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Case Report Open Access

Amaurosis Fugax and Cycloplegia in an Adolescent

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Abstract

Amaurosis fugax (AF), mainly occurring in adults, is defined as transient monocular loss of vision lasting from seconds to minutes and may be recurrent caused vascular insufficiency, and at timed referred to as retinal migraine. Searching the literature, the diagnosis of AF was based on the patients' reports that could not be examined during the acute short- lived events. We present herein a unique case of a 14-year-old male with recurrent AF attacks in whom we able to examine during the acute event. He started suffering from acute events of right monocular blindness for the last six months lasting up to five minutes at a frequency of 2 events per months. No preceding sensory or motor symptoms were experienced with no accompanying headaches following. No family history of headaches was detected. On examination of these acute events he lost his vision instantly and we observed marked dilatation of the right pupil without any response to light. The event of AF lasting commonly for up to 5 minutes and while he regained vision the right pupil was reactive again. The patient underwent a thorough investigation including MRA + MRA, echo-doppler of the carotid arteries, cardiac angiography and coagulation tests, all of which were normal. Therefore, no treatment or further investigations were suggested. On follow-up, he reported of still having episodes of acute monocular blindness which became less frequent and much shorter, not imparing his daily life activities. In conclusion, the combination of monocular blindness and cyclopgegia of the involved eye suggests hypoxic impairment of the retinal artery implying that AF along with cycloplegia during the acute event may be an unusual presentation of retinal migraine even in the absence of following pulsating headaches.

Keywords: Amaurosis fugax; Monocular blindness; Cycloplegia; Adolescence

Introduction

Amaurosis fugax (AF), mainly occurring in adults, is defined as transient monocular loss of vision lasting from seconds to minutes [1,2] usually reported in middle aged & elderly people. It is associated in them with atherosclerotic process of the ipsilateral internal carotid artery [3] representing in impending stroke [4]. The diagnosis of AF was mainly based on the patients' reports that could not be regularly examined during the acute short- lived episodes. Commonly, they described a "mosaic" or "jigsaw" pattern of isolated scotoma that progressively enlarged resulting in complete visual loss.

The amaurosis fugax study group [1] considered various multiple deficiencies other than cerebro-vascular associated with AF including coagulation and immunological reasons. However, a large bulk will remain undiagnosed. A possible additional mechanism to the evolution of AF is a reversible vascular process of a single retinal artery defined as retinal migraine occasionally associated with pounding headaches and accompanied with migraine headaches in family members [4,5].

AF is uncommon under the age of 40 years [6-8] and is especially very rare in the pediatric age group of whom the underlying etiology for the AF may be different and most patients remain undiagnosed. The early study by Poole et al. [7] in 1987 reported on 16 patients of whom only two were under the age of 20 years. Appelton et at [8] in 1988 reported on 5 teenagers with AF, speculating that AF in adolescents may be a migraine variant given extensive non- diagnostic investigations. The report by O'Sullivan et,al. [9] in 1992 on 9 patients with AF of whom three were adolescents. Ever since, no cumulative series of AF in people younger than 40 years were reported and especially no reports during adolescence except for a 15-year-old male reported in 2008 having retinal vasospasm secondary to rheumatic microemboli [10].

We present herein a unique case of a 14-year-old male with recurrent attacks of transient left – eye complete blindness along with cycloplegia.

Case Report

A 14- year-old boy presented to the emergency room at the mayer Children Hospital, Rambam Medical Center, Haifa, Israel with recurrent over the previous two months during which he suddenly loses sight in his right eye, & sees "black". These episodes last between 5-10 minutes not accompanied by any other symptom, including headache or pain in the affected eye. Thereafter, he gradually regains full eyesight and during that period admits to see double images.

He was born following normal pregnancy and delivery. His overall developmental milestones were all intact. Family history for headaches and migraine was negative as well as for transient ocular blindness.

On admission, physical, neurological and ophthalmologic examinations were all normal. During the hospitalization he again experienced an episode of acute eight-eye full blindness lasting for approximately 10 minutes, of which we could examine him. Examination during the attack of AF: the right pupil did not respond to direct light & remained dilated. This pupil did constrict in response to light directed to the contralateral eye, namely a relative puppilary

afferent defect (RAPD). Total right eye blindness and last for approximately five minutes gradually abated thereafter and disappeared parallel with recrudescence of the reactivity of the right pupil. A repeat ophtalmologic examination shortly the culmination of AF was unremarkable and no RAPD could be observed.

