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Neuro-Ophthalmology Subspecialty Highlight: With Dr. Sarah Thornton, Wills Eye Hospital

Joseph D. DeSimone, BS
Thomas Jefferson University

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By Joseph D. DeSimone, BS | Faculty Mentor: Sarah Thornton, MD

Introduction

Neuro-Ophthalmology is a subspecialty within ophthalmology that combines the complexities of nervous system disease with the intricacies of the ocular manifestations of those diseases. Neuro-ophthalmologists work closely with neurologists, neurosurgeons, rheumatologists, and other ophthalmologists in different subspecialties, such as retina and glaucoma, to get to the source of their patients' problems. Neuro-ophthalmology is typically a 1-year fellowship undertaken after residency training is completed, where physicians will learn the complex ways different diseases can manifest in the eye.

A typical day in the Neuro-Ophthalmology clinic varies, but usually involves managing patients with various diseases such as Myasthenia Gravis, Multiple Sclerosis, Optic Neuritis, Giant Cell Arteritis, Thyroid Eye Disease, and many others. A thorough history is essential for diagnosing these patients, as subtle details can give hints into the etiology of a patient's disease. Some common symptoms of neurologic disease include diplopia, sudden or transient vision loss, blurry vision, ptosis, headaches, or cranial neuropathies. A rigorous physical exam requires measuring visual acuity, visual fields, eye pressure, proptosis, extra-ocular

Dr. Sarah Thornton is an attending in the Neuro-Ophthalmology service at Wills Eye Hospital. She attended Sidney Kimmel Medical College, where she decided to pursue ophthalmology after rotating through the Wills Emergency Room as a third-year student: "I found the pathology so fascinating and the patients so grateful for an explanation and treatment of their eye condition. I also wanted to learn how to use this mysterious slit lamp device, which provided a direct view of the pathology in real time." After graduating from SKMC, Dr. Thornton pursued a residency in ophthalmology at Tufts Medical Center. She recalls being exposed to the "bread-and-butter" of ophthalmology, as well as interesting subspecialty clinics and time in the operating room. She enjoyed living in Boston, and discussed that Tufts has a dynamic balance of faculty supervision and autonomy that fostered her independent learning. A word of advice from Dr. Thornton: "When choosing a residency, I think it's helpful to think about your own values and prioritize the most important things for you, whether that be location, surgical volume, etc., and try to find a good fit." After graduating residency, she returned to Philadelphia for a Neuro-Ophthalmology fellowship under the guidance of Drs. Robert Sergott, Mark Moster, and Adam Debusk. She chose to pursue Neuro-Ophthalmology because she "always found those cases most interesting during residency and appreciated the analytical reasoning required to diagnose these challenging cases", and she "continues to be amazed by the diversity of pathology in the field." (Figure 1).

"When choosing a residency, I think it's helpful to think about your own values and prioritize the most important things for you."

Dr. Sarah Thornton
Figure 1

muscle ranges of motion, color vision, pupil responses to light and accommodation, as well as performing a slit lamp examination or using direct or indirect ophthalmoscopy. Diagnostic testing can involve a variety of imaging and lab tests depending on the patient. Imaging typically includes optical coherence tomography (OCT), visual field testing, computed tomographic (CT) or magnetic resonance imaging (MRI) scanning of the brain and orbits, and sometimes fluorescein angiography, electroretinography, and ocular ultrasound. Lab orders can also aid in diagnosing specific diseases, and some common tests used by neuro-ophthalmologists include inflammatory markers C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR), disease-specific antibodies in the serum, or genetic testing for rare hereditary diseases affecting vision, such as Leber's Hereditary Optic Neuropathy.

Clinical Pearls

Dr. Thornton shared some clinical pearls that medical students can use in clinical rotations in neurology and ophthalmology and beyond in clinical practice.

1. Pupillary Light Reflex: The pupillary response to light is a complex process but can give you great detail when evaluating a challenging neurological patient. Physicians test to see if the pupils are responding to light appropriately, and if the pupils appear symmetric or asymmetric. Normally, when light is shone into one eye, the retina will send a signal via the optic nerve (cranial nerve II) to both pretectal nuclei in the midbrain, which activate bilateral Edinger-Westphal nuclei, sending signals via both oculomotor nerves (cranial nerve III) to the sphincter pupillae muscles, resulting in bilateral pupillary constriction. If there is an asymmetric issue along the pathway, you will see a relative afferent pupillary defect, or RAPD, which occurs when the eyes dilate in response to light. RAPDs can be seen in a variety of diseases and lesions along the pathway to the brain, and some common etiologies include optic neuritis, ischemic optic neuropathy or compressive optic neuropathy, although

RAPD can sometimes be seen with severe asymmetric retinal disease.

