20/20 SIM Neuro-ophthalmology Supplemental Teaching Guide

*One-hour virtual interactive workshop guide for facilitators of the 20/20 SIM neuro-ophthalmology interactive session. Note that this guide is a supplement to the extensive educational content provided on the 20/20 SIM platform, which provides the prompts and clinical details, as well as hyperlinks to original sources of information.*

**Logistics:**

* Requires approximately one hour of curricular time for third- or fourth-year medical students.
* Designed to be conducted with an online video conferencing platform with screen share capabilities
* Recommend a facilitator familiar with both the 20/20 SIM platform and the contents of this teaching guide.

**Session Learner Objectives:**

* Identify and describe common neuro-ophthalmologic manifestations of neurologic conditions
* Explain the differential diagnosis, diagnostic work up, and evidence-based treatment of common neuro-ophthalmologic conditions
* Utilize interactive, case-based discussion to foster an enjoyable, student-focused e-learning environment via the 20/20 SIM platform

**Instructions:**

1. Welcome students on the videoconferencing platform and explain the importance of exposure to and basic understanding of high yield neuro-ophthalmology cases for all medical trainees, including key points:
   1. Current underexposure in medical education to neurology and ophthalmology across US institutions.
   2. Common and potentially life-threatening neurologic conditions often present with ophthalmologic manifestations.
   3. Given the projected shortages of neurologists and neuro-ophthalmologists, all trainees should be familiar with common neuro-ophthalmologic diseases
2. Share screen and open the 2020SIM.com website, ensuring all participants can see the homepage.
   1. Explain the concept of 2020SIM.com: a novel, online, case-based learning platform that can be accessed by anyone at any time. It includes high yield neuro-ophthalmology cases for students to complete with questions and answers, and hyperlinks to additional information from cited references. The website is also mobile-optimized and can be easily accessed from a cell phone.
   2. Explain that the purpose of the interactive session is to walk through 2-3 cases together as a group
   3. Introduce the top right banner of the website, the “Pick Your Case!” function, as well as reference pages including “Ophtho Abbreviations” and “Eye Anatomy.”
3. Go to “Pick Your Case!” and have students choose a case number at random from the neuro-ophthalmology series.
4. Identify a student to read the HPI.
5. Assess which answer choices on the differential students think are most likely given the HPI (see Case by Case Guide below for further information).
   1. Try to connect key points from the HPI to the differential with leading questions such as: “What are the key takeaway points in the HPI that help us narrow our differential?”
   2. Use open-ended questions to foster discussion of the differential.
   3. If students are not sure which diagnoses are most likely, have a student pick any answer choice and explain to the group what makes it a more or less likely diagnosis, or ask questions such as the following:

“Why might we have lower suspicion for amaurosis fugax?” or “What points in the history lead us away from suspecting meningioma?”

“What leads us to central retinal artery occlusion as opposed to central retinal vein occlusion? What is the difference?”

* 1. Try to highlight key differences in each diagnosis on the differential by asking open-ended questions such as:

“What would we expect in the history for temporal arteritis?”

“Even though we have chosen 3 possible diagnoses, is there anything else on this list that we could include?” Why or why not?”

1. Continue to read through the components of the case, including additional history, exam findings, workup, and management, following the prompts of the case and discussing the answer choices in a similar open-ended fashion. Consider guiding questions such as:

“What are the most pertinent aspects of the physical exam in this case?”

“How do these physical exam findings help narrow our differential?”

“How would ordering an MRI brain help us in this case?”

1. Identify a student to read aloud the discussion page. Highlight the hyperlinks to original sources of information and remind students to return to the website for further learning at their convenience.
2. Answer any outstanding questions from students.
3. Move on to the next case by returning to the “Pick Your Case!” page and having a student pick another number at random.
4. Aim to discuss 2-3 cases during one-hour session in a similar fashion.

**Case by Case Guide:**

Case 1:

DIAGNOSIS: Central Retinal Artery Occlusion (CRAO)

HISTORY OF PRESENT ILLNESS:

* An elderly person with vascular risk factors of high blood pressure, diabetes, high cholesterol, and recent stroke presents with sudden onset of monocular vision loss.
* In addition to the relevant cerebrovascular risk factor profile, highlighting the sudden onset and painless nature will help narrow the differential toward central retinal artery occlusion.