A diagnostic work- up to discern an underlying etiology included: A thorough laboratory work-up included a complete coagulation profile which was normal, an immunologic survey including anticardiolipin antibodies, anti-dsDNA Ab, ANAwhich were all negative with normal immunoglobulins. Cardiac echocardiography to disclose possible vegetations was unremarkable. Imaging studies included Doppler sonogram of carotid arteries with no evidence of any pathological hemodynamic interferance. CT Angiographie of brain & neck arteries was normal. Following cerebral MRI + MRA investigations were also unremarkable.

Following an unrevealing thorough diagnostic work-up no any treatment was offered and no further examinations were suggested. On follow-up asessment he reported of having episodes of acute monocular blindness which became less frequent and much shorter [6,11].

Discussion

The amaurosis fugax study group [2] divided the causes of adult AF into embolic, hemodynamic, ocular, neurologic and idiopathic. Within the ocular etiologies, anterior ischemic optic neuropathy and central artery or vein occlusion are included. Within the neurological etiologies, migraine is included. Retinal migraine with constriction of the retinal artery including visual symptoms commonly associated with pounding headaches. Anterior ischemic optic neuropathy that usually affects older patients with multiple atherosclerotic risk factors (e.g hypertension, diabetes mellitus), Increased viscosity may occur in polycythemia vera, leukemia, lymphoma, dysproteinemia. Despite thorough current investigations, as many as 40% of affected patients at any age of presentation will remain undiagnosed, as is the case of the adolescent reported here.

Searching the literature only approximately 11 pediatric cases were reported, most of them adolescents, and in most the underlying etiology could not be detected despite extensive investigations [6-9]. Poole at al [7] reported on a 5 – year –female with recurrent episodes of AF who was the daughter of a 23-year-old male with AF and another patient noted at the age of 13 years to have AF with recurrent episodes thereafter occurring up to 10 episodes per year. In both, an underlying etiology was not detected despite extensive investigations including carotid angiography. Appelton et [8] reported on 5 adolescents at age 15 - 17 years with recurrent episodes of transient monocular blindness referred for examination 8-12 months after the episodes had started. No precipitating factors were identified. The maximal duration of the visual loss was between 5-10 minutes in 4 patients and unusually lasted 20-30 minutes in 1 patient. Although the attacks were not accompanied by headaches or other features of migraine, four patients had either migraine at other times or had a positive family history of migraine. Extensive investigations were unremarkable including carotid angiograms which were normal performed in all four patients examined and no other source of emboli or evidence of hematological or connective tissue disorder could be detected. As such, given a family history of migraine, or migraine in the patient himself, the authors suggest that the etiology of AF in their patients ischemia of the retinal circulation leading to progression of visual loss from the center to the periphery of the visual field. They conclude that the paroxysmal, transient and recurrent nature of the episodes suggest that they may represent a form of ophthalmic migraine. They also state that in contrast to the outcome in adults, in whom AF is associated with increased incidence of stroke along with retinal artry embilsm of occlusion, the outcome in adolescents with idiopathic AF is excellent. The last reported series of AF by O'Sullivan et al. [8] on 3 patients during the pediatric age group at the age of 11, 14 15 years respectively having recurrent episodes of AF commonly lasting for 2-7 minutes. Doppler ultrasound of the carotid arteries was normal in all patients. The AF episodes faded and disappeared in most of their patients with no any report of permanent visual loss. As for an underlying etiology, the authors suggest that AF in young patients may be a migranous variant involving the choroidal and retinal circulation. Overall, they conclude that natural history of AF in young patients is benign and commonly the clusters will abate and ultimately resolve.

The latest patient reported recently [9], developed transient monocular blindness possibly attributed to retinal artery vasospasm and microemboli to the artery secondary to inadequately treated streptoccal carditis.

The patient reported here suffered from recurrent episode of transient left eye visual loss for the last two years that were initially frequent and disturbing but became less frequent and significantly shorter during the last 6 months. Previously he was completely healthy with no history of headaches and family history was negative for headaches or migraine. He underwent extensive up to date hematological and radiolodical investigations which were all normal. These included normal MRI of brain and magnetic resonance angiography, normal Doppler of the carotid arteries as well as normal thorough coagulation studies. As such, the underlying etiology of the recurrent episodes of AF in this adolescent as in others in whom most of they were idiopathic, are open to speculations. One could argue that the previous studies were reported before the era of MRI and MRA but arterial angiographies in those adolescents, A particularly curate measure, were all normal. Most articles dealing with AF record the history from the patients and only few succeed to observe the actual episodes as we were managed to observe in our patient and neurologically examine him. During the acute event of complete blindness we observed a relative puppilary afferent defect and cycloplegia of the left pupil which gradually resolved parallel to the recrudescence of vision.

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