ON NOT Associated with MS

• Neuromyelitis optica (NMO, Devic’s disease)
• Acute disseminated encephalomyelitis (ADEM)
• Chronic relapsing inflammatory optic neuropathy (CRION)
• Post-vaccination-associated optic neuritis

• Consider in:
  – Atypical age (younger, older)
  – Atypical features/clinical signs
  – Rapidly progressive disease
  – Poor recovery with usual treatment
Neuromyelitis Optica

- Optic neuritis + transverse myelitis
- Simultaneous or sequential
- SEVERE inflammation
- Marked disability
  - at 5 years: 50% blind or walking aid
- Worse prognosis:
  - # relapses 1st 2 years
  - severity of 1st attack
  - associated autoimmune dz
NMO: Diagnosis

- Difficult initially; may be monocular presentation
- Clinical presentation
- Spine MRI
- Aquaporin-4 antibodies (anti-NMO-IgG)
  - up to 80% of cases
- Median age = 40
- 9:1 female preponderance
- Other autoimmune conditions
NMO: Treatment

• Acute attacks:
  – high-dose steroids (ONTT protocol)
  – plasma exchange

• Maintenance:
  – prednisone + azathioprine (Imuran ®)
  – mycophenolate mofetil (CellCept ®)
  – rituximab (Rituxan ®)

• AVOID MS MEDS (especially β-interferons)
Infectious Optic Neuropathies

- Neuroretinitis
- Lyme Optic Neuropathy
- West Nile Virus
Neuroretinitis

- Young, healthy adults; no sex predilection
- 2/3 with viral prodrome
- VA loss mild to severe
- VF defect usually central/ceccocentral
- RAPD and dyschromatopsia*
- Optic disc edema (usually)
- Stellate maculopathy
Bartonella henselae

- Children > adults; M > F
- Cats (bite/scratch) >> dogs
- Macular star evolves over days to weeks
- *B. henselae* abs in serum
- Tx: doxycycline, rifampin, azithromycin
- Most often self-limited
Lyme Optic Neuropathy

- *Borrelia burgdorferi* (tick-borne)
- Erythema chronicum migrans
- CNS involvement = days/weeks post infection
- Tertiary stage = months/years
Lyme Disease: Exam

- Inflammation of the anterior segment
- Exudative retinal detachment
- Lyme meningitis:
  - Cranial neuropathies (multiple)
  - Papilledema due to increased ICP
- MRI non-specific +/- meningeal enhancement
- ELISA to screen, Western blot to confirm
- Intrathecal Lyme Abs: ceftriaxone 2g IV/d x 1 mo
West Nile Virus

- Fever, headache, lymphadenopathy, rash
- Increasing awareness of meningoencephalitis
- Retinitis, optic neuritis, uveitis
- Chan 2006: multifocal choroiditis most common
- Beware:
  - diabetes mellitus
  - age > 50
Inflammatory ON in Systemic Disease

42-year-old man, painless vision loss OS x weeks
• Progressive “loss of detail” OD
• BCVA: 20/30 OD, NLP OS
• Color (Ishihara): 10/10 OD, 0/10 OS
• Fields: Full to confrontation OD
• Pupils: 6 mm; Grade 4 APD OD
• 2 mm proptosis OS
• SLE: Unremarkable
• Fundus: dysplastic nerve OD, advanced atrophy OS
Repeat MRI...
Whole-Body PET
Sarcoidosis

- Multisystem granulomatous inflammation
- Lymphatic system, lungs
- Similar presentation to typical ON
- All ethnic groups (> in African Americans in US)
- CNS involvement ~5% of cases
- Ophthalmic involvement ~25%
- Consider if: recurrent vision loss with steroid w/d, conjunctival/iris nodules, uveitis, lacrimal gland enlargement
- Dx dependent on TISSUE
Giant Cell (Temporal) Arteritis:

- RARE > 60; prevalence increases with age
- Acute; vision loss is severe (<20/200)
- Pale, “chalky” edema +/- cotton-wool spots

**Risk Factors:**

- Age (mean age 70)
- Associated with PMR
GCA: Systemic Symptoms

- Headache 58%
- Jaw claudication 53%
- Weight loss 31%
- Malaise 22%
- PMR 22%
- Anorexia 20%
- Fever 11%
- Neck pain 11%
- Scalp tenderness 11%

GCA: Diagnosis

• Clinical Presentation
• Labs: ESR, CRP *may* be elevated
• Temporal Artery Biopsy (***)
GCA: EMERGENCY Treatment

Goals:
1. Decrease active inflammation
2. Improve vision in the affected eye (uncommon)
3. Prevent involvement of fellow eye
4. Decrease systemic complications of vasculitis

If GCA suspected:
1. Treat first; ask questions later…
2. Begin steroids immediately (min 80 mg/d)
3. Admit to hospital (with internist co-management)
4. IV steroids 250 mg every 6 hours x 3-5 days
5. Temporal artery biopsy, best if bilateral
GCA: Myths

1. Diagnosis requires systemic symptoms.
   - 21% present ONLY with vision loss (occult GCA).

2. Diagnosis requires elevated ESR.
   - Normal ESR does not exclude GCA.

3. Steroid therapy can be tapered by protocol.
   - Each patient must be treated individually.
   - Serial monitoring w/ ESR and CRP most useful.

4. GCA always "burns itself out."
   - Long-term (9-12 months) is required.
   - May need life-long tx to prevent vision loss.
Conclusions:

- Optic neuritis is MOST common
- May be first presentation of MS; initial MRI + clinical and radiographic monitoring REGULARLY
- Other conditions can mimic demyelinating ON
- Be suspicious of atypical features and evaluate accordingly