Amaurosis fugax in young people

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Abstract
Nine young adults (median age 19-5 years) who suffered from amaurosis fugax (AF) are described. The attacks of AF were short in duration and preceded by premonitory symptoms in five cases and by a migrainous headache in two. In five patients the visual loss progressed in a lacunar pattern unlike the 'curtain' pattern characteristic of AF in older patients. Investigation revealed no evidence of an embolic or atheromatous aetiology. In two cases a minor abnormality was found on echocardiography. We conclude that AF in young adults has a different clinical pattern and may have a different aetiology, possibly migrainous, compared with that seen in older patients. The pattern of visual loss in some of the cases suggests that the choroidal circulation rather than the retinal circulation is primarily affected. (Br J Ophthalmol 1992; 76: 660-662)

Amaurosis fugax (AF) is defined as transient monocular loss of vision lasting seconds to minutes and is attributed to ischaemia or vascular insufficiency.1 The impairment progressively may involve the entire visual field; rarely the visual loss lasts for several hours but full recovery of vision occurs. It is usually reported in middle aged and elderly people. A high incidence of ipsilateral carotid atheromatous disease has been found in these patients and fibrin platelet emboli from the carotid or from the heart are thought to be responsible.2,3 Treatment with aspirin may be effective in preventing further attacks.4,5 We have seen a group of young patients who reported a similar symptom; we define the clinical features of AF in young people and speculate on its pathophysiology.

Patients and methods
Nine patients presenting to the ophthalmic casualty department with AF were studied prospectively. The median age at onset was 19-5 years, range 11-30 years. There were six males and three females in the group and the average duration of follow up was 2-8 years.

HISTORY
The relevant features of the clinical history are summarised in Table 1. The history of AF described was quite distinct. The duration of AF was short; the median duration was 5-2 minutes (range 1-30 minutes). Five patients reported premonitory symptoms; these were tingling, numbness, or pain around the eye subsequently affected and preceded the AF by 2 minutes on average (range 30 seconds-15 minutes). In two cases the patients reported a typical migrainous headache preceding the AF.

The median number of attacks was 10-5 (range 4-21). The attacks occurred in clusters over days or weeks in seven of the nine patients and then faded in frequency (see Fig 1). In two cases the attacks occurred at regular intervals.

The pattern of visual loss described by five of the patients was quite different from the classic 'curtain' effect reported in older patients with AF. These patients reported that the vision was lost in a series of 'blobs' or lacunae throughout the field of vision which gradually coalesced into a complete loss of field. After a few minutes the vision returned in a reverse pattern with patches of vision eventually enlarging to a full field (see Figs 2-5). In the other four patients, three reported a non-specific blurring of the field of vision whilst one gave a history of a curtain effect.

As well as the migrainous headaches which preceded the AF in two patients, four other patients complained of migraine headaches at other times and eight of the nine patients gave a family history of migraine.

CLINICAL EXAMINATION
It was not possible to examine the patients during the attacks of AF. In four cases, relatives observed the patient during the attacks and noticed pupil dilatation in the affected eye. This resolved as the vision returned.

On clinical examination the positive findings were:
(1) A systolic murmur consistent with tricuspid and pulmonary regurgitation in a 14-year-old boy who had Crouzon's disease.
(2) Fundus changes in the affected eye consistent with inactive presumed ocular histoplasmosis syndrome in a 30-year-old woman.
(3) Bruits over both carotid arteries in a 14-year-old boy which on Doppler ultrasound were attributed to high flow, consistent with the patient's age.

No other patient had a carotid bruit, irregular
pulse, or other clinical cardiovascular abnormality. No retinal emboli or infarctions were seen in any of the patients. The rest of the oculomotor examination including tonometry and funduscopy was normal. Visual field testing and examination of the nervous system were also normal in all cases.

INVESTIGATIONS

All patients had a normal full blood count, erythrocyte sedimentation rate, urea and electrolytes, serum glucose, and fasting lipids. Anti-cardiolipin antibodies were not detected in the five patients who were tested.

