APPROACHING THE ED PATIENT WITH VISION LOSS

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HISTORY:
CHARACTERIZATION OF SYMPTOMS: In addition to standard symptom driven interview questions (onset, duration, provoking factors, relieving factors, associated symptoms etc.) probing questions regarding the visual symptoms are helpful to guide the exam and differential diagnosis:

- Severity of impairment – what specific things can they not see
- Positive symptoms (extra vision) vs. negative symptoms (visual loss)
- Movement images
- One eye or both eyes
- Central vs. peripheral vision

PHYSICAL EXAM:
VISUAL ACUITY: This is the vital sign of visual complaints. Pertinent parameters are which eye is tested, with or without glasses, at which distance (near or far) and what they can distinguish. In the absence of a calibrated chart, any visual stimulus can be used. If large text can't be discerned, ability to count fingers, identify hand motion or see light should be determined. Visual acuity that is dramatically different between the two eyes suggests a problem anterior to the chiasm.

COLOR VISION: Comprehensive tests of color vision are often not readily available in the ED setting. Some information can be gained by asking the patient to compare perception of a red object between the two eyes. Difference between the eyes is a positive finding.

VISUAL FIELD: This is the cardinal examination finding that guides neurological localization of vision loss. Unfortunately gold standard formal visual field testing that is used in the ophthalmology clinic, is not available in most neurology clinics or emergency departments, and the bedside test of confrontation visual fields has poor sensitivity.[1] However, specificity is reasonable. If detected, single eye loss localizes anterior to the optic chiasm, bitemporal visual field loss (i.e. outer field of each eye) localizes to the chiasm and homonymous defects (i.e. same field both eyes) localize to the visual pathways behind the chiasm. Neither lack of positive finding on confrontation visual fields nor lack of peripheral vision complaint excludes visual field loss.

ONE EYE/TWO EYES OR NEITHER: A simplified test of visual fields that can be easily done at the bedside is to have the patient sequentially cover each eye and describe how their symptom changes. A symptom that is exclusively in one eye is characterized by resolution when the affected eye is covered and persistence when the fellow eye is covered. This localizes anterior to the chiasm in the affected eye or optic nerve. A symptom that is in both eyes is characterized by persistence when either eye is covered. This localizes either to both eyes anterior to the chiasm, to the chiasm or behind the chiasm. A symptom that resolves when either eye is covered suggests that there is no lesion in the afferent visual pathway and that the symptom is related to ocular misalignment (see "Approach to Diplopia").

AFFERENT PUPILLARY FUNCTION: Anisocoria is NOT a test of afferent visual pathway function. Afferent visual function is characterized by the resting size of BOTH pupils given a certain light stimulus. One useful, practical finding is absence of pupillary constriction in either eye when light is shone in a blind eye – this localizes to a problem in that eye. In cases of less severe vision loss, the relative difference in pupillary response to light between the two eyes can be visualized using the swinging flashlight test. The flashlight is alternately shone in each eye, looking for constriction of the pupils when one eye is illuminated and dilation of the pupils when the other eye is illuminated. A subjective version of this test is to ask the patient if a standard light stimulus is perceived as equal in both eyes.

FUNDUSCOPIC EXAM: See "Examining the Ocular Fundus and Interpreting What you See"

NEUROLOGICAL EXAM AND LOCALIZE: Particular attention should be paid to symptoms and signs localizing to near the afferent visual pathway or suggesting neurological syndromes that can involve vision loss.

COMMON PATTERNS & EXAMPLES OF ETIOLOGIES:
OPHTHALMIC: Most causes of monocular vision loss are due to eye disease and are readily apparent on a full ophthalmic evaluation (i.e. dilated exam by an ophthalmologist). The exceptions are retrobulbar optic neuropathy and outer retinal disease, both of which can have normal slit lamp and dilated fundus examinations.

Retinal arteriolar occlusion is an ophthalmic diagnosis relevant to neuro-ophthalmology because the pathophysiology overlaps with that of ischemic stroke. It can be asymptomatic or cause partial monocular vision loss in the case of branch RAO or complete monocular vision loss as is the case in central RAO. Many advocate for treatment of this condition as is standard of care for acute brain stroke.[2]

MONOCULAR OPTIC NEUROPATHY: The most important cause of monocular vision loss relevant to neurologists is optic neuropathy. A monocular optic neuropathy can be asymptomatic or affect central and/or peripheral vision in one eye. Classically, there is associated change in color perception and relative afferent pupillary defect in unilateral cases. The differential diagnosis is broad and includes anterior (optic nerve head), intraorbital, intracanalicular and chiasmal etiologies. Swelling of the optic nerve head identifies acute anterior optic neuropathies. Acute retrobulbar optic neuropathies may have a normal funduscopic appearance. Chronic optic neuropathies are associated with optic nerve head pallor. It takes weeks to month following retrobulbar optic nerve injury for pallor of the optic nerve head (reflecting axonal loss) to develop.

