Chapter 5

# An Adolescent with Headache

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## Case History

This 12-year-old boy has a 3-year history of recurrent headache. The headache had a seasonal aggravation, worsening during the autumn and spring to more than two attacks per month, with only one attack per month in the summer. In the last 3 months, he reported an increase in frequency to one attack each week.

The head pain was bilateral, with frontal localization and rare involvement of the whole head. The headache commonly began as an awareness of a discomfort in the head that in a short time became painful, starting as a mild ache and increasing to moderate or severe intensity.

The pain was pulsating and throbbing to the beat of his heart and lasted for 5 to 6 hours. Most attacks began in the morning, during school, or at the end of a stressful day. Photophobia and phonophobia accompanied all the attacks, and nausea was present in about 50%. The head pain was aggravated by physical activity of any form.

Once a month, he described episodes of losing vision and transient amaurosis in the right eye. He would see a black spot starting at the center of his vision, expanding in size, and gradually involving the entire visual field, accompanying the head pain. These symptoms lasted for only 2 to 3 minutes. Usually, the headaches started during these visual symptoms. Very rarely, these visual symptoms began 10 minutes after the headaches started.

Furthermore, he described episodes of visual impairment with the same clinical characteristics without head pain, with a frequency of one episode in a month.

At the beginning, the patient had a good response to acetaminophen at a dose of 1,000 mg suppositories (patient weight: 55 kg). Acetaminophen was never effective for the visual symptoms. In the last few weeks, there has been no response to acetaminophen.

When taking his medical history, we noted hyperreactivity syndrome (irritability, infant colic, prolonged crying bouts) which occurred at 6 months of life, and recurrent abdominal pain at age 6 years. The family history revealed only that the mother has suffered from migraine without aura since adolescence.

## Questions on the Case

**Please read the questions, try to answer them, and reflect on your answers before reading the authors' discussion.**

- Is this a primary headache disorder?
- Which laboratory testing do you think could be useful in this case?
- Would you consider prescribing a preventive treatment? Which one?
- Which acute-care treatment should be tried next?

## Case Discussion

### Investigations

As with many neurologic problems, we believe that the key is to evaluate the whole patient rather than the isolated symptom.

The evaluation for amaurosis fugax is similar to that for transient ischemic attacks or stroke. However, an ophthalmologic examination was first done. Cardiac examination was performed to search for a source of emboli or to assess underlying cardiac disease. Blood tests, including erythro-

cyte sedimentation rate, and tests for prothrombotic state in this younger patient were performed. Imaging tests began with noninvasive carotid studies. Magnetic resonance imaging (MRI) or computed tomography scan can be done to rule out a clinically silent cerebral embolism.

We began with a complete physical and neurologic examination, which did not show any abnormalities. The ophthalmologic examination did not reveal any sign or symptom of vascular disease. Electroencephalogram (EEG) and visual evoked potential (VEP) tests were done and found to be normal. All blood tests, including erythrocyte sedimentation rate, tests for prothrombotic state, and antinuclear antibody test were normal.

Although cardiac examination was normal, in order to exclude any abnormalities of the cardiac and carotid systems, we decided to perform electrocardiogram, cardiac, and carotid ultrasound examinations, which were all normal. Brain MRI, done to exclude focal lesions, was normal.

The negative findings in the above investigations clearly suggest primary headache.

### Diagnosis

Clinical characteristics of the patient suggest multiple diagnoses:

1. Migraine without aura (code 1.1 according to *The International Classification of Headache Disorders*, 2nd ed). All the clinical features in our case are consistent with this diagnosis.
2. Retinal migraine (code 1.4 according to *The International Classification of Headache Disorders*, 2nd ed). Consistent with this diagnosis, our patient had fully reversible monocular negative-visual phenomena confirmed by the patient after instruction. He had normal ophthalmologic examination and we did not find other causes of amaurosis fugax. The headaches usually started during these visual symptoms. Rarely, the patient reported the visual symptoms after the headache began, but this is not considered in the International Headache Society classification.

