

Optic Neuropathies

Optic Neuropathies Presenting As Optic Nerve Swelling

Optic Nerve Swelling with Normal or Near-Normal Vision

Pseudopapilledema

Optic Disc Drusen

- Refractile, **calcified** nodules within the ON head (visible on **ultrasound** or **CT**); bilateral (80%)
- **VF loss** (75%): arcuate, enlarged blind spot, depression; field loss **may mimic glaucoma**
- **Histopathology**: basophilic deposits stains positive for **calcium** (alizarin red, von Kossa), mucopolysaccharides (Alcian blue)
- **Associations**: **RP**, **angioid streaks** (non-PXE), **pseudoxanthoma elasticum**

Pseudopapilledema

- Optic disc drusen
- Myelinated NFL
- Astrocytic hamartoma
- Physiologic crowding

Papilledema/Intracranial Hypertension

- Optic nerve swelling specifically in the context of **increased ICP**
- **Clinical presentation**
 - **Symptoms**: TVOs/↓ VA, diplopia (usu. CN VI); HA, pulsatile tinnitus
 - VA, color vision, and pupils initially normal, permanent visual loss late
 - **Acute**: hyperemia, telangiectasis, blurred margins, NFL edema/heme, cotton-wool spots, choroidal folds, **hyperopic shift**
 - **Chronic**: pale, loss of central cup, NFL gliosis, opticiliary shunt vessels, refractile bodies, subretinal neovascularization
 - **VF loss like glaucoma**: enlarged blind spot, nasal step, arcuate defects, generalized peripheral depression, central loss
- **Causes**: **IIH**, tumor, hydrocephalus, craniosynostoses (Crouson & Apert), meningitis, demyelinating polyneuropathies, POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, skin Δs), **cerebral venous thrombosis**

Medications Associated With Pseudotumor Cerebri: "**SANTA C**"

- **S**teroid (withdrawal)
- **A**ccutane
- **N**alidixic acid
- **T**etracyclines (minocycline, doxycycline)
- Vitamin **A**
- **C**yclosporine

Oral contraceptives have been implicated but link to PTC is controversial

Idiopathic Intracranial Hypertension (IIH, Pseudotumor Cerebri [PTC])

- **Diagnostic criteria**: (a) signs/symptoms solely attributable to ↑ ICP; (b) ↑ ICP; (c) normal CSF composition; (d) normal neuroimaging studies; (e) no other etiology identified (diagnosis of exclusion)
- **Mandatory studies**: MRI ± MRV w/contrast; LP (lateral decubitus) w/opening pressure; CSF cytology/culture, medication history
- **Medical treatment**: weight loss, Diamox, Topamax, Lasix
- **Surgical treatment**: VP/LP shunt, ON sheath fenestration/decompression

Optic Nerve Swelling with Decreased Vision

Non-Arteritic Anterior Ischemic Optic Neuropathy (NAION)

- **Most common acute optic neuropathy in patients > 50 yo**
- **Risk factors**: small C/D ratio ("disc at risk"), hypertension, diabetes; smoking, hyperlipidemia, hypotension, sleep apnea
 - **Medications**: interferon-alfa, **sildenafil**, **amiodarone** (mimics NAION)
- **Clinical presentation**: acute painless monocular visual loss, altitudinal VF defect (usually inferior), sectoral hyperemic edema
- **Clinical course**: typically stable, may worsen while edema present; fellow eye involved in ~15%
- **Management**: systemic risk factor optimization, ASA for vasculopathic risk factors, sleep study, no proven treatment