AMAUROSIS FUGAX

RODICA LASCU[1]

Clinical Emergency County Hospital Sibiu

Keywords: *Amaurosis fugax, arterial emboli, ocular hypoperfusion, arterial vasospasm*

Abstract: *Amaurosis fugax (Latin fugax meaning fleeting, Greek amaurosis meaning darkening, dark, or obscure) is a transient monocular visual loss.In 1990, the causes of amaurosis fugax were better refined by the Amaurosis Fugax Study Group, which has defined five distinct classes of transient monocular blindness based on their supposed cause: embolic, hemodynamic, ocular, neurologic, and idiopathic. Concerning the pathology underlying these causes (except. idiopathic), "some of the more frequent causes include atheromatous disease of the internal carotid or ophthalmic artery, vasospasm, optic neuropathies, giant cell arteritis, angle-closure glaucoma, increased intracranial pressure, orbital compressive diseaseand blood hyperviscosity or hypercoagulability. Amaurosis fugax este o scădere vizuală monoculară tranzitorie.*

Cuvinte cheie: *amauroza fugace, embol arterial, hipoperfuzarea oculară, vasospasm arterial*

Rezumat: *Amauroza fugace (fugax latin are sensul de efemer, amaurosis în greacă înseamnă întunecare, închisă la culoare sau obscur) este o scădere vizuală monoculară tranzitorie. În 1990, cauzele amaurozei fugace au fost stabilite de Grup Studiu Amaurosis Fugax, care defineşte cinci cauze ale orbirii monoculare tranzitorii: embolic, hemodinamic, ocular, neurologic şi idiopatic. În ceea ce priveşte patologia care stă la baza acestor cauze(cu excepţia idiopatică) unele dintre cele mai frecvente cauze includ boala ateromatoasă a arterei carotide interne sau oftalmice, vasospasm, neuropatii optice, arterita cu celule gigant, glaucomul cu unghi închis, creşterea presiunii intracraniene, boala compresivă orbitală şi hipervâscozitate sau hipercoagulabilitate.*

SCIENTIFICAL ARTICLE OF BIBLIOGRAPHIC SYNTHESIS

Amaurosis fugaux – General characters

The transient monocular visual loss occurs due to a reduction in retinal artery, ophthalmic artery or ciliary artery blood flow, leading to a decrease in retinal circulation which, in turn, causes retinal hypoxia.Emboli are described as coming from an atherosclerotic carotid artery, any emboli arising from vasculature preceding the retinal artery, ophthalmic artery, or ciliary arteries.

- Atherosclerotic carotid artery: Amaurosis fugax may present as a type of transient ischemic attack (TIA), during which an embolus unilaterally obstructs the lumen of the retinal artery or ophthalmic artery, causing a decrease in blood flow to the ipsilateral retina. However, a severely atherosclerotic carotid artery may also cause amaurosis fugax due to its stenosis of blood flow, leading to ischemia when the retina is exposed to bright light.Unilateral visual loss in bright light may indicate ipsilateral carotid artery occlusive disease and may reflect the inability of circulation to sustain the increased retinal metabolic activity Atherosclerotic ophthalmic artery: Will present similarly to an atherosclerotic internal carotid artery.
- Cardiac emboli: Thrombotic emboli arising from the heart may also cause luminal obstruction of the retinal, ophthalmic, and/or ciliary arteries, causing decreased blood flow to the ipsilateral retina; examples being those arising due to atrial fibrillation, valvular abnormalities including post-rheumatic valvular disease, mitral valve prolapse, and a bicuspid aortic valve, and atrial myxomas.
- Temporary vasospasm leading to decreased blood flow can be a cause of amaurosis fugax. Generally, these episodes are brief, lasting no longer that five minutes. These vasospastic episodes are not restricted to young and healthy individuals. Observations suggest that a systemic hemodynamic challenge provokes the release of vasospastic substance in the rentinal vasculature of one eye.

- Giant cell arteritis: Giant cell arteritis can result in granulomatous inflammation within the central rentinal artery and posterior ciliary arteries of eye, resulting in partial or complete occlusion, leading to decreased blood flow manifesting as amaurosis fugax. Amaurosis fugax caused by giant cell arteritis may be associated with jaw claudication and headache but it is possible for these patients to have no other symptoms.

