Non-organic visual loss

Patients can present to eye departments with various signs and symptoms (mostly symptoms) with no obvious organic cause. These patients can be labelled with any of a wide range of diagnoses such as functional visual loss, functional overlay, psychosomatic reaction and malingering.

Assessment

It is important to approach such cases with an open mind and not to have any pre-conceptions in order not to miss any subtle organic disease. History of the presenting complaint, the way the patient describes their symptoms and the patient involvement and interaction with healthcare personnel are very important as they can reveal key features that help reach the correct diagnosis.

It is also important to have a level of understanding of the patient's medical, as well as social, background to know if there is any motivation for functional loss of vision.

Before diagnosing the patient with a functional visual loss, it is crucial to perform a full ocular examination and sometimes other investigations (such as OCT, neuro-imaging or electrophysiology) to confirm a normal visual pathway from the structural point of view, as well as the functional point of view.

Neuro-ophthalmology non-organic presentations

1. Afferent visual pathway: visual acuity and visual fields
2. Efferent visual pathway: ocular motility and alignment
3. Pupillary size and reaction
4. Other: such as lid position and facial sensation.

Visual acuity

It is perhaps one of the commonest referrals to neuro-ophthalmology where the level of visual acuity cannot be explained by the findings of full ocular examination. It is important as mentioned above to rule out any possible organic cause, such as refractive errors, keratoconus, mild ocular media changes and subtle maculopathies by taking a detailed history and performing a full ocular assessment. These visual complaints could be psychogenic or due to malingering.

Patients with reduced visual acuity and no obvious ocular cause are usually labelled as possible cases of optic neuropathy (despite a normal looking optic nerve), however, it is very unusual for true optic neuropathy patients to have visual loss without colour vision abnormality, so an assessment of colour vision is a key part of the assessment.

It is also important to assess pupillary reaction looking for relative afferent pupillary defect (RAPD) as the absence of it suggests that either the patient does not have a unilateral optic neuropathy or has a bilateral symmetrical optic neuropathy.

There are various features and clinical tests that should alert the physician to the functional nature of the complaints. The use of these tests depends on the level of the perceived visual loss.

Complete loss of vision: Patients tend to avoid looking into the examiner's face or wear dark glasses despite the lack of photophobia. Other tests include using the OKN drum to elucidate a nystagmus (vision at least 1.3 LogMAR) or using a large mirror in patients claiming perception of light (PL) or no perception of light (NPL) vision. It is also useful to ask such patients to sign their name or touch tips of the fingers from both hands (patients with organic loss of vision should be able to do this as it's a proprioception function).

Partial loss of vision: between 0.3 LogMAR and hand movement (HM). It is a bit more difficult to assess the true level of vision in such patients. In some, certain tests reveal no loss of vision at all (complete functional visual loss) or partial organic visual loss, where the patient's presenting vision is lower than what their organic disease can cause (functional overlay). None of these tests is fully reliable but combined they can provide enough evidence to suggest the functional nature of the condition. Tests include using prisms (four base out) to detect fixation movements, fogging the better eye while testing the other eye, using +/- lenses (to trick the patient into believing that they are given glasses), using red-green glasses with a duochrome screen, assessing patient stereopsis (tends to correlate well with visual acuity, 60" of stereopsis needs 0.3 LogMAR in the worst seeing eye), varying the distance of vision chart or assessing near vision.

Visual field loss: Various types of visual fields defects can be functional. Common examples would be generalised constriction of visual fields, cloverleaf type defects or spiralling defects on Goldmann's perimeter. It is important to remember that patients can fake any field defect with absolute reliability.

When functional defect is suspected, it is useful to perform confrontational visual field testing. The presence of tunnel vision instead of funnel vision suggests functional loss (fields should normally enlarge as the distance between patient and examiner increases).

Monocular field defects, bi-nasal and bi-temporal defects should reduce or disappear on binocular testing if the cause is organic.

It is also useful to test the patient's saccadic eye movements towards a non-seeing part of their field to show the functional nature of the defect.

Efferent pathway

Monocular diplopia is not a neuro-ophthalmic condition. The majority of these cases are secondary to refractive or ocular media abnormality and usually patients describe ghosting of the images rather than true diplopia. The true monocular diplopia is almost invariably non-organic.

Common functional presentations to the efferent pathway include:

Spasm of the near reflex: This could present with large but variable esotropia, miosis, accommodation spasm (and myopic shifts) and limitation of abduction (which need to be differentiated from a VI nerve palsy by testing ductions rather than versions). Observation of the patient, doll's head movements and assessment of ductions and looking for pupillary changes are all useful if identifying the functional nature of the condition, although some patients might need neuro-imaging to rule out mid brain lesions.

Convergence insufficiency or paralysis: It is sometimes useful to ask the patient to read a long paragraph or look up the time from their watch or mobile screen.

Paralysis of horizontal or vertical gaze: These could be assessed by chair rotation or doll's head movements.

Voluntary nystagmus or saccadic oscillations: These are usually difficult to maintain for a long time and observing the patient's face while these movements are

happening usually reveals the nature of the condition.

Pupillary reaction and size

The most common presentation is bilaterally or unilaterally dilated non-reactive pupils following installation of dilating agents. These pupils invariably do not constrict to pilocarpine 2% contrary to cases of III nerve palsy or tonic pupils.

Lid position or movement presentations

These include pseudo / voluntary ptosis, blepharospasm, lid retraction and lid apraxia.

Summary

Functional complaints are common in neuro-ophthalmology clinics. It is important to approach these cases in a systemic way to avoid missing an organic disease (Table 1). It is important to assess the structure and the function of the visual pathways and efferent pathways and take a full detailed history. It is also sometimes necessary to perform some investigations such as neuro-imaging and electrophysiology testing. Once the examiner is satisfied regarding the nature of the condition, management is usually to assure the patient and family that there is no cause for concern and discharge the patient.

References

1. Miller NR, Subramanian P, Patel V. *Walsh and Hoyt's Clinical Neuro-Ophthalmology: The Essentials*. Third edition. Lippincott Williams and Wilkins; 2015.
2. Schiefer U, Helmut W, Hart W (eds). *Clinical Neuro-Ophthalmology. A Practical Guide*. Springer; 2007.

Table 1: Common conditions that can mimic or misdiagnosed as functional visual loss.

Uncorrected or missed refractive errors or amblyopia
Mild corneal abnormalities such as keratoconus
Mild ocular media opacities
Subtle maculopathies
Paraneoplastic retinopathies
Subtle or early optic neuropathies
Hereditary optic neuropathies (LHON)
Small compressive orbital or intracranial lesions
Higher visual functions abnormalities

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