orbital diseases, trauma
- Vasospasm after a subarahnoidian hemorrhage

Clinical characteristics:
- Sudden loss of vision finally to blindness
- The fundus examination shows pallor of the retina, and the fluorescein angiography points out retinian or choroidian perfussion deficits

CENTRAL RETINAL VEIN OCCLUSION

Ethiology:
- Hypertension
- Vasculitis
- Glaucoma
- Hypercoagulable states
- Iatrogenic
- Tumoral retrobulbar compression
- Graves disease

Classification: depending on the risk of developing neoformation vessels due to the retinal ischemia:
- Ischemic: hemorrhagic retinopathy.The outcome is poor, with loss of vision, finally to blindness.
- Non-ischemic: venous retinopathy. The outcome is benign, with recovery of vision, almost complete (sometimes the only sequela is the presence of a central scotoma)

Clinical characteristics:
- Painless, unilateral sudden visual loss.
- Fundus examination shows venous dilatations, retinian hemorrhages, papilar edema and neoformation vessels. This examination is not able to differentiate the ischemic type from the non-ischemic one, nor precocious or late in the evolution of the disease.

Treatment: there are not yet discovered real benefit methods for this disease
- Anticoagulants and antiagregants are forbidden!!!
- Hemodilution when there is a viscous state
- Sistemic or local corticotherapy
- Acetazolamida
- Drugs for lowing the intraocular tension
- Surgical treatment: laser-therapy for the non-ischemic type for inducing chorioretinian venous anastomosis
- Panretinian photocoagulation for preventing the appearance of neovascularisation in the ischemic type

OCCLUSION OF RETINAL VEIN BRANCHES

- Represents the second major cause of affectation in the retinian vessels after the diabetic retinopathy
- No sex predilection
- Risk factors: hypertension, cardiovascular diseases, glaucoma
- The most often affected are the upper temporal veins
- Clinical characteristics: visual loss in the affected teritory
- The fundus examination shows superficial hemorrhages, papilar edema, venous congestion

- The fluorescein angiography shows colateral and neoformation vessels, papilar edema, lack of cappilars vessels, serous retina detachments
- Treatment: photocoagulation (when the papilar edema is present or the visual acuity is very affected)
- Outcome: depending on the extension of the lesions, the presence of papilar edema and the changes of the retinian vessels

TRANSIENT ISCHEMIC ATTACK

- Represents an acute episode, with sudden onset and a duration between 2 and 15 minutes
- The ocular manifestation of a TIA is the appearance of a transitor monocular blindness (amaurosis fugax), having a duration of seconds to several minutes. This entity represents an emergency because it can predict a final stroke.
- Ethiology:
– In elderly people the mechanism is mainly atherosclerotic, associated with carotidian stenosis
– In younger people it is frequently associated with migrena, antiphospholipidic syndrome or posttraumatic carotidian dissection
- The paraclinical exploration: imagistic methods, ecodoppler, biological risk factors
- Treatment:
– Antiagregants
– Anticoagulants
– Endovascular therapy: carotidian stent or endarterectomy

REFERENCES

1. **Arnold M, Koerner U, Remonda L, et al** – Comparison of intra-arterial thrombolysis with conventional treatment in patients with acute central retinal artery occlusion. *J Neurol Neurosurg Psychiatry* 2005; 76: 196-9
2. **Hayreh SS, Joos KM, Podhajsky PA, Long CR** – Systemic diseases associated with nonarteritic anterior ischemic optic neuropathy. *Am J Ophthalmol* 1994 Dec 15; 118(6): 766-80
3. **Klein R, Klein BE, Jensen SC, et al** – Retinal emboli and stroke: the Beaver Dam Eye Study. *Arch Ophthalmol* 1999 Aug; 117(8): 1063-8
4. **Schmidt D, Schumacher M** – Stage-dependent efficacy of intra-arterial fibrinolysis in central retinal artery occlusion (CRAO). *Neuro-ophthalmology* 1998; 20: 125-141.
5. **Weber J, Remonda L, Mattle HP, et al** – Selective intra-arterial fibrinolysis of acute central retinal artery occlusion. *Stroke* 1998 Oct; 29(10): 2076-9
6. **Zweifler RM** – Management of acute stroke. *South Med J* 2003 Apr; 96(4): 380-5