Normal Tension Glaucoma.

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Definition & Epidemiology.

- Progressive optic neuropathy that mimics POAG without elevated IOP.
- 33% of POAG.
  - Close to 2/3 in Japan.
  - More common in females? (questionable)
    - Live longer.
- POAG and NTG coexist in same families.
IOP

- CNTGS has 24 mm Hg as cut off.
- Most studies use 21 mm Hg or less.
- Many patients have asymmetric damage with symmetric IOP.
- Drop in IOP may slow but not halt disease \(^1\).
  - 30% drop from baseline for treated.
  - Deterioration ratio. Control vs. Treated, 3:1.

Blood Flow

- Increase incidence of migraine and vasospasms.
  - Ca\(^{+2}\) channel blockers may help.
- Nocturnal hypotension.
- Sleep apnea (OSAS)
- Lower pulsatile ocular blood flow.
- Reduced peripapillary blood flow.
Immune related?

- Elevated antibodies to retinal proteins.
- 12% of NTG show increase level of monoclonal gammopathies.
  - Similar to progressive peripheral neuropathies with monoclonal paraproteinemia.
- 30% prevalence of autoimmune disorders.
Natural History of NTG

NATURAL HISTORY OF VISUAL FUNCTION AND RETINAL GANGLION CELL (RGC) LOSS WITH TIME

20/20 VF - wnl
20/40 VFD - mild
<20/200 VFD - island

Glaucoma associated RGC loss

Average life expectancy

Projected normal age-related RGC apoptosis rate

50 Years
100 Years
150 Years

Time course of condition
Natural History of NTG

NATURAL HISTORY OF GLAUCOMA PROGRESSION

OHT
20/20 VF - wnl
EARLY GLAUCOMA
20/30 VFD - mild
MODERATE GLAUCOMA
<20/200 VFD - island
ADVANCED GLAUCOMA

5 Years 10 Years 15 Years

Time course of condition

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GC-Drawings.CNV
Overlap Syndromes

NTG
Normal Tension Glaucoma

BT
Brain Tumors

HTG
High Tension Glaucoma

Others
Differential Diagnosis of cupping

- Compressive optic neuropathy
- Congenital disc anomalies - Pit, Coloboma, Morning glory syndrome, Tilted disc, Macrodisc, Papillorenal syndrome
- A-AION >>>>>> NA-AION
- Hereditary optic neuropathies - LHON, DOA, ROA, DIDMOAD
- Post-inflammatory optic neuropathy - MS
- Toxic optic neuropathy - Methanol, Paint thinner, Nutritional
- Traumatic optic neuropathy
- Periventricular leukomalacia - Prematures
- Other - Paraneoplastic, Advanced ARMD, etc.
Growth-Hormone-Secreting Pituitary Adenoma

Nerve-field mismatch in OS. Disc shape & cup asymmetry

24 y/o AA male

VA R- 20/20 L- CF
47 y/o WF
VA: R - 20/40 - cupped
L - 20/20
DX: Right Internal Carotid Artery Aneurysm
Compressive Optic Neuropathy - Glioma

DX: Chiasmal Glioma

MRI: Fat Chiasm

9 y/o WM
No NF

Band ONA, Pseudo Cup

DX: Chiasmal Glioma
Congenital - Coloboma

VF - Normal
IOP - Stable & normal
Course - No deterioration or change
Congenital - PapilloRenal Syndrome

No central vessels.
Multiple cilio-retinal vessels.
Cupped center.
Associated Renal disease.
Congenital - Tilted Disc

Common in myopes.
Difficult to tell from glaucoma.
Congenital - Macro Discs

Large-sized disc, Horizontal cup.

Normal-sized disc.

