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PRACTICAL NEUROLOGY

Transient Blindness

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There can be few more dramatic symptoms than sudden loss of vision. Even when blindness lasts only a short period concern is felt by both patient and doctor that the symptom may recur and persist. The practical management of each case, especially with regard to further investigations, depends on the clinical features and their diagnostic possibilities.

Blindness or "blackouts" may occur in both eyes simultaneously as a result of vasovagal attacks or in any condition in which there is a drop in systemic blood pressure affecting the ophthalmic arteries. (The term blackout was first used by pilots diving steeply during the earlier days of air transport.) Transient blindness ("amaurosis fugax") is far commoner in one eye than both. Surprisingly, monocular blindness may not be noticed unless the normal eye is closed. Not infrequently the patient may complain of loss of vision in one eye and examination reveals a homonymous hemianopia. For these reasons a careful history and a thorough examination are always essential.

Migraine

Though only about one in four patients has a visual aura, migraine is a common cause of transient loss of vision, since it affects more than 10% of the population.

A first attack of migraine starting with central scotomata can be extremely worrying (as I recently experienced when driving along Regent Street during a traffic jam). Within minutes, however, central vision becomes normal as the shimmering teichopsia spreads to the periphery, leaving the classical unilateral throbbing headache.

The visual aura usually lasts from 10 to 30 minutes and may take many different forms, including flashing or coloured lights, field defects, or fortification spectra. The ensuing headache with nausea and photophobia lasting several hours renders the diagnosis easy. In the unusual cases in which the visual phenomena occur alone, a positive family history of migraine (obtained in the majority of cases) is supportive evidence for the diagnosis, and reassurance may confidently be given. Rarely does the teichopsia last as long as an hour, but persistent field defects such as homonymous hemianopia do occur, though fortunately rarely.

Carotid Insufficiency

The commonest cause of transient blindness in one eye is carotid insufficiency. This condition has a predilection for the male sex in the second half of life and is commoner on the left side. Typically the patient describes a curtain coming from above or below or laterally and spreading to affect half or the whole field of vision within about half a minute. Occasionally the curtain may go half way up and then another curtain come down to produce complete blindness. The blindness persists for two to five minutes or longer and clears in the same way as it started. The attacks tend to be repeated,

but their severity, duration, and frequency are variable, even in the same patient. During the attack the pupil is non-reactive, the retinal vessels look narrow, and emboli may be seen travelling along the retinal arterioles. The emboli consist of either fibrin and platelets or cholesterol, the latter seen as glistening refractile bodies at the bifurcations of vessels and appearing wider than the lumen. These (Hollenhorst) bodies are evidence of atheroma in vessels leading from the heart to the ophthalmic artery.¹

Though other factors besides emboli may play a part in the transient blindness of carotid disease, e.g. decreased blood flow or retinal artery spasm, there is little doubt that this symptom is one of the commonest presentations of carotid insufficiency. As it may be a prodrome of permanent blindness or a devastating stroke (approximately 50% within 3 years) it is advisable to consider arteriography in every patient under the age of 70 years. The carotid vessels should be carefully palpated for evidence of decreased pulsation. This sign is more easily elicited in complete than partial carotid occlusion, and because the external carotid artery may be mistakenly palpated a more useful test is auscultation of the neck for a carotid bruit.

Carotid arteriography may reveal an atheromatous plaque in the first two or three centimetres of the internal carotid artery (carotid stenosis) and consideration should then be given to surgical removal of the atheromatous area (thrombendarterectomy). Surgical interference is usually contraindicated in complete occlusion of the internal carotid artery, since the thrombus usually extends into the cranial portion and attempted removal rarely results in restoration of blood flow. Repeated attacks of amaurosis fugax which suddenly stop may be due to complete occlusion of a stenotic vessel. Thrombendarterectomy does not always stop the attacks even when postoperative arteriography reveals no abnormality. Because each case has to be treated on its individual merits and since few centres have a sufficiently large number of cases it is difficult to evaluate this treatment with a controlled trial and the precise value of surgery is still not settled.

When carotid arteriography is negative this does not exclude the diagnosis of embolism, which may have arisen from the large vessels in the chest, and for this reason arch-aortography may need to be performed.

Cortical Blindness

Bilateral homonymous hemianopia is usually due to occlusion of both posterior cerebral arteries, often associated with atheroma of the basilar artery. The homonymous field defects may appear consecutively. Loss of vision is often complete at the onset but persists only for a few minutes, recovery of the central fields being the rule, so that visual acuity may become normal. Similar phenomena are sometimes seen after vertebral arteriography or ventriculography, and the blindness associated with these neuroradiological investigations usually has a good prognosis.

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In cortical blindness there is complete loss of all visual sensation including perception of light; the blink reflex—reflex lid closure in response to threatening gestures—is absent, and yet the pupillary responses to both light and accommodation are retained, and the fundus has a normal appearance.

Temporal Arteritis

At least 25% of patients with temporal (giant cell) arteritis become blind in one or both eyes. There may be warning attacks of transient blindness, and immediate treatment with steroids can save vision. The diagnosis should be suspected in any patient over the age of 55 with headache or generalized pains (polymyalgia rheumatica); if the sedimentation rate is over 50 mm. in 1 hour steroids should be given without waiting for the result of temporal artery biopsy, which is not infrequently negative for giant cells.