Migraine is a common disorder in the differential diagnosis of transient monocular blindness, especially in the age group of our patient.

Amaurosis fugax refers to transient, reversible loss of vision, typically monocular and of short duration. With its many causes, it can be a benign occurrence or can represent a life-threatening emergency.

Since the examination is often normal in patients with transient monocular blindness, the history is critical in determining optimal patient management.

The specific pattern of the visual loss can be helpful. The classic "shade" coming down over the course of several seconds marks the etiology as being vascular. Although the exact mechanism of the "shade" is unknown, it is strongly indicative of thromboembolism. Other important causes of transient monocular blindness are the transient visual obscurations of raised intracranial pressure or those associated with optic disc anomalies, narrow angle glaucoma, and corneal surface problems.

## Management Strategies

First of all, we stopped the acetaminophen because it was not effective. As the other common analgesic agents were ineffective, and the patient and his family had no cardiovascular disease or hypertension, we decided to use sumatriptan nasal spray as acute treatment at a 10 mg dose, with excellent efficacy on the migraine pain. (Editorial note: In the United States, the European Union, and Canada, only 5 and 20 mg sumatriptan nasal sprays are available, and no triptan is recommended to be given to patients below the age of 18 years. Some doctors do use it in adolescents and even younger children off-label, and there are few reported adverse events.)

We decided to treat the patient preventively with a calcium channel blocker, due to the high frequency of attacks in the last few months. We used flunarizine at the dosage of 5 mg daily, in the evening. We obtained an important reduction of frequency (to less than one attack per month). The drug was stopped after 3 months as we try to avoid problems with the neuroendocrine axis.

The flunarizine was effective for the patient's visual symptoms too.

## Case Summary

The patient was a 12-year-old boy with a history of migraine without aura and retinal migraine. Amaurosis fugax was the key symptom of this case. With its many causes, it can be a benign occurrence or can represent a life-threatening emergency. Furthermore, amaurosis fugax is quite rare in this age group.

For these reasons, we performed complete physical, neurologic, cardiac, and ophthalmologic examinations, EEG and VEP tests, blood tests, and an MRI scan. All of these were normal.

Acetaminophen was no longer effective. In the absence of cardiovascular disease and hypertension, we decided to use sumatriptan nasal spray for migraine without aura attacks, with efficacy and good tolerability. Flunarizine was used as the preventive medication with success.

## Overview of Amaurosis Fugax

The differential diagnosis of transient monocular visual loss includes several entities. Microembolic disease originating from the heart or the carotid–ophthalmic sys-

tem is the most common cause in older individuals. It is well known that emboli can temporarily lodge in the central retinal artery and cause amaurosis fugax. These retinal emboli may be visualized on ophthalmologic examination as Hollenhorst plaques in the arterioles composed of cholesterol components, or they can be fibrin or platelet emboli, calcific emboli, or foreign intravascular material.

In young patients, fibromuscular dysplasia of the carotid artery can be a source of thrombosis and/or embolization. Systemic vasculitis may cause amaurosis fugax, but are rarely the presenting symptoms of these multisystem disorders. Hypercoagulable states, including the antiphospholipid-antibody syndrome, have been described in many young adults with retinal infarction. In these patients, amaurosis fugax can be the only symptom of this coagulopathy.

A less common cause of amaurosis fugax is vasospasm. It has been suggested that vasospasm is due to altered responses to vasoactive substances that are released by platelets. Another uncommon cause of amaurosis fugax is hypotension. Hypotension usually causes transient bilateral visual loss. Unilateral symptoms may occur if there is preexisting asymmetric, occlusive vascular disease.