The Swinging Flashlight test is used to look for a RAPD. To perform this test, the examiner will shine light into one eye to attain maximum constriction, then quickly switch the light source to the other eye and back again. Normally, the second eye should respond with the same pupillary constriction as the first eye as a result of the consensual response. If the eye does not respond at all to the light source and remains dilated, then there should be concern for optic nerve damage. Light shone in the unaffected eye will cause both eyes to constrict. Conversely, light shone in the affected eye will cause both eyes to dilate (Figure 2).1 This is also termed a Marcus Gunn Pupil.

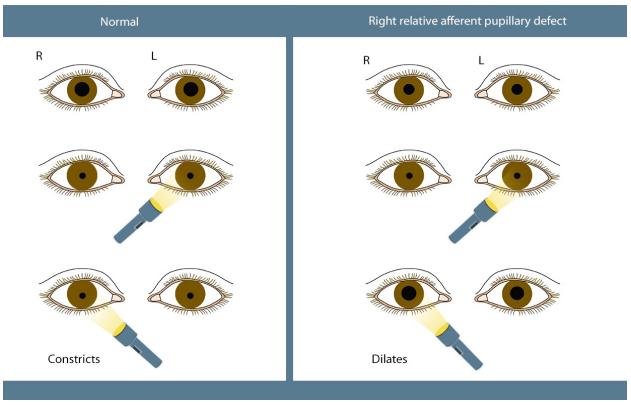

Figure 2

2. Bell's Palsy Examination: Bell's palsy is idiopathic inflammation and dysfunction of the peripheral facial nerve (cranial nerve VII) that results in facial weakness or paralysis that typically affects one side of the face.[2] Patients with Bell's Palsy will have unilateral facial drooping, drooling, difficulty closing the eye on the affected side, and difficulty smiling. Bell's palsy can limit the ability to blink, which can lead to painful corneal irritation and dryness leading to damage and vision problems. Evaluation of Bell's Palsy involves asking the patient to perform the following movements to test cranial nerve VII function and to differentiate an upper and lower facial nerve lesion:

- "raise your eyebrows" → affected side will not show forehead wrinkling and brow furrowing
- "smile" → affected side will have a flat facial contour
- "puff out your cheeks" → affected side will not be able to expand relative to the unaffected side
- "forcefully close your eyes" → affected side will not be able to fully close their eyes due to weakness of the frontalis muscle
- "stick out your tongue" → to check for lesions to the hypoglossal nerve (cranial nerve XII), will deviate toward affected side of nerve injury
- stroke forehead, upper face, and lower face bilaterally → to check for lesions to the ophthalmic (V1), maxillary (V2), and mandibular (V3) divisions of trigeminal nerve (cranial nerve V) sensations, respectively

Clinical Challenge

A 28-year-old woman with a past medical history of obesity, type 2 diabetes mellitus, obstructive sleep apnea, and migraines since childhood presented to the Wills Eye Hospital Emergency Department with one month of positional headaches, transient visual obscurations, and blurry vision in her right eye (OD). The best corrected visual acuity was 20/200 OD and 20/60 in her left eye (OS) with a 2+ APD OD. She had full color plates but was

slower OD. Fundoscopic exam revealed bilateral disc edema. Magnetic resonance imaging of the brain and orbits revealed slight protrusion of both optic nerves and a partially empty sella. MRV showed short segment stenoses at the bilateral transverse sigmoid sinuses. She was unable to undergo bedside lumbar puncture (LP) and was admitted for IV acetazolamide (Diamox) and a fluoroscopy-guided LP. The LP had an opening pressure of 46 cm H2O. Results of cerebral spinal fluid analysis and serology tests were negative