Retinal Artery Spasm

Periods of blindness in one eye or both eyes, lasting from one minute to several hours, can occur in hypertensive encephalopathy, uraemia, and eclampsia.² Central vision again recovers before peripheral, and it is presumed that these patients have spasm of the central retinal arteries, as may those rare patients with Raynaud's disease who occasionally have intermittent blindness. The diagnosis in these cases is usually easy, as it is in those cases of transient blindness following haemorrhage. This usually occurs between the third and seventh days after repeated haematemeses or menorrhagia, and recovery takes place within a few hours. Fundoscopy reveals pallor of the fundus, together with narrowing of the arteries and blurred margins of the disc edges. In spite of treatment to control haemorrhage and blood transfusions the blindness may persist in some cases. Though occlusion of the central retinal artery causes persistent blindness it may be preceded by transient attacks.

Central Angiospastic Retinopathy

Periodic loss of central vision of short duration lasting two to three seconds associated with orbital pain which subsides has been attributed to transient vasospasm.³ The condition is bilateral but asymmetrical in its severity. The patient may complain of the change of shape of objects—i.e., metamorphopsia. During the acute stage there is a dull grey-red circular area of macular oedema which is slightly elevated with a sharp margin, and there is loss of the foveal reflex. When there is evidence of retinal artery spasm many different treatments have been used—ergotamine, phenobarbitone, belladonna, glyceryl trinitrite—but there is little evidence that they are of value.

Papilloedema

Transient blindness is not uncommon in pronounced papilloedema and usually lasts for only a few seconds. Changes in posture such as sudden standing or lying down tend to produce it, as does nervous tension. Though no changes in the retinal artery can be seen the blindness probably results from transient ischaemia of the nerve head.⁴ These cases need urgent referral for full investigation. The longer the papilloedema has been present, the more likely the possibility of secondary optic atrophy. Patients with pituitary tumours may have a sudden onset of transient blindness which lasts from a few moments to an hour or more, probably due to bleeds into the vascular intrasellar tumour. The presence of field defects (complete bitemporal hemianopia is "classical" but exceptional) and signs of hypopituitarism (lack of axillary hair, smooth pale skin) renders a lateral radiograph of the skull mandatory (the sella will be abnormal in nearly all cases).

Transient blindness may also occur after head injury

in children. Mild blows to the frontal or occipital areas, even without loss of consciousness, may be followed by blindness within a few minutes but the vision usually returns within a few hours.

Optic Neuritis

In the vast majority of cases of optic neuritis the loss of vision is unilateral and may take hours or more usually days before it is complete. There is often a history of pain on movement of the eye with some alteration of pupillary reflexes on the affected side. Improvement in vision usually occurs within a few weeks after the onset, and the prognosis is excellent in 95% of cases.

Patients who have had optic neuritis may complain of decreased vision during a hot bath, or exercise, or following a meal (Uhthoff's sign). The same symptom is less commonly seen affecting one or both eyes in patients in whom there is no evidence of a previous attack of optic neuritis. In all cases, it is presumed that the vascular supply to the optic nerve is tenuous, so that cutaneous, muscular, or splanchnic vasodilatation produces transient ischaemia.

Other Causes

Transient loss of vision may occur in encephalomyelitis following vaccination or exanthematic fevers and in meningitis and encephalitis. The presence of other signs makes the diagnosis of these cases fairly obvious.

Hysterical blindness was commoner in wartime, but may be seen in children, especially those with dyslexia. Either or both eyes may be affected, and the onset is usually abrupt. In spite of the complaint of blindness these patients find their way about without bumping into things and there is a marked "belle indifférence." There is often contraction of the visual fields or tubular fields, and repeated visual field testing can usually catch the patient out, as it reveals marked variability. Tremor of the closed lids or blepharospasm is usual. The pupillary responses are normal, as in ophthalmoscopic examination. Recovery usually occurs within a few days to a few weeks; the condition may, however, last years, and there is a tendency to recurrence, with hysterical manifestations elsewhere.

Acute glaucoma can cause sudden onset of blindness, but there is usually pain. Treatment consists of decongestants and reassurance, giving acetazolamide 250 mg. four-hourly and 4% eserine and 2% pilocarpine eye drops. When the acute attack is over peripheral iridectomy may need to be performed to prevent further attacks.

Rarer causes of transient blindness² are anoxia, exposure to electrical flashes, arc-welding lights, electrical shock, or patients who gaze at an eclipse for a long time. A similar thing is seen in snow blindness, from which the patient always recovers. Transient blindness can complicate operations performed in the sitting position or even occasionally follow an epileptic fit, but whether hypotension or spasm of the retinal arteries is the cause is unknown.

Complete blindness, usually transitory, may occur hours or even days after chest operations owing to the accidental introduction of air into the blood stream, or after fractures of medullated bones, owing to fat emboli.

Inquiry should always be made about drugs taken, since these may produce temporary blindness—an example is extract of male fern, used in the treatment of tapeworm infestation.

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