OPTIC NEURITIS

" Optic neuritis is an acute monocular loss of vision caused by focal demyelination of the optic nerve "

The most common cause of unilateral painful vision loss in a young adult, typically, in patients ranging from 15-45 years.

The diagnosis itself is usually made clinically. It is the initial presentation in ~ 20% of cases of multiple sclerosis (an illness which is much more prevalent at *high latitudes*).

Pathophysiology.

Inflammatory process leading to activation of peripheral T-lymphocytes which cross the blood-brain-barrier & cause a delayed type hypersensitivity reaction culminating in axonal loss.

Clinical Features.

Patients are typically otherwise young & healthy. There may be a history of preceding viral illness. There is a female preponderance (3:1).

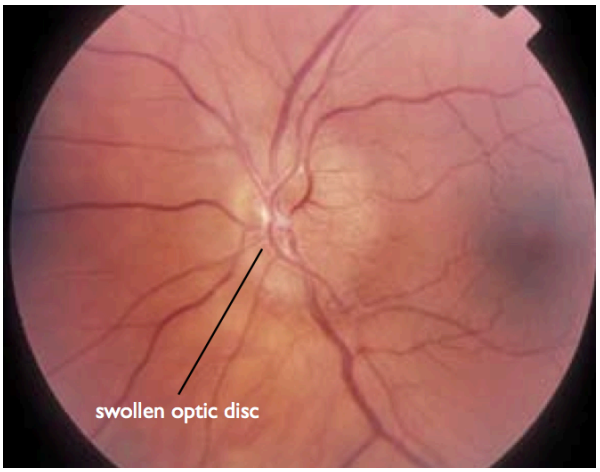
Triad for diagnosis:
1) visual loss
2) periocular pain
3) dyschromatopsia

Symptoms include:

- Painless loss of vision
  - hours to days
  - subtle vs profound
- Ocular pain (w/ eye movement)
- Reduced visual acuity (ranging from minimal loss to no light perception), colour & contrast vision
- Usually *unilateral*, though can be bilateral

Signs.

- Relative Afferent Pupillary Defect (RAPD) = Marcus-Gunn pupil
- Decreased colour vision > visual acuity
- ? Patchy visual field defects
- ? Swollen optic discs



Aetiologies / Differential Diagnoses.

Corticosteroid-responsive optic neuropathies.

- SLE, Sarcoidosis
- Behcet syndrome (vasculitis w/ oral & genital ulceration + uveitis)
- Autoimmune or chronic-relapsing inflammatory optic neuritis
- Neuromyelitis optica (ON associated with myelitis)

Other inflammatory conditions.

- Post-infectious, post-vaccination
- Acute disseminated encephalomyelitis (ADEM)

Compressive optic neuropathies.

- Primary tumours (gliomas, meningiomas, pituitary, craniopharyngioma)
- Metastases
- Aneurysms

Ischaemic optic neuropathies.

- Anterior / Posterior ischaemic optic neuropathy
- Giant cell arteritis
- Diabetic papillopathy

Infective conditions.

- TB, Syphilis, Bartonella (cat-scratch disease), Lyme disease
- Viral (measles, mumps, zoster, varicella, EBV)
- Cryptococcus, Toxoplasmosis
- Periorbital cellulitis / sinusitis.

Toxic / Nutritional neuropathies.

- B12 deficiency, ethanol/methanol, heavy-metals.

Investigations.

In a typical case of optic neuritis, without any clinical signs & symptoms of a systemic disease the yield from diagnostic tests is extremely low.

- **MRI**
    - *An important prognosticator !!*
    - Normal @ baseline = 25% risk of MS
        - 16% @ 5 years
        - 22% @ 10 years
    - One or more lesions = 75% risk of MS.

- **CSF**
    - The presence of oligoclonal bands correlates with later development of MS.
    - Those with oligoclonal bands usually have abnormal MRIs (therefore CSF sampling is unnecessary).
    - Reserve lumbar puncture for atypical presentations.

The Natural History.

Visual acuity reaches its poorest within 1 week, then will slowly improve over the next several weeks.

Spontaneous visual improvement should occur in >90% of patients within 2-3 weeks.

- 93% have VA of > 6/12 @ 1 year.
- 70% have VA of > 6/6 @ 1 year.

Progression to MS.

- 30% of patients presenting with acute optic neuritis develop *multiple sclerosis* within 5 years.
- 50% of clinically isolated optic neuritis go on to develop a second MS-defining episode by 15 years.

Treatment.

The role of steroids (predominately IV methylprednisolone) remains somewhat controversial.

- Initial studies (following 3 days of IV therapy) showed a reduced rate of MS development over 2 years of follow up.
- Subsequent review of the same cohort at 5 years post-treatment revealed no significant difference in the rate of development of MS.

RCT data on high-dose oral methylprednisolone vs placebo showed improved recovery at 1 & 3 weeks of followup, but no effect at 8 weeks (or in subsequent attack frequency).

- No role in long-term visual outcome.

Treatment with oral prednisone alone *increases risk* of recurrent optic neuritis.

Meta-analysis data (from 12 RCTs) confirms that whilst high-dose IV methylprednisolone is effective in improving short-term visual recovery, there is no significant benefit in long-term outcome.

There are specific circumstances however, where corticosteroids should be offered:

- Monocular patients
- Severe bilateral visual loss
- Occupations requiring normal visual acuity

DOSE = 1 gram IV methylprednisolone for 3 days.

IV-Ig has no beneficial effect.

References.

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