ADDITIONAL HISTORY:

* Note that the patient had a prior episode of transient unilateral vision loss lasting a few minutes (amaurosis fugax).
* Carotid doppler demonstrates ipsilateral internal carotid artery plaque, which like served as the source for an embolus to the central retinal artery supplying the retina

PHYSICAL EXAM:

* Vitals: notable for mild hypertension
* Visual acuity reduced to hand motion in the left eye (OS) and relatively normal vision in the right eye (OD).
* The relative afferent pupillary defect (rAPD) in the left eye signifies impaired light perception in the left eye due to retinal or optic nerve pathology.
* Fundoscopic exam reveals a classic finding in CRAO of a pale retina and a cherry red spot.

BONUS QUESTION:

Other conditions associated with a cherry red spot on fundoscopic exam include Tay-Sachs and Neimann-Pick Disease.

TESTS TO ORDER:

STAT CT head and CT angiography of the head and neck should be immediately performed to assess for cerebral infarction and compromise to the cerebral blood supply that warrants revascularization. Additional testing will be performed to assess the etiology of the CRAO and cerebrovascular risk factor profile.

DIFFERENTIAL DIAGNOSIS:

* Ischemic optic neuropathy: another cause of sudden painless vision loss. This is a general term and there are different causes of ischemic optic neuropathy. A key distinction is whether it is *arteritic* (due to inflammation/vasculitis of an artery supplying the optic nerve, for example temporal arteritis), or *nonarteritic* (due to vascular ischemia).
* Amaurosis fugax: vision loss would be transient.
* Cataract: less likely given the timing and sudden onset of vision loss.
* Acute angle closure glaucoma: also acute in onset and an ophthalmologic emergency but associated with prominent pain.
* Macular degeneration: more insidious in onset and results in loss of central vision.
* Retinal detachment: another ophthalmologic emergency that is acute and painless but is often associated with floaters, flashes, curtaining of vision).
* Diabetic retinopathy: associated with similar risk factor profile but more insidious in onset.
* Central retinal vein occlusion: extent of vision loss may not always be as profound; results in scattered hemorrhages throughout the macula.
* Vitreous hemorrhage: also causes painless loss of vision, may be associated with floaters.

Case 2:

DIAGNOSIS: Idiopathic Intracranial Hypertension (IIH)

HISTORY OF PRESENT ILLNESS:

* A young adult female presents with headaches. She states the headaches are worse in the morning. It is important to note that she has gained 20 pounds recently.
* She has been having brief episodes of blurred vision when bending over, also known as transient visual obscurations (TVO), which are a symptom of elevated intracranial pressure.
* She also has noticed a “swooshing” sound, which is one way patients may describe associated pulsatile tinnitus.

ADDITIONAL HISTORY:

* Note that the patient was recently prescribed isotretinoin, which is associated with IIH.
* She has a family history of migraines which can raise suspicion for migraine.

PHYSICAL EXAM:

* Vitals: notable for BMI of 31 kg/m2
* Her neurological exam is non-focal
* Fundoscopic exam reveals optic disc swelling

TESTS TO ORDER:

Because of the signs and symptoms of increased intracranial pressure, we need to first obtain imaging to rule out a mass lesion. The next step is to order MRI brain with MRV. It is not safe to proceed with lumbar puncture until a mass lesion has been ruled out.

DIFFERENTIAL DIAGNOSIS:

* Acute angle closure glaucoma: less likely as the vision changes are brief, transient, and painless.
* Meningitis: less likely given the chronicity of symptoms and absence of associated infectious signs.
* Migraine without aura: although common in young healthy adults, no migranous features such as nausea, vomiting, photophobia, phonophobia.
* Subdural hematoma: although when large can result in signs and symptoms of elevated intracranial pressure, often associated with trauma history.
* Sinusitis: would not expect associated TVO and pulsatile tinnitus.
* Cluster headache: typically present as brief bursts of excruciating pain associated with autonomic symptoms such as lacrimation, not TVO and pulsatile tinnitus.
* Tension headache: although also common in young healthy adults, would not expect signs and symptoms of elevated intracranial pressure.
* Intracranial mass: must be ruled out given the signs and symptoms of elevated intracranial pressure.
* Primary open angle glaucoma: unlikely to cause headache and does not cause tinnitus.
* Vitreous hemorrhage: usually presents as painless monocular vision loss.
* Hypertensive emergency: can cause headaches but would not expect signs of elevated intracranial pressure.
* Ocular migraine: would not expect signs of elevated intracranial pressure.

