Optic Nerve: Edema, Iritis, Ophthalmitis
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Case history

- Rapid onset: demyelinating, inflammatory/infectious, ischemic, traumatic
- Gradual onset: toxic/nutritional, hereditary, compressive

Optic neuropathy features

- Reduced visual acuity
- Decreased color vision
- Relative afferent pupillary defect
- Visual field defect
- Optic disc appearance
  - Swelling
  - Pallor
  - Cupping
- Optic disc atrophy
  - Sectoral or diffuse
  - Loss of reddish hue (pallor)
  - +/- Attenuation of retinal arteries
  - +/- Cupping
  - NFL loss

Temporal atrophy causes
- Toxic and nutritional ON
- Autosomal dominant ON
- Leber’s hereditary ON
- Optic neuritis
- Superior/Inferior atrophy
  - Ischemic
- Band or bow tie atrophy
  - Chiasm or optic tract

- Differentiation from retinal disease
  - Visual acuity
  - Vision characteristics
  - RAPD
  - Sense of brightness
  - Color vision
  - Refractive error
  - Associated signs

- Differentiation from glaucoma
  - Loss of VA, color vision, and VF occur late
  - Cupping
    - More profound in glaucoma
    - Focal neuroretinal rim loss
    - Normal, pink rim tissue

- Congenital optic nerve anomalies
  - Malinserted disc
  - Tilted Disc
  - Optic nerve hypoplasia
  - Megalopapilla
  - Morning glory disc anomaly
• Peripapillary staphyloma

• Colobomatous optic disc

• Optic pit

• Myelinated nerve fiber layer

• Optic disc drusen
  o Physiologic cup is absent
  o Lack of hyperemia, dilated capillaries, or vessel obscuration
  o NFL retains its normal pattern, and the elevation does not extend to NFL
  o Anomalous vascular patterns
  o Visual field defects
    ▪ Buried drusen: 5-39%
    ▪ Visible drusen: 71-86%
    ▪ Arcuate defects
    ▪ Concentric constriction
    ▪ Enlarged blind spot

  o Investigation
    ▪ Ultrasound
    ▪ CT
    ▪ Autofluorescence
    ▪ OCT

➢ IIH and Papilledema

• Papilledema: disc edema due to raised intracranial pressure (ICP)

• Systemic symptoms
  o Headache (90-98%)
  o Nausea and vomiting (40%)
  o Pulsatile tinnitus (16-60%)
  o Dizziness

• Visual symptoms
  o Transient visual obscurations (32-80%)
  o Horizontal diplopia (30-32%)
  o Photophobia

• Normal color vision and VA
• No RAPD

• VF:
  o Enlarged blind spot (>90%)
  o Arcuate, central, paracentral, altitudinal
  o Overall restriction in late papilledema

• Optic nerve appearance
  o Bilateral, symmetric disc edema
  o Paton’s lines
  o Lack of spontaneous venous pulsation

• Investigation
  o MRI with contrast
    ▪ Empty sella
    ▪ Dilation of ON sheath
    ▪ Flattening of posterior globe
  o MRV
  o Lumbar puncture: >250 mmH2O

➤ Demyelinating optic neuritis

• Ocular symptoms
  o Acute vision loss in one eye
  o Pain behind the eye that is worsened with eye movement (87-92%)
  o Photopsia

• Symptoms of MS
  o Numbness
  o Vertigo
  o Loss of balance

• Signs
  o VA: 20/20 to NLP
  o Decreased color vision
  o Decreased contrast sensitivity
  o RAPD
  o Visual field defects
    ▪ Diffuse loss (48%)
    ▪ Focal defects (52%)
• Optic nerve appearance
  o Retrobulbar optic neuritis (67%)
  o Optic nerve pallor after 4-6 weeks

• Prognosis
  o 93% have at least 20/40; 74% have 20/20
  o Color vision defects and RAPD may persist

➢ NAION

• Etiology
  o Mean age 61-66 years
  o Caucasians
  o Males = females

• VA:
  o 67-75% are better than 20/200
  o Half are better than 20/30 to 20/60

• Signs
  o Reduced color vision
  o Inferior altitudinal visual field defect
  o Disc at risk
  o Diffuse or segmental disc edema