Finally, a growing body of evidence suggests that migraine, or a migraine-like vasospastic disorder, may be the most common cause of amaurosis fugax in younger patients. In favor of this etiology are the following:

- The frequency of headache during episodes, which is different in carotid embolic disease, which is usually painless
- The rarity of permanent visual loss or other neurologic deficits when compared to patients with carotid embolic disease
- The favorable response of some patients to calcium channel blockers
- The rarity of observed retinal emboli
- The numerous well-documented observations of retinal vasospasm during episodes
- The high frequency of personal and family history of migraine

We conclude that amaurosis fugax occurring in younger patients is probably associated with a more benign clinical course than that seen in older persons, and that migraine is a likely cause for the episodes of visual loss in a majority of this group.

## Selected Readings


Adams HP, Buder MJ, Biller J. Nonhemorrhagic cerebral infarction in young adults. Arch Neurol 1986;43:793–6.

Blau JN, MacGregor EA. Retinal migraine. Lancet 1993;342:1185.

Bruno A, Corbett JJ, Biller J, et al. Transient monocular visual loss patterns and associated vascular abnormalities. Stroke 1990;21:34–9.

Evans RW, Daroff RB. Expert opinion: monocular visual aura with headache: retinal migraine? Headache 2000;7:603–4.

Guidetti V, Moscato D, Ottaviano S, et al. Flunarizine and migraine in childhood. An evaluation of endocrine function. Cephalalgia 1987;7:263–6.

Guidetti V, Ottaviano S, Pagliarini M. Childhood headache risk: warning signs and symptoms present during the first six months of life. Cephalalgia 1984;4:236–42.

Guidetti V, Russell G, Sillanpää M, Winner P, editors. Headache and migraine in children and adolescents. London: Martin Dunitz; 2002.

Headache Classification Subcommitee of the International Headache Society. The international classification of headache disorders: 2nd ed. Cephalalgia 2004;24 Suppl 1:1–160.

Inan LE, Uysal H, Ergun U, et al. Complicated retinal migraine. Headache 1994;34:50–2.

Gupta VK. Visual function impairment in migraine: cerebral versus retinal deficit. Cephalalgia 1993;13:431–3.

Levine SR, Deegan MJ, Futrell N, Welch KM. Cerebrovascular and neurologic disease associated with antiphospholipid antibodies: 48 cases. Neurology 1990;40:1181–9.

O'Sullivan F, Rossor M, Elston JS. Amaurosis fugax in young people. Br J Ophthalmol 1992;76:660–2.

Pandit JC, Fritsche P. Permanent monocular blindness and ocular migraine. J R Soc Med 1997;12:691–2.

Rumi V, Angelini L, Scaioli V, et al. Primary antiphospholipid syndrome and neurologic events. Pediatr Neurol 1993;9:473–5.

Sjaastad O. Transitory isolated, global blindness and headache the possible relationship to migraine. Funct Neurol 1986;1:467–71.

Slavin ML. Amaurosis fugax in the young. Surv Ophthalmol 1997;41:481–7.

Solomon S. Migraine variants. Curr Pain Headache Rep 2001;5:165–9.

Tippin J, Corbett JJ, Kerber RE, et al. Amaurosis fugax and ocular infarction in adolescents and young adults. Ann Neurol 1989;26:69–77.

Winterkorn JM, Kupersmith MJ, Wirschafter JD, et al. Brief report: treatment of vasospastic amaurosis fugax with calcium-channel blockers. N Engl J Med 1993;329:396–8.


## Editorial Comments

This very interesting case of migraine presenting with episodes of amaurosis fugax is carefully worked up and evaluated by Professor Guidetti and colleagues. Although most younger migraineurs do not have such unusual visual symptoms, it is very helpful to see yet another way that migraine can worry us clinically. We would like to point out that although the judicious use of triptans in children under 18 years of age is often done around the world, it is not currently recommended in the United States, the European Union, and Canada, and in many other countries.

Final diagnosis:

Migraine with amaurosis fugax