GLAUCOMA ON A SUNDAY

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I. Glaucomatous optic neuropathy
Trobe JD, Glaser JS, Cassady J, Herschler J, DR Anderson. Nonglaucomatous excavation of the optic dsic. *Arch Ophthalmol* 1980; 98 (6) 1046-50.
A. Characteristics:
1. Generalized enlargement of cup
    a. Earliest change
2. Focal enlargement of cup
    a. ISNT rule
    b. Atrophy most typically occurs at inferior and superior poles
3. Superficial splinter hemorrhage
4. Loss of nerve fiber layer
5. Translucency of neuroretinal rim
    a. Increased cupping and pallor within the cup; NOT pallor of neuroretinal rim
6. Development of vessel overpass
    a. Nasalization of central retinal artery and central retina vein
7. Cup asymmetry
8. Peripapillary atrophy (beta zone)

II. Unilateral pallor
A. Optic nerve glioma
NR Miller. Primary tumours of the optic nerve and its sheath. *Eye* 2004; 18, 1026-1037
- Most common tumor of optic nerve, about 1% of all intracranial tumors
1. In children, almost always benign benign
    a. 75% of patients with optic nerve gliomas become symptomatic in first decade of life. 90% become symptomatic during the first two decades of life.
    b. Relationship between optic nerve glioma and NF1 well established. Reported incidence of NF1 among patients with optic nerve or chiasmal gliomas ranges from 10 to 70%
    c. Most grow slowly, self-limited, and some spontaneously regress
2. In adults, almost always malignant
    a. Second to the eighth decade of life
    b. No satisfactory treatment. Short-term success follows treatment with combined radiotherapy and chemotherapy Death is result in most cases.

B. Anterior ischemic optic neuropathy (AION)
Quigley J, DR Anderson. Cupping of the optic disc in ischemic optic neuropathy. *Trans Sect Ophthalmol Am Acad Ophthalmol Otolaryngal* 1977; 83 (5): 755-762.
1. Non arteritic AION
    a. Most common acute optic neuropathy among individuals over the age of 50
    b. Typically idiopathic. Presumably results from vascular dysregulation and impaired microcirculation of optic nerve head.

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c. Characteristics: Sudden vision loss, disc swelling, visual field defects. Fellow eye crowded, "cupless" disc or disc at risk

d. Management: No proven treatments. Manage vascular risk factors

2. Arteritic AION

a. Vasculitic occlusion of the posterior ciliary arteries.

b. Approximately 50% of eyes with AAION develop optic disc cupping

c. Characteristics: history of jaw claudication, scalp tenderness, weight loss, headache, and fatigue.

d. Management: systemic corticosteroids, prompt ESR, CRP, CBC, and temporal artery biopsy to confirm granulomatous vasculitis.

III. Bilateral optic nerve pallor

A. Tertiary syphilis

WL Bruetsch. Syphilitic optic atrophy. *AMA Arch Ophthalmol* 1953; 50 (2): 273-274.

1. Infection by spirochete *Treponema pallidum*

2. Systemic disease

a. Primary syphilis characterized by painless chancre, which appears 2 to 6 weeks after infection at the site of inoculation.

b. Secondary syphilis occurs 4 to 10 weeks following infection. Characterized by non-specific symptoms, such as fever and malaise, and generalized rash, which often involves the palms and soles.

c. Latent syphilis, disease clinically undetectable, can persist for many years.

d. Tertiary syphilis, characterized by neurologic and cardiovascular manifestations, occurs months to years following infection and may produce significant morbidity.

3. Ocular manifestations

a. Affects all ocular structures- conjunctivitis, episcleritis, scleritis, interstitial keratitis, chorioretinitis, retinitis, vasculitis, vitritis and panuveitis.

b. Uveitis, which may be unilateral or bilateral, is one of the most frequent ocular manifestations. Granulomatous features, including large keratic precipitates, iris nodules. Elevated intraocular pressure associated with uveitis, or so-called Inflammatory Ocular Hypertension Syndrome.