Associated signs and symptoms that can help to localize an optic neuropathy include
- Orbit: proptosis, orbital pain, extra-ocular movement limitations
- Orbital apex: ipsilateral CN III, IV, VI, V1 dysfunction
- Skull base/pituitary: CN III, IV, VI, V1, V2, sympathetic pathway dysfunction

Important considerations for monocular optic neuropathies that require rapid diagnosis or intervention include
- Optic neuritis (particularly NMO spectrum, which may necessitate escalation of immunotherapy to induce remission)[3]
- Other inflammatory optic neuropathy (e.g. sarcoidosis, which may require IV immunotherapy for treatment)
- Arteritic ischemic optic neuropathy, which carries short term risk of fellow eye involvement and stroke[4]
- Acute compressive (e.g. pituitary apoplexy, which is associated with acute adrenal insufficiency)[5]
- Fungal orbital apex syndrome[6]

BILATERAL OPTIC NEUROPATHY: The differential diagnosis is similar as for monocular optic neuropathies. The symptoms and signs differ due to bilateral nature and likely absence of relative color vision changes and relative afferent pupillary defects since these findings rely on comparison between the eyes. Conditions that are more likely to cause acute bilateral optic neuropathies are:
- Papilledema due to elevated intracranial pressure, which can cause blindness and may be presenting sign of malignant brain tumor, venous sinus thrombosis or meningeal disease[7]
- Pituitary apoplexy causing acute chiasmal compression (see above)

OPTIC TRACT: Lesions of the optic tract cause homonymous visual field loss in both eyes (i.e. same field of both eyes) and a contralateral afferent pupillary defect. This is due to the fact that the tract contains more fibers from the contralateral than ipsilateral eye. Because the optic tract is composed of presynaptic retinal ganglion cells, chronic lesions are associated with bilateral optic nerve head pallor. Isolated visual symptoms are rare, but can occur due to compression from a sellar or suprasellar process (e.g. pituitary tumor or craniopharyngeoma) or retinal ganglion cell pathology (e.g. optic neuritis).

LATERAL GENICULATE NUCLEUS: Lesions to the lateral geniculate nucleus are associated with homonymous contralateral visual field loss in both eyes. Isolated visual symptoms are rare but can occur from either anterior circulation (anterior choroidal artery) or posterior circulation (posterior choroidal artery) strokes.

OPTIC RADIATIONS: Dysfunction of the optic radiations is associated with visual field loss in the opposite field (both vertical and horizontal) in both eyes. For example vision loss in the upper right visual field can be caused by dysfunction of the left temporal radiations (Meyer's loop). Due to the distributed nature of the radiations, it is

rare for injury to cause isolated visual symptoms and it is typical to have other symptoms/signs that localize to the relevant parietal/temporal lobe. Differential diagnosis is that for parenchymal lesions of the brain including stroke, tumor and demyelination amongst others.

OCCIPITAL LOBE: Similar to optic radiation dysfunction, dysfunction in an occipital lobe is associated with visual field los in the opposite field (both vertical and horizontal) in both eyes. Unlike visual field defects localizing to the optic radiations, those from occipital lobe dysfunction are commonly isolated. The differential diagnosis is that for parenchymal lesions of the brain including stroke, tumor and demyelination amongst others.

Midline (e.g. falcine meningioma) or distributed processes (e.g. reversible posterior leukoencephalopathy syndrome can cause bilateral occipital lobe dysfunction and even blindness on a cerebral basis. In these cases pupil reactivity is normal and patients may be agnostic to their vision loss and confabulate (Anton Syndrome). Lesions to the occipital pole cause central vision loss.

TRANSIENT VISION LOSS: Transient vision loss can be caused by a variety of conditions.[8] Neurologic and ophthalmic exams may offer clues regarding etiology. Though much time can be spent trying to distinguish eye from brain events based on history, it is often difficult to determine conclusively. A more important distinction is whether the symptoms may have been due to ischemia. Diffusion weighted imaging brain MRI can be useful in the short term as it may identify persistent or concurrent ischemia in the brain even after symptom resolution.[9] TIA definitions include symptoms due to either retinal or brain ischemia and guidelines should be followed if either of these are suspected.[10]

TESTING TO CONSIDER:

MRI ORBIT provides detailed imaging of the orbital and canalicular portions of the optic nerve and includes fat saturated images, which are helpful for visualizing enhancement separate from the background T1 hyperintensity caused by orbital fat.

MRI PITUITARY/SELLA provides detailed imaging of the optic chiasm.

MRI BRAIN is the test of choice to exclude parenchymal lesions such as stroke, demyelination or tumor directly or indirectly (i.e. via elevated ICP and papilledema) causing vision loss.

MRA neck or CTA neck are useful for evaluating for large vessel contributions to ischemia. MRA head or CTA head are useful for evaluating for intracranial vascular stenosis.

MRV head is helpful to evaluate for venous sinus thrombosis as a cause of elevated intracranial pressure causing optic neuropathies from papilledema.

Lumbar puncture is helpful for measurement of intracranial pressure in the setting of bilateral optic neuropathies and for evaluation of meningeal processes.

Echocardiography is helpful to identify cardiac sources of embolism.

Cardiac rhythm assessment is helpful to identify atrial fibrillation or other rhythm disturbance as a possible etiology of ischemic vision loss.

Serological testing is important for evaluation of possible giant cell arteritis (ESR, CRP) as well as evaluation of other lesions discovered on exam or imaging.

Temporal artery biopsy is important for evaluating for possible giant cell arteritis.

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