ABSTRACT

The course utilizes the jeopardy format to provide an interactive lecture on optic nerve conditions. Clinical characterization of optic nerve anomalies, specifically congenital vs. acquired etiologies, is emphasized. In addition, patho-physiologic correlates are discussed along with treatment and/or management modalities.

LEARNING OBJECTIVES

1. To review the normal anatomy and physiology of the optic nerve.

2. To differentiate congenital versus acquired optic nerve conditions.

3. To review the clinical characteristics of the optic nerve conditions.

4. To review the diagnostic clinical criteria, including lab testing and imaging, which may facilitate in disease diagnosis.

5. To review therapeutic medical and surgical options which may be available for treatable optic nerve diseases.

KEY WORDS

Optic nerve, congenital, acquired, treatment, neurological
I. The optic nerve
   A. Anatomy
   B. Vasculature
II. Anomalous optic nerves
   A. Congenital
      1. Minimal cupping
      2. Usually no hemorrhages
      3. Vessels emerge centrally from disc
      4. Disc may be small or large, with relative normal vessel caliber
   B. Acquired
      1. Hyperemia
      2. Disc / retinal hemorrhages
      3. Abnormal blood vessels
      4. Cotton wool spots
      5. Loss of spontaneous venous pulsation
III. Congenital optic nerve anomalies
   A. Optic nerve drusen
      1. Sporadic inheritance pattern
      2. Ophthalmoscopically visible at surface with irregular yellow glistening structures – histopathologically, presumed to be associated with accumulations of axoplasmic debris
      3. Minimal cup evident with possible hemorrhages
   B. Optic pit
      1. Grayish round depression within the disc margin tissue (typically infra-temporal)
      2. Variable size of depression; typically ranging from ¼ to ½ disc diameter
      3. Serous detachment of the sensory retina
   C. Tilted disc
      1. Fuchs coloboma and fundus ectasia (commonly infero-temporal)
      2. Temporal (commonly superior-temporal) visual field defect
      3. Situs inversus
   D. Hypoplastic optic nerves / Congenitally full nerves
      1. Vision may range from normal to subnormal levels
      2. Double ring sign
      3. Systemic associations
   F. Coloboma of the optic nerve
      1. Excavation of the optic nerve associated with faulty closure of the fetal fissure
      2. White & excavated area—typically located at the inferior portion of the optic nerve
      3. Disc malformations are associated with facial & skull defects
IV. Acquired optic nerve conditions
   A. Optic neuritis
      1. Vision loss and color vision loss
      2. Typically unilateral; however, may be bilateral
      3. Orbital pain upon eye movement
      4. Relative afferent pupillary defect
      5. Critical differential diagnoses
         a. Severe systemic hypertension
         b. Compressive optic neuropathy
         c. Multiple sclerosis
B. Bilateral disc edema
   1. Bilateral swollen and hyperemic optic nerves
   2. Papillary or peripapillary hemorrhages
   3. Critical differential diagnoses
C. Pseudotumor cerebri / Idiopathic intracranial hypertension
   1. Papilledema
   2. Normal CT / MRI of the brain
   3. Increased intra-cranial pressure on lumbar puncture with normal cerebral spinal fluid composition
D. Ischemic optic neuropathies
   1. Non-arteritic anterior ischemic optic neuropathy (N-AION)
      a. Sudden painless vision loss
      b. Afferent papillary defect
      c. Optic nerve swelling, with possible disc hemorrhages, progressing to pallor
      d. Disc at risk optic nerve head configuration in contra-lateral eye
   2. Arteritic ischemic optic neuropathy (A-AION)
      a. Sudden painless vision loss
      b. Associated symptoms include prodrome, headaches, jaw claudication, and scalp tenderness
      c. Association with giant cell arteritis
      d. Increased incidence in patients’ with polymyalgia rheumatica
F. Diabetic papillopathy
   1. Disc swelling → unilateral or bilateral
   2. Blurring of disc margins
   3. Striate hemorrhages
   4. Cotton wool spots
G. Hypertensive optic neuropathy
   1. Unilateral or bilateral disc swelling
   2. Blurring of disc margins
   3. Hemorrhages
   4. Cotton wool spots
H. Compressive optic neuropathy
   1. Slow progressive optic nerve disease
   2. Possible dyschromatopsia
   3. RAPD
   4. Optic nerve appearance (Swollen, normal or atrophic)
I. Glaucoma

V. Conclusions
A. Congenital vs. acquired
B. Ocular vs. systemic etiology for acquired entity
C. Treatment and management options