Perspectives on Neuro-Ophthalmology

Interpreting Patient History

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Going blind: clarification — both eyes or one; totally black, blurred, or double; distant or far or both; constant or intermittent; most common findings — dry eyes; refractive error; convergence insufficiency; monocular or binocular diplopia; fear

Had a stroke: clarification — other areas affected by stroke (eg, weakness, numbness); most common findings — unilateral visual loss (eg, ischemic optic neuropathy [ION]); bilateral visual loss (eg, homonymous visual field defect); no visual loss (eg, refractive error, dry eyes); complicated migraine event

My eye is killing me: clarification — point to or outline pain; transient or permanent; often constant pressure with intermittent sharp pain; most common findings — corneal erosion or dry eye syndrome; bilateral pain rules out hemi-trigeminal neuralgia

Cannot read eye chart: clarification — can patient see wall, screen, or light; ask patient to blink and insist they read chart; most common findings — malingering; dry eyes

Cannot see out of right eye: clarification — cover right eye; whether getting worse; most common findings — true visual loss; right hemianopic defect; ptosis

Cannot read anything: clarification — can patient see wall, screen, or light; can they see print — cover right eye; does eye hurt when moved (often indicates optic neuritis); duration of worsening

Ptosis: determine whether asymmetric or related anisocoria present; decide whether treatment appropriate or necessary given side effects; speaker discourages patients with myasthenia gravis (MG) from undergoing treatment if ptosis only problem; causes — congenital; age-related; Horner syndrome; MG; retraction of other eyelid; early hemifacial spasm (small intermittent contraction of orbicularis triggered by squeezing eyes shut and opening 3 or 4 times); treat with botulinum toxin (BTX); brow ptosis; previous Bell palsy; long-term contact lens wear (look for giant papillary conjunctivitis [GPC]); dry eye syndrome (causes adherence)

Examinations and diagnoses: Tensilon, ice, or rest tests helpful; benign essential blepharospasm with symmetric mild contraction may relax if patient distracted for 5 to 6 min; examine older photograph; if ptosis present, consider chronic progressive external ophthalmoplegia (CPEO; ruled out by response to Tensilon); cases of overlapping MG and blepharospasm respond to BTX; blepharochalasis (overhanging fold of skin can hide eyelid retraction caused by thyroid eye disease); in extreme blepharospasm, lid apraxia can cause difficulty opening, even if spasm relaxed; painful Horner syndrome in young to middle-aged men presents with neck pain, headache, anisocoria, and ptosis possibly hidden by eyelid folds

Proposis: difficult to obtain consistent measurements when evaluating progression; possibly caused by contralateral enophthalmos from old orbital fracture; confirm with computed tomography (CT); some inequality of size normal; evaluate sizes of hands and feet or old photograph to identify congenital differences; unilateral proposis and ptosis indicates problem in orbit; unilateral proposis and retraction indicates thyroid eye disease (check family history)

Diplopia: determine whether monocular, binocular, or both and whether comitant or incomitant; comitant (equal strabismus in all fields of gaze) suggests decompensated preexisting eye condition; common causes (monocular) — refractive error, dry eyes, or cataract; common causes (bilateral) — decompensated preexisting phoria; congenital superior oblique palsy; acquired cranial neuropathy; acquired myopathy (restrictive or paretic); objective examinations — cover-uncover or corneal light reflex; subjective examinations — speaker uses red light test with fixation light as screening tool; more sensitive than objective testing; Risley prism test detects 0.5 to 1.5 diopters (D) of vertical deviation; decompensated phorias — decereption after cataract surgery (patient may have microstrabismus and difficulty with fusion); head trauma; possibly related to age or stress

Congenital superior oblique palsy: presentation includes longstanding head tilt; fusion of >1.5 D vertically indicates congenital fourth nerve palsy or MG; examination — 3-step test; look for overaction of inferior oblique; treatment — perform myectomy because recession only delays scarring problems

Acquired cranial neuropathy: presentation — complete or partial third, fourth, or sixth nerve palsy; intracranial ophthalmoplegia (INO); speaker considers ischemic mononeuropathy similar to ION; look for coexisting vasculopathies (eg, lipid disease); skew deviation — normally right fourth palsy shows right hypertropia that worsens on left gaze and vice versa;

Educational Objectives

The goal of this program is to improve the diagnosis and treatment of neuro-ophthalmologic disorders. After hearing and assimilating this program, the clinician will be better able to:

1. Interpret patients’ histories and descriptions of their symptoms and complaints.
2. Diagnose and treat ptosis, proposis, diplopia, and various neuropathies and myopathies.
3. Determine whether patients with symptoms of optic neuropathy or third nerve palsy should be referred to a neuro-ophthalmologist.