Doppler ultrasound of the carotid vessels and echocardiograms were performed in all cases. The results are summarised in Table 2. The only abnormal findings were on two of the echocardiograms. The 14-year-old boy who had a systolic murmur was confirmed to have pulmonary and tricuspid incompetence. A 28-year-old male patient who had no detectable clinical abnormality was found to have a patent foramen ovale on echocardiography.

Outcome

The median follow-up time of this group was 2-8 years (range 0.5-3.7). In seven of the nine cases the natural history was that of clusters of attacks over weeks to months which then faded out spontaneously. In the remaining two patients the attacks occurred more regularly either monthly or bimonthly and have persisted. No patient has suffered permanent loss of visual acuity, visual field, or any other neurological deficit.

Low dose aspirin was prescribed as therapy in five cases but appeared to have no effect on the clinical course. One woman who had regular repetitive attacks accompanied by migrainous headaches was treated with pizotifen which brought incomplete relief of her symptoms.

Discussion

Sudden transient loss of vision in young patients is relatively rare. There have been previous reports of small numbers of patients whose median ages were in the mid-20s.9,7 Our patients were younger and included two children under 14 years who gave clear accounts of visual loss.

The history elicited from five of our patients was quite distinct from that typically reported by older patients with AF. The characteristic sequence of a 'curtain' dropping over the vision was found in only one of our cases. Furthermore we did not find retinal emboli or infarcts in any of our patients and there was no evidence of atheromatous carotid disease. The patients we treated with aspirin reported no change in their symptoms and none of the group have subsequently suffered neurological or cardiac events. The absence of these findings suggested that the cause of AF in our group of patients was not carotid or cardiac thromboembolism.

There is some clinical evidence to suggest that AF in young patients may be migrainous in origin.4,9,10 Appelton et al11 call this variant of migraine where visual loss occurs without headache 'acephhalic migraine' and suggest that it has a benign prognosis in young patients.11 This study provides further evidence to support this hypothesis. Migraine is often a familial disorder,12 and eight out of nine of our patients had a personal or family history of migraine. The episodes of AF were preceded by sensory premonitory symptoms in five cases, which could have been due to ischemia from vasospasm in the territory supplied by the ophthalmic artery. The pupillary dilatation reported to us, which occurred during the attack of AF may imply involvement of the ciliary ganglion circulation in the migrainous process and has previously been reported in attacks of migraine.13 Furthermore the cluster pattern of attacks is a feature of other migrainous disorders such as cluster headaches.14

Patients with migraine have been reported to suffer from central retinal artery occlusion and ischaemic optic neuropathy.15,16 However the pattern of visual loss reported in five of our patients is best explained by a process affecting the choroidal circulation. Conner has previously reported on pigmentary changes in the fundus following migraine attacks and has suggested that the choroidal as well as the retinal circulation can be affected.17 The lacunar or 'blob' pattern of visual loss described by some of our patients may correspond to the lobular arrangement of the blood supply in the choriocapillaris.18,19

The choroidal circulation of primates has a very high blood flow6 and it is estimated that 70% of blood in the globe is within the choriocapillaris.1 The blood vessels of the choroid in primates have a plentiful sympathetic and parasympathetic nerve supply.20,21

Many of the features of migraine can be explained by vasospastic events.22 Stimulation of the sympathetic nervous supply to the choroid, which would occur during a migrainous attack, caused a 60% reduction in flow in cats.23 Reduced blood flow through the choriocapillaris would
account for visual loss, so the potential mechanism for choroidal migraine exists.

Previous reports indicated that mitral valve prolapse in young patients may be responsible for the symptom of AF, though we were unable to confirm this link. However it is worth noting that patients with mitral valve prolapse have a high incidence of migraine. In summary, we report on nine young patients with AF who in contrast to older patients with AF do not have carotid vessel disease. There is no indication for carotid angiography in the investigation of these patients. We present suggestive evidence that their symptoms may be migrainous in origin and we propose that the choroidal as well as the retinal circulation may be affected in migraine. The natural history of AF in young patients is benign and clusters of attacks tend to resolve completely.

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