Case 3:

DIAGNOSIS: Optic Neuritis

HISTORY OF PRESENT ILLNESS:

* A young adult with a history of migraines and hypothyroidism presents with painful unilateral vision loss.
* Note that she endorses pain around the eye and with eye movement, which is important for narrowing the differential.
* She had a recent illness, which can trigger certain autoimmune diseases.

PHYSICAL EXAM:

* Decreased visual acuity and decreased perception of red on the left (ie., red objects seen through the left eye appear significantly more dull than when seen through the right eye. This “red desaturation” is associated with optic nerve pathology).
* rAPD in the left eye signifies impaired light perception in the left eye due to retinal or optic nerve injury.
* Fundoscopic exam of the left eye shows optic disc swelling.
* Focal neurologic findings of right leg and foot sensory loss and hyperreflexia suggest pathology in the brain along with the optic nerve.

TESTS TO ORDER: MRI of the brain and orbits are ordered initially to assess for structural and inflammatory lesions and are more sensitive than CT. Eventually, a lumbar puncture may help determine the etiology.

DIFFERENTIAL DIAGNOSIS:

* Scleritis: also associated with autoimmune disease but would not expect optic nerve swelling rAPD, or focal neurologic deficits on exam.
* Migraine with aura: would not expect painful vision loss, optic nerve swelling, rAPD, and focal neurologic deficits.
* Ischemic optic neuropathy: strong consideration given optic nerve findings but would not expect associated focal neurologic deficits. This is also more typically seen in older patients with vascular risk factors.
* Amaurosis fugax: typically painless transient vision loss.
* Central retinal vein occlusion: typically painless vision loss.
* Meningioma: does not typically cause symptoms from mass effect due to extra-axial location and slow growth, but when symptomatic more likely to cause headache or seizure. However, rarely, a meningioma within the orbital apex could present with visual symptoms although they would most likely be painless.
* Retinal detachment: typically painless and associated with floaters, flashes, and curtaining of vision.
* Thyroid orbitopathy: also autoimmune mediated and uncomfortable but patients usually experience a sensation of grittiness in the eye, photophobia and eyelid retraction and exophthalmos.
* Central retinal artery occlusion: typically painless vision loss.

Case 4:

DIAGNOSIS: Cranial nerve III palsy due to compressive posterior communicating (Pcomm) artery aneurysm

HISTORY OF PRESENT ILLNESS:

* An adult with hypertension and diabetes mellitus presents with sudden onset of double vision persisting over hours.
* When she covers each eye individually her double vision resolves, thus this is binocular double vision (i.e. it’s present when both eyes are open and resolves with closing one eye). Binocular vision complaints clue us in that the pathology is in the brain as opposed to the eye itself. Monocular diplopia is usually a result of uncorrected refractive error.
* The double vision worsens when looking towards the left. It is important to remember the eye muscles involved in left lateral gaze, including the lateral rectus on the left (innervated by CN VI) and medial rectus on the right (innervated by CN III).
* The “droopy” right eye lid suggests ptosis, which can be due to weakness of the levator palpebral superioris that is responsible for lifting the eyelid. Consider discussing the difference in appearance of ptosis from a CN III palsy vs. ptosis from Horner’s syndrome. The associated pupillary changes with each are opposite.
* She has a family history of an early death from a brain bleed, which could have been caused by a ruptured aneurysm.

PHYSICAL EXAM:

* External exam shows that the right eye is closed (ptosis due to levator palpebrae weakness).
* Visual acuity is worse in the right eye, and the right pupil is mid-dilated and not reactive to light due to damage to the sphincter pupillae and parasympathetic fibers in setting of compression of CNIII.
* The diagram showing extraocular movements displays that the right eye is limited in looking up and to the left due to impairment of the superior rectus and medial rectus.