- Malignant hypertension can cause ischemia of the optic nerve head leading to transient monocular visual loss.
- Drug abuse-related intravascular emboli
- Iatrogenic: Amaurosis fugax can present as a complication following carotid endarterectomy, carotid angiography, cardiac catheterization, and cardiac bypass.

Ocular origin of the Amaurosis fugax:

- Optic disc drusen
- Posterior vitreous detachment
- Closed-angle glaucoma
- Transient elevation of intraocular pressure
- Orbital hemangioma
- Orbital osteoma

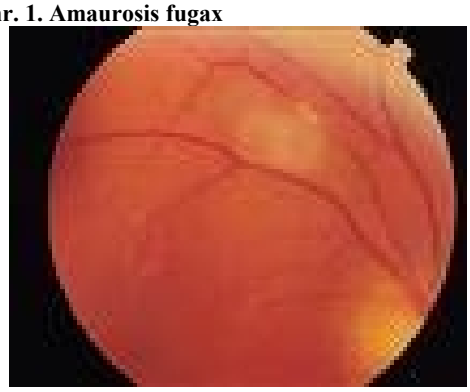
Neurologic origin of the Amaurosis fugax:

[1]Corresponding Author: Rodica Lascu, Aleea Infanteriştilor street, Ist B Flat, IIIrd Floor, Ap.25, Sibiu, România; e-mail: lascughrodica@yahoo.com; tel +40-720547341

Article received on 03.05.2010 and accepted for publication on 14.05.2010
ACTA MEDICA TRANSILVANICA September 2010; 2(3)200-202

- Optic neuritis
- Compressive optic neuropathies

Figure nr. 1. Amaurosis fugax



- Papilledema: "The underlying mechanism for visual obscurations in all of these patients appear to be transient ischemia of the optic nerve head consequent to increased tissue pressure. Axonal swelling, intraneural masses, and increased influx of interstitial fluid may all contribute to increases in tissue pressure in the optic nerve head. The consequent reduction in perfusion pressure renders the small, low-pressure vessels that supply the optic nerve head vulnerable to compromise. Brief fluctuations in intracranial or systemic blood pressure may then result in transient loss of function in the eyes." Generally, this transient visual loss is also associated with a headache and optic disk swelling.
- **Multiple Sclerosis can cause amaurosis fugax due to a unilateral conduction block, which is a result of demyelination and inflammation of the optic nerve, and "...possibly by defects in synaptic transmission and putative circulating blocking factors."**
- Migraine
- Pseudotumor cerebri
- Intracranial tumor
- Psychogenic

TMVL caused by vascular arterial ischemia

TMVL most often results from impaired perfusion in the ophthalmic, retinal (central or branch retinal arteries), choroidal (posterior ciliary arteries), or optic nerve (posterior ciliary arteries) circulation (Figs. 4-6 and 4-7)

There are three main mechanisms responsible for episodes of vascular arterial TMVL. They comprise

1. arterial emboli that originate in proximal arteries or the heart (usually to the ophthalmic artery, central retinal artery or its branches),
2. ocular hypoperfusion secondary to hemodynamic impairment (stenosis or occlusion of the aortic arch, carotid or ophthalmic arteries, reduced cardiac output or systemic hypotension),
3. arterial vasospasm (usually involving the central retinal artery). Each of these mechanisms may occur separately or in association with each other. The characteristics of the episode of TMVL and the fundus appearance help characterize the mechanism (Figs. 4-8, 4-9, 4-10, 4-11, 4-12, and 4-13).

1 Retinal Emboli

TMVL was first linked to retinal arterial emboli 50 years ago when white fragments were observed by ophthalmoscopy to travel through the retinal arterial vessels during episodes of TMVL. These emboli originate most often from an atherosclerotic plaque at carotid bifurcation (Fig. 4 – 14) and less commonly from the aortic arch or ophthalmic artery (fig. 4 – 15). Patients with this symptom typically complain of TMVL that lasts a few minutes at most.

2 Anterior Circulation Stenosis

Severe stenosis of the carotid or ophthalmic arteries or stenosis of the aortic arch (in severe aortic arch atherosclerosis or Takayasu arteritis) may cause TMVL by hypoperfusion rather than embolism.