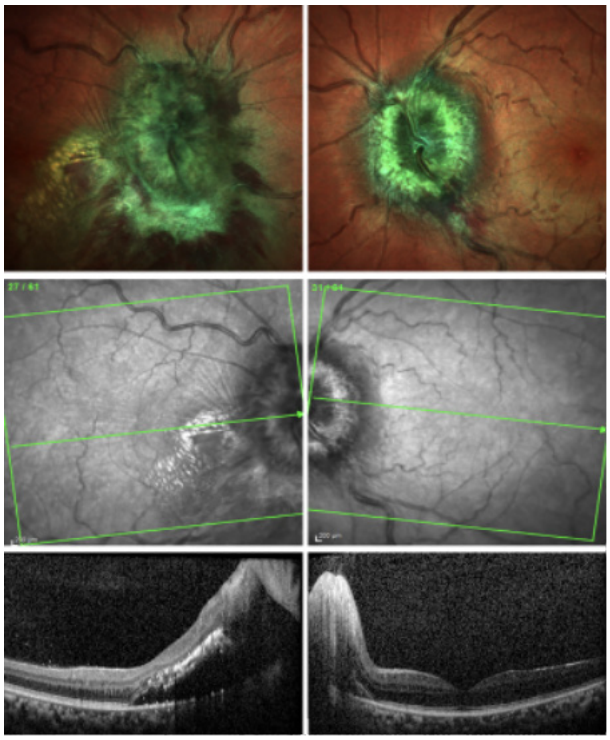

Figure 3

for infectious and inflammatory etiologies, including syphilis, sarcoidosis, Lyme disease, and tuberculosis. Serum studies revealed sickle cell trait and a hemoglobin A1c of 6.8%. She was discharged and was to continue Diamox 1 gram three times per day.

One week later in the Neuro-ophthalmology clinic, she underwent OCT (shown below, Figure 3). OCT showed severe bilateral disc edema, OD worse than OS. There was retinal nerve fiber layer (RNFL) edema extending into the fovea with exudates, chorioretinal folds, and peripapillary hemorrhages OD. Left eye showed RNFL edema with early tracking not extending beyond the prepapillary region, Paton's lines, and peripapillary hemorrhages. Inferior nerve thickness was 445 µm OD and 426 µm OS.

What is the next best step in the management of this patient?

A. continue Diamox and suggest weight loss
B. prescribe Dorzolamide-Timolol eye drops
C. consult Neurosurgery for placement of a ventriculoperitoneal shunt
D. consult Glaucoma for tube shunt placement

Answer: C

Discussion

The differential diagnosis for papilledema includes idiopathic intracranial hypertension, intracranial mass, obstructive hydrocephalus, meningitis, or cerebral venous sinus thrombosis.[3] This patient was diagnosed with pseudotumor cerebri (PTC), also known as idiopathic intracranial hypertension, a syndrome consisting of increased intracranial pressure, headaches, and visual loss not attributable to a mass lesion or underlying infection or malignancy.[4] PTC is normally a more insidious condition, allowing time to

treat medically and with weight loss. More rarely, PTC has a fulminant presentation, and consideration for surgical intervention is warranted. This patient's intracranial pressure was not responding appropriately to medical therapy with acetazolamide alone, so surgical intervention was recommended (eliminating choice A). This patient has increased intracranial pressure, so methods to lower intraocular pressure would be ineffective in treating PTC (choices B and D). The patient was referred to Neurosurgery for ventriculoperitoneal (VP) shunt placement (choice C). When considering surgery for PTC, options include optic nerve sheath fenestration, VP shunting, or venous sinus stenting. The overall rate of visual improvement seems to be equivalent across surgical interventions, and there is insufficient evidence to recommend or reject any one surgical intervention over another at this time.[5,6]

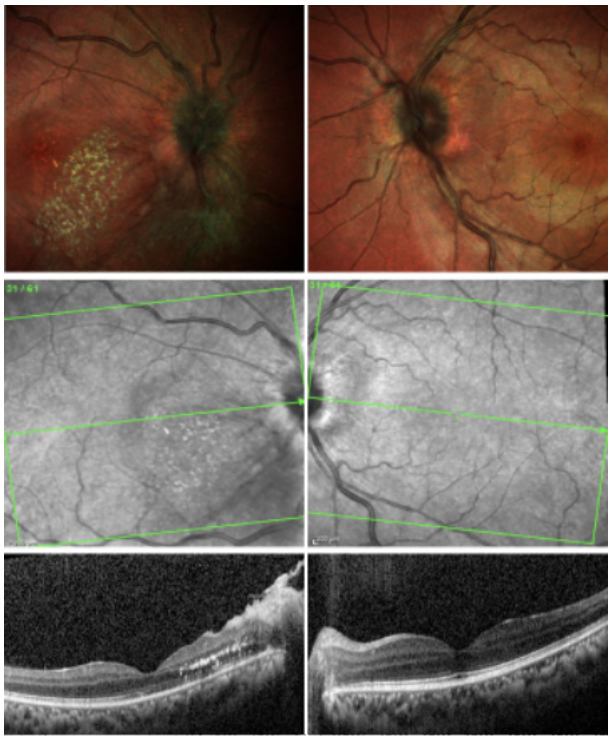

Figure 4

Two months after shunt placement, the patient was evaluated again by Neuro-Ophthalmology. Visual acuity was 20/40 OD and 20/20 OS and color plates were full in both eyes. OCT showed significant improvement in papilledema, now measurable with inferior thickness 124 µm OD and 109 µm OS (Figure 4).

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