BONUS QUESTION: this is an emergency! A Pcomm aneurysm causing a CNIII palsy is at imminent risk of rupture. Every effort must be made to preserve visual function and prevent further compromise.

IMAGING: The axial and coronal CT angiogram images demonstrate a right sided Pcomm aneurysm.

BONUS QUESTION: subarachnoid hemorrhage is the most common life-threatening complication of this patient’s presentation.

DIFFERENTIAL DIAGNOSIS:

* Horner’s syndrome: classic triad of miosis, ptosis, anhidrosis but does not typically cause diplopia.
* Myasthenia gravis: should be considered in any patient presenting with double vision, although presentation unlikely to be so acute and symptoms are often intermittent and worse with fatigability.
* Bell’s palsy: CN VII palsy causes facial weakness rather than double vision; an eyelid problem sometimes seen in Bell’s palsy is inability to close the eye, which this patient does not complain of.
* CN IV (trochlear nerve) palsy: weakness of the trochlear nerve, which innervates the superior oblique and allows for intorsion of the eye, may also cause double vision but would not be associated with ptosis.
* CN III (oculomotor nerve) palsy: the oculomotor nerve controls movement of both the eye and the eyelid. A right sided CN III palsy may cause difficulty with vertical gaze and adduction (leading to double vision), as well as ptosis.
* CN VI (abducens nerve) palsy: although lateral rectus weakness can cause horizontal diplopia that worsens when looking left, would not be associated with ptosis or pupillary abnormality.
* Temporal arteritis: most common in patients >60 years and causes headache, jaw claudication, and constitutional symptoms in addition to vision loss.
* Internuclear ophthalmoplegia (INO): a lesion in the medial longitudinal fasciculus causes impaired adduction of the affected eye, along with nystagmus of the contralateral abducting eye, and would cause diplopia but not ptosis. It would also not be associated with a or pupillary abnormality.

Case 5:

DIAGNOSIS: Homonymous superior quadrantanopia due to temporal lobe mass

HISTORY OF PRESENT ILLNESS:

* A 45 year old man presents with a few months of memory impairment and functional decline
* The patient endorses headaches and fatigue, which are nonspecific but important associated symptoms.
* He saw an ophthalmologist due to trouble reading, and was then referred to a neurologist. In discussion, consider reasons why an ophthalmologist may refer to a neurologist (i.e. what visual impairments suggest a lesion of the central nervous system).

PHYSICAL EXAM:

* Mental status exam notable for deficits in attention and memory.
* Visual field examination notable for right homonymous superior quadrantanopia (ie., the upper right portion of each eye, which is the right superior temporal field and the left superior nasal field) as seen in the visual field test.

TESTS TO ORDER: The cognitive impairment and focal deficits in an otherwise healthy adult warrant an MRI brain with contrast to evaluate for a structural/mass lesion. Serologic studies such as TSH, HIV, RPR, vitamin B12, CBC, and CMP are low cost ways to assess for potentially reversible causes of cognitive impairment.

DIFFERENTIAL DIAGNOSIS:

* MCA stroke: would likely be acute in onset.
* Vascular dementia: unlikely in a relatively young adult without vascular risk factors; typically results in step-wise decline.
* Acoustic neuroma: would be more likely to result in hearing loss and imbalance rather than affect cognition.
* Vertebrobasilar insufficiency: would expect transient brainstem symptoms such as vertigo, diplopia, dysarthria, dysphagia.
* HSV encephalitis: unlikely given the chronicity and absence of associated fever, headache, seizures; has a high mortality rate when untreated.
* Early onset Alzheimer’s dementia: a consideration in an adult less than 65 years with memory changes but would not expect associated focal neurologic deficits.
* Creutzfeldt-Jakob disease: although rare, a concern in anyone with rapid onset changes in memory and personality; would not expect focal neurologic deficits and a structural lesion on imaging.
* Vitamin B12 deficiency: an important potentially reversible cause of memory impairment.
* Glioblastoma: high on the differential for a structural lesion in this age group.
* Hypothyroidism: another important potentially reversible cause of memory impairment, fatigue, and other symptoms).
* PCA infarct: similar to MCA stroke, unlikely given the temporal course of the presentation.