THE NECESSITY OF THE NEURO RETINAL RIM

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COURSE DESCRIPTION: This course discusses the importance of the neuro retinal rim in visual function. Emphasis is placed on the critical analysis of the neuro retinal rim structure for such abnormalities as rim loss, pallor, and edema. A case-based approach highlights sometimes subtle neuro retinal rim findings, which when accurately assessed, can lead to proper diagnosis and treatment, and possibly save vision or life.

COURSE OBJECTIVES:

1. To understand the importance of the neuro retinal rim in visual function.
2. To appreciate the critical analysis of the neuro retinal rim when assessing the optic disc.
3. To realize the importance of matching the optic disc appearance with visual function.
4. To gain a better understanding of the etiologies of neuro retinal rim loss, and when this rim loss may be associated with a non-glaucomatous etiology.
5. To identify neuro retinal rim pallor and understand the necessary work-up to rule out the many possible causes.
6. To appreciate edema of optic disc and realize its varied potential mechanisms.

COURSE OUTLINE:

I. NEURO RETINAL RIM / OPTIC DISC ASSESSMENT

   a. Structure
      i. Fundus Examination
         1. Optic disc
            a. Neuro retinal rim
               i. Thickness (disc area)
                  1. Normal (I>S>N>T)
                  2. Thinned
                  3. Obliterated
               ii. Color
                  1. Pink
                  2. Pale
                     a. Optic neuropathy
                     b. Damage anterior to the lateral geniculate nucleus (LGN)
                        i. Ganglion cell axons synapse in the LGN
c. Can take up to 1 month to develop pallor after damage to axons

iii. Thickness (elevation)
   1. Normal
   2. Elevated
      a. Edema
      b. Anomalous Disc
         i. Disc drusen
         ii. Hypoplastic disc
         iii. Other
   3. Atrophic

b. Cupping
   i. Normal
   ii. Increased
      1. Glaucomatous cupping
      2. Non-Glaucomatous cupping

c. Margin
   i. Flat
   ii. Elevated
      1. Edematous
      2. Not edematous

ii. OCT
   1. Neuro retinal rim thickness
   2. Retinal nerve fiber layer thickness

b. Function
   i. VA
   ii. Color vision
   iii. Pupil Testing
      1. RAPD = unilateral or asymmetric optic nerve damage
   iv. Visual Field Testing
      1. VF should match the optic disc appearance

II. NEURO RETINAL RIM LOSS

a. Neuro retinal rim thinning (cupping)
   i. Glaucomatous
      1. Primary Open Angle Glaucoma
      2. Normal Tension Glaucoma
         a. Diagnosis of exclusion
b. Need to rule out other causes of neuro retinal rim loss
   i. Work-up may consist of
      1. Lab testing
      2. Neuro-imaging
      3. Carotid ultrasound

ii. Non-Glaucomatous
   1. Compressive
      a. Sellar mass
         i. Pituitary adenoma
         ii. Craniopharyngioma
      b. Orbital mass
   2. Ischemic
      a. Ischemic optic neuropathy
         i. Arteritic (more likely to cause NRR thinning / cupping)
         ii. Non-arteritic
      b. Other

III. NEURO RETINAL RIM PALLOR

a. Patterns of pallor
   i. Diffuse
   ii. Sectoral
      1. Temporal
      2. Bow-tie / band

b. Causes of pallor / optic neuropathy
   i. Compressive
      1. Location
         a. Orbital Mass
         b. Sellar mass
         c. Mass compressing optic tracts
      2. Etiology
         a. Primary tumor
         b. Metastasis
   ii. Traumatic
   iii. Infectious
      1. Examples of infectious associations with optic neuropathy
         a. Syphilis
         b. Lyme disease
      iv. Inflammatory
         1. Example of inflammatory associations with optic neuropathy
a. Sarcoid

v. Auto-immune
   1. Example of auto-immune associations with optic neuropathy
      a. Sjogren’s syndrome
      b. Neuromyelitis optica (NMO) / Devic’s Disease

vi. Nutritional / Toxic
   1. Vitamin B12 deficiency
   2. Folate deficiency
   3. May be associated with alcoholism

vii. Demyelinating
   1. Multiple sclerosis

viii. Mitochondrial

ix. Others

c. Work-up
   i. Lab testing – possibly including
      1. CBC
      2. ESR
      3. C-reactive protein
      4. Platelet count
      5. Vitamin B12
      6. Folic acid
      7. Methylmalonic acid
      8. Homocysteine
      9. ANA
      10. SSA / SSB (Sjogren’s)
      11. Rheumatoid Factor
      12. Anti-ds DNA
      13. ACE
      14. RPR
      15. FTA-ABS
      16. Lyme titer / Lyme IgG and IgM
      17. Serum protein electrophoresis
      18. NMO-IgG Antibody test

   ii. Neuro-imaging
      1. MRI or CT (if MRI contraindicated)
         a. Orbits
         b. Brain

   iii. Lumbar puncture
      1. When warranted
         a. Etiology remains unknown despite above work-up
         b. Progressive worsening of optic neuropathy
IV. NEURO RETINAL RIM EDEMA

a. Patterns of edema
   i. S>I>N>T
      1. Papilledema
   ii. Diffuse
   iii. Sectoral

b. Laterality
   i. Unilateral
      1. Optic neuritis
      2. Anterior ischemic optic neuropathy
   ii. Bilateral
      1. Papilledema
      2. Possibly arteritic anterior ischemic optic neuropathy

c. Associated Findings
   i. Paton’s lines
   ii. Obscuration of retinal vessels at disc margins
   iii. Hemorrhages
      1. Rare in optic neuritis
   iv. Macular star
      1. Neuroretinitis
         a. Can occur up to 1-2 weeks after disc edema

d. Causes of edema
   i. Increased intracranial pressure
      1. Causes
         a. Idiopathic Intracranial Hypertension
         b. Infectious / meningitis
         c. Venous sinus thrombosis
      2. Work-up
         a. MRI
         b. MRV
         c. Lumbar puncture (if not contraindicated)
   ii. Optic Neuritis (papillitis)
      1. Causes
         a. Demyelinating
         b. Infectious
         c. Inflammatory
d. Idiopathic

2. Possible work-up
   a. MRI
   b. FTA-ABS / RPR
   c. ANA
   d. ACE
   e. Lyme Titer

iii. Neuroretinitis
   1. Causes include
      a. Cat-scratch disease
      b. Syphilis
      c. Lyme disease
      d. Toxoplasmosis
      e. Tuberculosis
      f. Herpes and other viruses

2. Possible work-up
   a. Bartonella Quintana (for cat-scratch)
   b. Bartonella Hensalae (for cat-scratch)
   c. FTA-ABS
   d. Lyme titer / Lyme IgG and IgM
   e. Toxo IgG and IgM
   f. Chest x-ray / CT

iv. Anterior Ischemic Optic Neuropathy (AION)
   1. Types
      a. Arteritic (AAION)
         i. Cause
            1. Giant cell arteritis (GCA)
      b. Work-up
         i. CBC
         ii. ESR
         iii. C-reactive protein
         iv. Platelet count
         v. Temporal artery biopsy
      c. Non-Arteritic (NAION)
         i. Risk factors
            1. Hypertension
            2. Diabetes
            3. Hypercholesterolemia
            4. Tobacco use
            5. Sleep Apnea
         ii. Work-up
            1. R/O GCA