4. Diagnosis

a. Two types of antibody-based serum tests: non-treponemal and treponemal

i. Non-treponemal tests, Rapid Plasma Reagin (RPR) and Venereal Disease Research Laboratory (VDRL), detect antibodies directed against host antigens, such as cardiolipin, which are released following tissue damage induced by *T. pallidum.*

- Advantages: quantifiability, reflect both disease activity, response to therapy, test for reinfection. Disadvantage: RPR and VDRL have limited sensitivity—which has been suggested to be as low as 70 percent.

ii. Treponemal tests, Fluorescent Treponemal Antibody Absorption (FTA-ABS) and Microhemagglutination *Treponema pallidum* (MHA-TP), measure serum antibodies directed specifically against the *T. Pallidum*, and are highly sensitive.

- Disadvantage: not reliable in gauging response to treatment,

iii. While both treponemal and non-treponemal tests are quite specific, false positive results- collagen vascular disease, advanced age and HIV infection.

5. Treatment

a. Penicillin is treatment of choice for all stages of syphilis. Acquired syphilis with ocular involvement should be treated as neurosyphilis with intravenous penicillin G, 18 to 24 million units (MU) daily for 10 to 14 days, followed by intramuscular procaine penicillin, 2.4 MU weekly for three weeks.

b. Patients with penicillin allergy- tetracycline, doxycycline, chloramphenicol, ceftriaxone and the macrolide antibiotics. No antibiotic other than penicillin has been proven to be effective for the treatment of syphilis. Penicillin desensitization, recommended in patients with neurologic involvement.

c. Serologic testing is required both to confirm the diagnosis and to monitor response to therapy. Patients with ocular syphilis should undergo CSF testing. All patients reported to local health authority. All patients tested for HIV.

B. Autosomal dominant optic atrophy

Kline LG, JS Glaser. Dominant optic atrophy: the clinical profile. *Arch Ophthalmol* 1979; 97 (9): 245-251.

1. Most common hereditary optic neuropathy. Estimated disease prevalence 1:50,5000.
2. Typical onset of visual loss is in first or second decade of life, Visual acuity loss is commonly bilateral, symmetric and is usually mild, slow, and insidious. More than 80 % of patients maintain better than 20/200 vision. Color vision deficits also invariably present.
3. Pathophysiology

a. Mutations in OPA1 gene on chromosome 3q- encodes for mitochondrial-targeted protein that appears to be involved in mitochondrial membrane biogenesis and stabilization of membrane integrity.

4. Diagnosis

a. Optic disc atrophy shows focal, wedged-shaped temporal optic atrophy, however, diffuse atrophy may be present. Central, centrocecal and paracentral scotomas are most common visual field defects.

5. Management

Carelli V, La Morgia C, AA Sadun. Mitochondrial dysfunction in optic neuropathies: animal models and therapeutic options. *Curr Opin Neurol* 2013; 26 (1) 52-58.

a. No established medical treatment. Nutritional supplements such as vitamin B12 and C, Coenzyme-Q10, and lutein have been suggested to reduce reactive oxygen species induced stress in optic nerve. Topical agents deemed to be neuroprotective or

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antiapoptotic, such as bromonidine, have also been recommended, although, evidence anecdotal.

IV. Compressive visual pathway lesions
A. Pituitary adenoma
Ahmed II, Feldman F, Kucharczyk W, GE Trope. Neuroradiologic screening in normal-pressure glaucoma: study results and literature review. *J Glaucoma* 2002; 11 (4) 279-286.
1. Intracranial masses in only 6% of normal tension glaucoma patients. Likelihood of identifying intracranial lesions increases in patients younger than age 50, visual acuity below 20/40, optic nerve rim pallor, or vertically aligned VF defects.
2. Most common cause of chiasmal syndrome. Present at any age.
3. Classification based on size. Microadenoma (<10 mm) or Macroadenoma (>10 mm). May be secretory or nonsecretory. Classically, the nonsecretory tumor presents with vision loss, whereas patients with secretory tumors are usually referred to ophthalmologists for evaluation after having already been diagnosed because of endocrine derangements.
4. Life-threatening complication: pituitary apoplexy. Results from acute hemorrhage or infarction of tumor. Reported incidence varies between 0.6% and 10% of all pituitary adenomas.