3 Hypotension

Reduced cardiac output or systemic hypotension may also produce TMVL. Although TMVL is not typically an isolated symptom of systemic hypotension, which generally also causes lightheadedness, confusion, and binocular visual loss, the combination of drop in systemic blood pressure and asymmetric anterior circulation stenosis may cause TMVL alone, particularly orthostatically induced TMVL.

4 Chronic Ocular Hypoperfusion

Chronic ocular hypoperfusion of any mechanism may be associated with transient but prolonged visual loss (several minutes to hours) and positive visual phenomena. It may be induced by situations that further decrease perfusion pressure (postural change) or increase retinal oxygen demand (exposure to bright light). Borderline ocular perfusion may not be able to maintain retinal metabolic activity when blood flow is diverted to other tissues as after eating a meal or during exercise. Chronic hypoperfusion of the eye may also induce delay in the regeneration of visual pigments in the photoreceptor layer of the retina, resulting in blurred or absent vision that persists until regeneration of visual pigment occurs. Impaired dark adaptation may be a consequence of this phenomenon. In these cases, examination often shows venous stasis retinopathy or the ischemic ocular syndrome (dilated retinal veins, retinal hemorrhages, retinal or iris neovascularization, ocular hypotony or hypertony, anterior chamber cells and flare, cataract, and corneal edema) (Figs 4-11 and 4-12)

5 Other Causes

Less common causes of TMVL are vasculitis and radiation toxicity. Giant cell arteritis commonly causes TMVL by compromising the optic nerve circulation, more commonly than the retinal arterial low. TMVL from isolated choroidal ischemia is rare and should point to a vasculitic process such as giant cell arteritis (Fig. 4-13). TMVL is rarely a premonitory sympthom of ischemic optic neuropathies. In those cases, arteritic (rather than nonarteritic) ischemic optic neuropathy should be suspected.

Idiopathic TMVL in Young Individuals (Vasospasm)

Young people who have no evidence of vasculopathy may have episodes of TMVL secondary to reversible vasospasm of retinal arteries. Rare case reports have documented this phenomenon. Such vasospasm may be the basis for the TMVL of so – called retinal migraine, which remains a debated entity. In listening to the patient`s history, it is impossible to distinguish TMVL as an isolated symptom of vasospasm from TMVL of other causes. There-fore, vasospasm should remain a diagnosis of exclusion.

6 Natural History of TMVL

The natural history of patiens with TMVL depends on the age of the patient and the etiology of the TMVL (Table 4-2)

7 Retinal Stroke

A major adverse outcome is persistent visual loss, mostly resulting from branch or central retinal artery occlusion (Figs. 4-15 and 4-16). Besed on several natural history studies, the aggregate risk of permanent ipsilateral visual loss is about 1 % to 2 % per year.

8 Cerebral hemispheric stroke

TMVL may also herald a cerebral infraction (Fig. 4-17). When carotid occlusive disease is related to atherosclerosis, TMVL is a marke of systemic atheromatous disease and is associated a higher risk of vascular death.

The NASCET study showed a 25% 3 year risk of

stoke in patients with hemodynamically significant carotid stenosis causing ipsilateral TMVL, cerebral hemispheric transient ischemic attack (TIA), or mild stroke. However, the risk of stroke doubles in patients presenting with a hemispheric TIA compared with an episode of TMVL (see later).

Death

The risk of death in patients with TMVL and atheromatous carotid stenosis is approximately 4% per year, mainly related to myocardial infarction. Patients with retinal and hemispheric TIAs are equally vulnerable.

These data suggest that TMVL is a marker for systemic arteriosclerosis and should prompt immediate comprehensive patient evaluation

REFERENCES

1 Fisher C: Transient monocular blindnes versus amaurosis fugax. Neurology 1989; 39:1622-1624.
2 Gautier JC: Amaurosis fugax. N Engl J Med 1993; 329: 426-428.
3 Kidd PD, Newman JN, Biousse V: Neuro-Ophthalmology, Butterworth-Heinemann, Oxford 2008: 1-44, 94-134, 332.
4 Biousse V, Newman JN: Neuro-Ophthalmology Illustrated. Thieme, New York, 2009: 1-27; 41-75; 146-155; 194-206; 256-260.