5.4.11. Patient With Decreased Vision: Classification & Management (XI): Posterior Optic Neuropathies (IV)

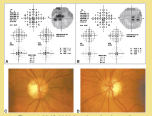
Posterior ischemic optic neuropathy

- rare
- diagnosis of exclusion
- clinical presentation
  - vision loss
    - abrupt
    - severe
  - RAPD
  - initially normal-appearing optic disc
- 3 distinct scenarios
  - perioperative
    - spine, cardiac, and head or neck procedures
    - bilateral
    - profound
    - patient and perioperative factors
    - multifactorial
      - substantial blood loss, hypotension, and prolonged anesthesia time
      - some patients with PION lost < 500 mL of blood and had systolic blood pressure >90 mm Hg
  - arteritic
    - giant cell arteritis
  - nonarteritic
    - risk factors and clinical course similar to those of NAION
- diagnostic workup
  - neuroimaging
    - patients with no history of previous surgery
    - GCA work-up in older individuals
- treatment
  - high-dose corticosteroid treatment is indicated for cases of proven GCA
  - poor prognosis for vision recovery

Optic Atrophy

- nonspecific
  - chronic phase of any of the optic neuropathies
- clinical presentation
  - vision loss
  - RAPD
  - optic atrophy
- diagnostic workup
  - when historical features and clinical signs do not suggest a specific cause
  - level of optic nerve function
    - visual acuity
    - color vision
    - perimetry testing
  - degree and pattern of atrophy
    - stereoscopic views
    - fundus photography
    - optical coherence tomography
    - retinal nerve fiber layer thickness
  - neuroimaging
    - MRI of the brain and orbits
      - gadolinium contrast
      - fat suppression
    - warranted in any case without a clear cause
    - rate of compressive lesions in patients with isolated optic atrophy
      - 20%
  - low yield in screening for
    - syphilis
    - sarcoidosis
    - vasculitis
    - vitamin B12 deficiency
    - folate deficiency
    - metal toxicity
  - for patients without a history or findings suggestive of these specific diseases
    - in the appropriate clinical or historical setting, the clinician should pursue laboratory evaluation
- treatment
  - observation
    - reasonable for patients with negative test results
    - reassess if the condition worsens or new findings develop

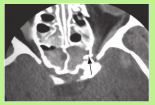
Toxic or nutritional optic neuropathy

- clinical presentation
  - gradual, progressive
    - more rapid onset may occur occasionally
  - painless
  - bilateral and symmetric
  - vision loss
    - early
      - subtle depression of central visual sensitivity
        - Amsler grid testing
        - perimetry testing
        - within the central 10°
    - late
      - central scotoma
      - decrease in visual acuity and color vision
  - optic atrophy eventually develops
  - mild to moderate optic disc edema
    - rare
- implicated medications
  - most common
    - ethambutol
    - isoniazid
    - linezolid
    - chloramphenicol
    - hydroxyquinolines
    - penicillamine
    - antineoplastic drugs
      - cisplatin
      - vincristine
  - methanol
  - ethylene glycol
    - rapid onset of severe bilateral vision loss with prominent disc edema
  - amiodarone
    - vision loss and disc edema
    - features differentiating from NAION
      - subacute onset
      - bilateral
      - diffuse rather than altitudinal visual field loss
      - slow resolution of optic disc edema over months
  - Interferon alpha
    - bilateral NAION
  - lead ingestion
    - in children
  - ethanol abuse
    - indirectly by causing malnutrition
  - dietary deficiencies
    - vitamin B12
      - 
      - Figure 4-23 (© 2020 American Academy of Ophthalmology)
      - temporal optic atrophy similar to AD optic atrophy
    - folate
    - thiamine
  - tobacco use
    - evidence is questionable
  - See Table 4-7
- differential diagnosis
  - optic neuropathies
    - maculopathies
    - hereditary
    - compressive
    - demyelinating
    - infiltrative
- diagnostic workup
  - diagnosis may be difficult, particularly with vague complaints and little objective abnormality
  - specific vitamin deficiencies are detected only infrequently on blood testing
  - neuroimaging
    - should be performed routinely to rule out a compressive etiology
- treatment
  - stopping medication
  - replacement of dietary deficiencies.
- prognosis
  - good if optic atrophy has not occurred
  - recovery is highly variable
  - occurs slowly over several months

Infiltrative optic neuropathy

- etiology
  - granulomatous inflammation
    - sarcoidosis
    - syphilis
    - tuberculosis
    - fungal infections
  - lymphoma
  - metastasis to the optic nerve
    - rare
    - breast or lung carcinoma
  - carcinomatous infiltration of the meninges at the skull base
    - involvement of multiple cranial nerves
    - optic nerves involved in 15–40%
  - onset may precede, coincide with, or follow diagnosis of the underlying malignancy
- clinical presentation
  - progressive, often severe, vision loss
    - progresses over days to weeks
  - often associated with pain
  - 1 or both eyes
  - ± other cranial nerve involvement
    - with retrobulbar infiltration, the optic nerve head may appear normal initially
    - optic disc cellular infiltrate creates a swollen appearance that may be distinct from that of simple optic disc edema
- evaluation
  - indications
    - to rule out compressive lesions
    - to confirm pachymeningeal or meningeal infiltration
  - MRI of the brain and orbits
    - fat suppression
    - gadolinium contrast
    - ± diffuse thickening and enhancement of the dura and the surrounding subarachnoid space
    - abnormalities may not be visible in the early stages
  - serologic testing
    - myeloproliferative, inflammatory, and infectious disorders
  - CSF analysis
    - malignant cells, elevated white blood cell count, and elevated protein level consistent with a neoplastic, infectious, or inflammatory cause
    - sensitivity of a single lumbar puncture is low
      - repeat testing is often necessary
- timely, correct diagnosis is essential
  - identification of associated malignancy or systemic disease may be life-saving.
  - in malignancies, palliative radiation therapy may substantially improve vision, even though the long-term prognosis is poor
    - median survival for meningeal carcinomatosis ranges from 4–9 weeks, even with aggressive therapy
  - in infectious or inflammatory disorders, antimicrobial or corticosteroid therapy may partially reverse damage

Traumatic optic neuropathy

- trauma to the head, orbit, or globe
  - direct optic nerve damage
    - optic nerve avulsion
    - laceration by bone fragments
    - laceration by foreign bodies
    - compressive optic neuropathy
      - intraorbital or intrasheath hemorrhage
  - indirect optic nerve damage
    - most common
    - may occur with relatively minor head injury
    - trauma to frontal or maxillary bones
      - transmitted forces damage the optic nerve at the orbital apex
        - shear forces on the nerve and possibly its vascular supply at its intracanalicular tether point
- clinical presentation
  - vision loss is typically immediate and often severe
    - 24%–88% no light perception vision at presentation
  - RAPD
  - optic disc usually appears normal at onset
    - becomes atrophic within 4–8 weeks
  - external evidence of injury may be scarce
- management
  - neuroimaging
    - 
    - Figure 4-24 (© 2020 American Academy of Ophthalmology)
  - orbital or cranial surgery may be necessary
    - may not affect the prognosis for the optic nerve
  - therapy for indirect traumatic optic neuropathy is controversial
    - prognosis for vision recovery has been regarded as poor
      - numerous reports describe spontaneous recovery of some visual function in a significant number of cases
    - International Optic Nerve Trauma Study
      - no clear benefit for intravenous corticosteroids
      - no clear benefit for optic canal decompression
      - study of more than 10,000 head-injury patients
        - high-dose corticosteroid group had a higher rate of mortality than the placebo group

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