➢ AAION

• Etiology
  o Mean age: 75-76 years
  o Women
  o Caucasians

• Systemic symptoms
  o Headache
  o Pain on chewing
  o Pain and tenderness of temporal artery or scalp
  o Malaise
  o Anorexia
  o Weight loss
  o Fever
  o Joint and muscle pain
  o 20% have no systemic symptoms
• Ocular symptoms
  o Vision loss over hours or days
  o Usually unilateral but may be bilateral
  o Transient visual obscurations
  o Diplopia

• Signs
  o Severe vision loss
    ▪ Mean: 20/400
    ▪ Worse than 20/200 in 70-83%
  o Decreased color vision
  o RAPD
  o VF: altitudinal defect
  o IOP: low
  o Pallid edema of the optic nerve

• Prognosis
  o Systemic symptoms resolve within one week of steroid treatment
  o Optic atrophy with cupping after 6-8 weeks

➢ Inflammatory optic neuritis

  • Most common cause is sarcoidosis
  • Other evidence of intraocular inflammation is usually present
  • Investigation
    o MRI of brain and orbits with contrast
    o Laboratory testing

➢ Infectious optic neuritis

  • Various causes result in similar clinical signs and symptoms
    o Viral
    o Bacterial
    o Parasitic

  • Often differentiated based on associated signs and laboratory findings

  • Syphilis
    o History of painless ulcer at site of sexual contact
    o Uveitis
    o Interstitial keratitis
- Cat-scratch disease
  - Associated with neuroretinitis
  - History of scratch or bite from a cat 1 week to 3 months prior
  - Symptoms: fever, malaise, headache
  - Serologic testing: increased antibody titer

- Parainfectious optic neuritis
  - 1-3 weeks after a viral infection
  - Most common in children
  - Usually bilateral
  - CSF analysis
    - Lymphocytic pleocytosis
    - Elevated protein
  - Neuroimaging: enlargement and enhancement of optic nerves
  - Visual prognosis is excellent

- Post-vaccination optic neuritis
  - 1-3 weeks after vaccination
  - Spontaneous recovery
  - Vaccinations
    - Anthrax
    - Rabies
    - Hepatitis B
    - Tetanus
    - Variola
    - Combined smallpox and diphtheria
    - Combined measles, mumps, and rubella
    - Influenza

- Compressive optic neuropathy
  - Lesions in the orbit
    - Tumor
      - Optic nerve glioma
      - Optic nerve meningioma
      - Hemangioma
      - Lymphangioma
      - Dermoid cyst
      - Malignancy
    - Infection
    - Inflammation
      - Idiopathic inflammatory orbital pseudotumor
    - Thyroid eye disease
➢ Other optic nerve tumors
  • Astrocytic hamartoma
  • Melanocytoma
  • Secondary tumors (metastasis, lymphoma, leukemia)

➢ Toxic/metabolic optic neuropathy
  • Slowly progressing vision loss over weeks or months
  • Mild, bilateral disc swelling
  • Optic atrophy, most commonly of the temporal disc, occurs in later stages
  • Associated findings:
    o Vortex corneal dystrophy
    o Band keratopathy
    o Nystagmus

➢ Hereditary optic neuropathy
  • Autosomal Dominant Optic Atrophy
    o Wedge-shaped excavation of the temporal disc
  • LHON
    o Hyperemic optic nerve with obscuration of the disc margins
    o Retinal blood vessels become tortuous and dilated
    o Pallor is most pronounced in the temporal area
    o Attenuation of retinal arteries
    o Non-glaucomatous cupping

➢ Traumatic optic neuropathy
  • VA: 20/20 to NLP
  • Decreased color vision
  • RAPD
➢ Conclusion

● Careful history

● Age
  o <40 years: look for signs of typical optic neuritis
  o >40 years: look for signs of AION

● Neuroimaging, serologic testing, lumbar puncture looking for compressive, infiltrative, infectious, inflammatory, toxic or nutritional, or hereditary optic neuropathy