5. Identify and refer patients with Tolosa-Hunt syndrome and amaurosis fugax.

Faculty Disclosure

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skew deviation shows left hypertropia that worsens on left gaze (similar to inferior rectus paresis) and vice versa; mimics double vision; usually accompanied by balance disorders or ataxia; dolicioctasia of intracranial vessels not uncommon; may cause progressive compressive optic neuropathy

Acquired Myopathies

Paretic myopathies: MG — onset either abrupt or insidious, systemic or ocular; use ice test or others; CPEO — usually long-standing, chronic, and progressive for decades; family history variable; important to rule out MG; MG — most patients with ocular MG progress to systemic MG; ptosis and diplopia common; “pseudo-INO” (pseudo-weakness of medial rectus mimicking brainstem disorder) but really MG; CPEO Plus syndrome can affect balance and optic atrophy; convergence and divergence insufficiency — diplopia in patients >60 yr of age often caused by divergence insufficiency or convergence insufficiency; for divergence insufficiency, avoid surgically correcting medial rectus muscles (may produce convergence insufficiency); exercises do not work for convergence insufficiency; prescribe prism reading glasses

Case example: man 20 yr of age had pain in right eye, left orbital swelling 3 mo previously (treated), right orbital swelling 5 wk previously; improved after treatment with oral steroids; examination showed mild persisting discomfort of right medial canthus; imaging showed thickening and inflammation of right medial rectus; migration from orbit to orbit suggests inflammatory myositis rather than thyroid eye disease

Forced ductions to differentiate between paretic and restrictive myopathy: administer drop of topical anesthetic in eye; use cotton-tipped swab to force eye to look up

Restricted myopathies: caused by accidental or iatrogenic orbital fractures, thyroid eye disease (determine whether stable or progressive), orbital inflammatory syndrome (determine whether fibrotic or lymphocytic), or infiltrative (consider Mucor if diabetic)

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Five Key Diagnoses

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Key diagnoses: consider referral — posterior ischemic optic neuropathy (PION); third nerve palsy (unless aneurysm ruled out); chronic optic neuritis; Tolosa-Hunt syndrome (THS); retinal migraine (on first visit); characteristics — all diagnoses made on basis of exclusion; alternative etiologies for THS and retinal migraine potentially life threatening; often show negative results on imaging, producing false sense of security; make diagnoses retrospectively (after everything ruled out and patient better or stable); always refer if THS progresses or retinal migraine atypical (eg, corneal ulcer, retinal detachment)

PION and giant cell arteritis (GCA): ischemic lesion that does not cause swelling of optic nerve; presents as acute unilateral loss of vision, relative afferent pupillary defect (RAPD), hand motion visual acuity, and normal fundus; do not diagnose retrobulbar optic neuropathy in elderly; in young woman, diagnose acute unilateral retrobulbar optic neuropathy as optic neuritis; in elderly patients, diagnose PION until proven otherwise; PION considered GCA until proven otherwise; can occur after surgery; nonarteritic form — anterior ischemic optic neuropathy (AION) shows swollen disc during acute phase of nonarteritic form of AION

Optic neuropathy: broad category of localizing disease; optic neuritis indicates idiopathic, inflammatory, or demyelinating disease; MRI — necessary to rule out compressive lesion, but does not show abnormality in PION; ischemic optic neuritis does not exist (only optic neuritis ischemic); incorrect treatment with steroids could harm patient; consider fundus fluorescein angiography (FFA) in PION of elderly to differentiate retrobulbar optic neuritis from compressive lesion or ischemia (choroidal perfusion deficit) in cases where no swelling visible

Criteria for diagnosis of PION: clear cause for retrobulbar ischemia (eg, GCA or after spine or cardiac procedure involving systemic hyperperfusion); MRI ruled out tumor; if findings consistent with GCA, consider FFA; RAPD establishes optic neuropathy; optic atrophy develops over time and remains stable thereafter; do not treat with steroids if fungus present; Optic Neuritis Treatment Trial (ONTT) showed age range for optic neuritis of ≤46 yr; diagnosis of optic neuritis rare in elderly

Third nerve palsy: elderly patient had ptosis and third nerve palsy; GCA remained possible; perform imaging study to look for compression; diplopia in elderly patients may also indicate GCA; if not GCA and pupil involved, consider aneurysm; pupil-involved third nerve palsy considered aneurysm of posterior communicating artery (PCA) until proven otherwise; perform angiography (ie, MRA or CTA); rate of mortality after rupture of aneurysm 50% at scene; 50% of survivors permanently impaired