PROBLEM - Determining disc size from a simple disc exam, since normal discs vary so much.
Acute phase photo
Autosomal Dominant Optic Atrophy (Kjer type)
Hereditary optic neuropathy - LHON

Acute Phase

Chronic Phase

Leber’s Hereditary Optic Neuropathy
Hereditary optic neuropathy

DIDMOAD

- Diabetes Insipidus,
- Diabetes Mellitus,
- Optic Atrophy,
- Deafness
(Wolfram’s Syndrome)

Autosomal Recessive
WFS1 gene defect

19 y/o WM
20/200 OU
Post-Optic Neuritis optic neuropathy

1998

After MS optic neuritis

30 y/wM, ON-IS-OD:
Rcrr:
23 yrs later:
OD-20/50,
OS - 20/20

1998

2003

2003
Perinatal hypoxic brain damage. Associated neurologic deficits.
Traumatic Optic Neuropathy

16 y male
Fell from bike & hit forehead.
V- HM. Rx: Megadose steroids. V-200

Acute - 16 days.
Chronic - 90 days.
Paraneoplastic Optic Neuropathy

VA:
OD - 20/16
OS - 20/25

Vitritis - cells
Cups
Thin vessels

T
OD - 15
OS - 13

Teratoma-associated Paraneoplastic Retinopathy
Paraneoplastic Optic Neuropathy

Initial VF’s
OS OD

Post-Rx periocular steroids
OS OD

Multicystic Anterior Mediastinal Teratoma
Diagnosis & Differential

• Presentation.
  • Optic nerve cupping similar to POAG without elevated IOP.
    • Peripapillary atrophy.
    • In general quite similar to POAG.
  • Disk hemorrhage no longer exclusive of NTG, it is only a predictor of future damage.
Diagnosis & Differential

- VFs.
  - Similar to POAG but appears more advance that optic nerve status.
  - Deeper, steeper & closer to fixation?, more often superior?
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<th>Basic Diff Dx</th>
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<tr>
<td><strong>NTG</strong></td>
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Work up steps

1. History, Age
2. Visual Acuity and Color vision !!!
3. Pupils
4. Clinical course
5. Visual fields - Essential & Basic
6. Optic nerve head
7. NeuroImaGing = NIG
8. Other tests: Lab, X-R, CSF, AG
Diagnosis & Differential

- Work-up.
  - Diurnal curve.
    - At least 3 readings.
  - Optic nerve documentation.
    - Photograph, drawing.
    - GDx, HRT-II or OCT.
  - VF.
    - SWAP?
Work-Up: MRI

MRI low yield in asymptomatic patients (2/53)²

- INDICATIONS FOR MRI:
  - HISTORY - Headaches, Eye pains, Progressive visual loss, Mental changes, Diplopia, Endocrine changes, Neurologic deficits, Sudden onset.
  - OTHER - Age < 50 yrs, VA < 20/40, Dyschromatopsia,
  - VFs - Vertical step, Junction scotoma.
  - COURSE - Progression after 25-30% IOP drop.
  - ONH - NRR pallor, Pallor > cupping.
  - ONH & VF - Mismatch of findings.

² AAO: Optic nerve head and RNFL. Oph 1999;106:1414-1424
Diagnosis & Differential

• Work-up.
  • Labs.
    • CBC, r/o anemia.
    • VDRL/FTA.
    • Sed rate & C-reactive protein.
      • Temporal Arteritis.
  • Carotid doppler.
  • Sleep studies.
    • Benefits of CPAP?
Waiting & observation are options in some glaucomas: NTG, OHT

PEARLS FOR OBSERVATION:
1- Watch out for rapid progressors.
2- Do IF: few &/or low risk factors.
3- Close FU required:
   1st 2 years, do VF’s q 3-4 mos.,
   after 2 yrs, do VF’s 2x/year.
Indications for Treating NTG

Consider when there are Risk Factors as:

--Females, Migraine.
--Vasospastic phenomena: Raynaud’s, Cold hands & feet, Prinsmetal angina.
--Severe VFD’s at presentation.
--Subjective worsening of vision.
--Disc hemorrhages.
Treatment.

- Initial target.
  - 30% from baseline.
  - Adjust with frequent VF's and imaging.
- Medical Therapy.
  - PFG.
  - Topical CAI's
  - Alpha 2 agonists.
- ALT/SLT.
- Sx.