



Unexplained Visual Loss

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One of the most vexing problems faced by ophthalmologists and neurologists is visual loss which cannot be explained by obvious abnormalities of the eye. There are basically two variations to this problem: a) subnormal acuity with normal examination and b) normal acuity but symptoms of visual loss. In this situation it is tempting to plunge into expensive laboratory and radiological investigations. However, before doing this, the neurologist must be certain that the problem has been examined logically and that the subtleties of neuro-ophthalmic examination have not been missed.

In the case of unexplained visual loss (UVL) the fundamental problem lies in separating optical problems from neuro-retinal ones. As a neurologist, I shall not discuss optical problems but would rather focus attention on the day to day problems that affect the various levels of the visual pathways.

To separate optical problems from neuro-retinal diseases the first step in the diagnostic testing is the pin-hole test. If the acuity of vision improves, it is obviously an optical problem whereas if it remains unchanged, it is a neuro-retinal problem. Once a neuro-retinal problem is established, the logical next step in the diagnostic process is the pupillary examination. For this purpose, the swinging flashlight test is performed. In a normal patient, when the flashlight is swung from one eye to the other, there is no pupillary dilatation. However, if there is any lesion in the afferent pathways, the pupil will dilate. This test is very rapid, reliable and a sensitive method of establishing an **afferent pupillary defect**, particularly in populations with dark eyes and in whom the consensual light reflex is difficult to interpret when performed in the traditional way. It separates asymmetric optic nerve disease from all other causes of UVL.

When an afferent pupillary defect accompanies visual loss, the clinician's further approach to the problem depends upon the duration of visual loss and on examination of the visual fields. In such a situation, the charting of the visual fields will reveal two basic types of defects: a nerve fibre bundle-type of defect and a hemianopic defect.

UVL WITH AFFERENT PUPILLARY DEFECT & NERVE FIBRE BUNDLE-TYPE DEFECT

This type of defect indicates a retinal or pre-chiasmal optic nerve lesion and the patterns of field defects are a) arcuate scotoma due

to involvement of the arcuate fibres secondary to ischaemic optic atrophy or glaucoma, b) altitudinal field defect due to occlusion of one of the posterior ciliary or retinal arteries secondary to atheroma in an individual with hypertension or diabetes, c) central or centrocecal scotoma due to involvement of the maculopapillary bundle secondary to demyelination, metabolic toxic optic neuropathies or infiltration of the optic nerve, d) non-specific generalised constriction of the visual fields. Data from representative studies have shown that such cases are usually non-compressive (97%). The history is important in differentiating the various causes and although improvement is the rule only a small number of patients will respond satisfactorily to therapy.

Having established a retinal or optic nerve defect, at times careful funduscopy may reveal the presence of systemic diseases which predominantly involve the CNS. In this group the main conditions are the phacomatoses, certain infections and infestations and retinitis pigmentosa. The four main syndromes included in the phacomatoses are neurofibromatosis, tuberous sclerosis, angiomas of the retina and CNS - Von Hippel Lindau syndrome and encephalotrigeminal angiomas - Sturge Weber syndrome. Syndromes added to the phacomatoses are ataxia telangiectasia, A-V aneurysms of midbrain and retina known as Wyburn-Manson syndrome, Kippel-Trenuanay-Weber syndrome, hereditary haemorrhagic telangiectasia - Osler Rendu Weber syndrome and Riley's syndrome.

CNS disorders with retinitis pigmentosa include mitochondrial cytopathies - Kearn-Sayer's syndrome, Refsum's disease, Laurence Moon Biedl syndrome, Usher's syndrome, abetalipoproteinaemia and Tangier's disease. These patients commonly have mental retardation, cerebellar ataxia, ophthalmoplegia, hearing loss and peripheral neuropathy.

Common CNS infections/infestations which can be diagnosed on the basis of fundal examination are tuberculosis, Eale's disease and neurocysticercosis.

UVL WITH AFFERENT PUPILLARY DEFECT AND A HEMIANOPIC FIELD DEFECT

With this combination, the lesion usually lies at the junction of the optic nerve with the chiasma or at the chiasma itself. At this level of the visual pathways, the field defects have sharp vertical borders. However, a marked feature is the considerable variability

in the field defect because of the anatomical variations. Some of the patterns of field defects due to this anatomical variation may give us a clue to the aetiological factor and is exemplified by the junctional scotoma. The lesions with this type of field of vision defect are usually compressive and produce progressive impairment of vision in the ipsilateral eye. These lesions are commonly pituitary adenomas, craniopharyngioma, sphenoidal ridge or sellar meningioma, chiasmal glioma, chordoma, ectopic pinealoma or massive third ventricular dilatation. These space occupying lesions usually expand slowly and headaches may be absent if the increase in the ICP is slow. Secondly, the patient may not appreciate a uni or bitemporal field defect and may be brought by their relatives for "poor eyesight" as they keep bumping into objects on either side.

UVL WITHOUT AFFERENT PUPILLARY DEFECT

In such cases the neurologist must be guided by a different set of possibilities. The main differential diagnoses lie between amblyopia, macular disease, cone-rod dystrophy, bilateral symmetrical optic neuropathy, chiasmal and post-chiasmal lesions and feigned visual loss. Amblyopia is a purely ophthalmic problem and will not be discussed here. Neurological disorders with macular degeneration include Tay-Sach's disease, Niemann-Pick disease, metachromatic leukodystrophy and neuronal ceroid lipofuscinosis. All these cases have progressive visual failure but the afferent pupillary reflex is preserved till a late stage of the disease.

If amblyopia and macular degeneration have been excluded, one must again turn to the duration of the illness and the visual fields. Normal field of vision would mean that the patient has no organic disease and that his symptoms were misinterpreted or that he has a very subtle optic nerve disorder or a cone-rod dystrophy. For subtle optic neuropathy, one may have to rely on visual-evoked potentials and for cone-rod dystrophy on colour vision. Tunnel fields are very characteristic of hysterical visual

loss or malingering. Once these conditions are excluded, one can easily locate the lesion in the post-chiasmatic area which includes the optic tract, lateral geniculate body, optic radiation and visual cortex. Isolated lesions of the optic tract and radiations are rare. In this part of the visual pathways, strokes and neoplasms form the main aetiological lesions. The clinical evaluation of a patient with a homonymous hemianopia is a common problem. It invariably indicates a structural parenchymatous lesion within the hemispheres and I have yet to see this type of a defect produced by an extrinsic compression like a subdural haematoma or a convexity meningioma. It is usually associated with neurological symptoms and signs which help in the specific location of such lesions. These patients usually have normal acuity but are brought again for poor vision as they cannot read or keep bumping into objects. Certain syndromes of higher mental functions are associated with visual dysfunction. These include alexia with or without agraphia and visual agnosia. The full spectrum of visual agnosias include cortical blindness, cortical visual distortion, disorders of colour vision, visual object agnosia and prosopagnosia.

Hence, when dealing with a patient with apparent unexplained visual loss, one must remember that there is always a logical explanation for it. The clinical strategy includes careful history taking, a high index of suspicion and basic clinical examination in order to arrive at an accurate clinical diagnosis. Expensive laboratory and radiological investigations should be used judiciously in order to confirm the clinical diagnosis and not unnecessarily because a visual loss is apparently unexplained.

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