Neuromyelitis optica (NMO): ONTT showed 97% of patients with optic neuritis recovered VA better than 20/200 in first 6 mo regardless of treatment; refer patients who do not recover; chronic optic neuritis seen in patients with severe secondary and progressive form of optic neuritis from secondary and progressive multiple sclerosis (MS); most patients with MS have relapsing remitting form showing relapse and remission of optic neuritis also

Leber hereditary optic neuropathy: consider in young male patients with central scotoma and retrobulbar optic neuropathy; central scotoma does not improve and other eye becomes affected; imaging studies appear normal

Diagnosis: no diagnostic test available for THS; CT insufficient; MRI required; refer patients; review of literature (La Mantia et al, 2006) showed only 21% of 208 reported cases met diagnostic
criteria for THS; 50% of patients with negative CT results had lesion by MRI; 33% had normal neuroimaging results

Ocular migraine: case example — 60-year-old woman experienced loss of vision (as if curtain came down) that lasted 10 min and resolved; first event; examination of eye normal; condition incorrectly diagnosed as ocular migraine; altitudinal presentation indicates ischemic event; diagnostic criteria — review of literature (Hill et al, 2007) found only 5 of 16 reported cases met diagnostic criteria of International Headache Society (IHS) for retinal migraine (now called retinal vasospasm); diagnosis made by exclusion; IHS criteria (ie, ≥ 2 attacks of fully reversible monocular vision loss with migraine headache); migraine mechanism (ie, spreading depression of electrical cortical neuronal activity); no residual vision loss on visual field test; key findings of amaurosis fugax — if onset altitudinal, odds ratio (OR) 3.7 for ipsilateral internal carotid artery (ICA) stenosis of 70% to 90%; if mode of onset unknown, OR for ICA stenosis only 0.5; diagnosis of retinal migraine requires multiple attacks; first presentation of altitudinal vision loss in elderly patient indicates ICA stenosis; in young patient, dissection

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The California State Board of Registered Nursing (CA BRN) accepts courses provided for AMA Category 1 credit as meeting the continuing education requirements for license renewal.
1. Which of the following disorders is least likely to be found in a patient who complains of bilateral eye pain?
   (A) Corneal erosion  (B) Trigeminal neuralgia  (C) Dry eye syndrome

2. Ptosis is often caused by any of the following, except:
   (A) Horner syndrome  (B) Myasthenia gravis  (C) Previous orbital fracture  (D) Chronic progressive external ophthalmoplegia

3. The most likely diagnosis for a patient who has unilateral proptosis and eyelid retraction is which of the following?
   (A) Orbit fracture  (B) Thyroid eye disease  (C) Horner syndrome  (D) Hemifacial spasm

4. Binocular diplopia is often caused by any of the following disorders, except:
   (A) Decompensated preexisting phoria  (B) Congenital superior oblique palsy  (C) Acquired cranial neuropathy  (D) Refractive error

5. Prism reading glasses are more effective for treatment of convergence insufficiency than exercises.
   (A) True  (B) False

6. Referral to a neuro-ophthalmologist should be considered for patients whose conditions are consistent with which of the following diagnoses?
   (A) Posterior ischemic optic neuropathy  (B) Tolosa-Hunt syndrome  (C) Chronic optic neuritis  (D) A, B, and C

7. The most likely diagnosis for an elderly patient who presents with acute unilateral loss of vision, relative afferent pupil defect, hand motion visual acuity, and normal fundus is which of the following?
   (A) Optic neuritis  (B) Tolosa-Hunt syndrome  (C) Giant cell arteritis  (D) Retinal migraine

8. A patient with pupil-involved third nerve palsy is likely to have an aneurysm of the posterior communicating artery.
   (A) True  (B) False

9. A patient whose optic neuritis does not remit and who has transverse myelitis should be treated with which of the following agents?
   (A) Interferon β-1a  (B) Interferon β-1b  (C) Glatiramer acetate  (D) Immunosuppressive therapy

10. Choose the correct statement about amaurosis fugax.
    (A) If the onset is altitudinal, the most likely diagnosis is retinal vasospasm
    (B) If there are ≥2 attacks of fully reversible monocular vision loss with migraine headache, the most likely diagnosis is retinal vasospasm
    (C) A literature review found that 83% of patients with amaurosis fugax met the diagnostic criteria for retinal vasospasm
    (D) If there is residual vision loss on a visual field test, the most likely diagnosis is retinal vasospasm