Optic Neuropathy Grand Rounds : Differentiating Glaucomatous vs. Non-Glaucomatous Optic Neuropathies

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I. Assessment of the Optic Nerve

A. Components of the Optic Nerve

1. Axons: comprise 90 per cent of the nerve
2. Astrocytes
    a) Maintain stable biochemical and structural environment
    b) Replace atrophic axonal tissue
    c) Form laminar tissue
3. Blood vessels
    a) capillaries—form a continuous meshwork that maintains the blood-brain barrier
    b) Central Retinal artery—feeds capillary plexus via 3 subdivisions
    c) Central Retinal Vein
4. Extracellular space: Small percent of tissue volume but shifts in volume and chemistry play an important role in pathologic conditions

II. Anatomical Factors Affecting Optic Disc Appearance

A. Size of the scleral canal determines cup size

1. The larger the scleral diameter, the larger the opening
2. Varies among individuals; tissue volume passing through it is more constant
3. Any “leftover” space is present in the central portion, resulting in a larger or smaller physiologic cup

B. Angle of exit of scleral canal from the eye determines the characteristics of the margin

1. <90 degree angle means RPE may end before the edge of the canal, resulting in choroidal or scleral crescent and a sharp edge

2. >90 degree angle results in more shallow sloping of the edge

III. Optic Nerve Assessment:

A. Cup

1. Determined anatomically by size of scleral canal
2. Pathologic abnormalities can result in acquired disc changes
    a) Edema can result in nerve head congestion which can obliterate the cup
    b) Atrophy can result in expansion

B. Neuro-retinal Rim

    a) Comprised of arcuate, nasal and papillomacular bundles
    b) Many neuro-ophthalmic processes have a predilection for the papillomacular bundles while others target arcuate and nasal bundles
    c) Thickness should follow the rule of "ISNT"
    d) Color is derived from fiberoptic transmission of capillary hues along healthy nerve fibers and is determined anatomically by disc size
        i. Smaller disc size results in more densely packed axons with pinker appearance
        ii. Larger disc size results in more loosely packed axons that have less "pinkness"
        iii. Color varies circumferentially with changes in density of nerve fiber
        iv. Disease processes that destroy axons or capillaries or cause increase in capillary dilation will also affect color

C. Disc Margins

    a) Degree of distinction and structural appearance is determined anatomically
    b) Acquired changes most frequently result from edematous processes
    c) Anatomical variations and pathologic process can both affect the contour of the disc margin.

D. Vasculature

    a) Anatomical variation affect branching patterns and degree of tortuosity
    b) Pathologic processes may result in loss of vasculature with subsequent death of axons and disc pallor. Capillary dilation may result in hyperemia and edema.
    c) Sheathing of larger vessels, tortuosity, changes in vessel diameter, telangiectasis or shunt formations are all indications of a pathologic process

E. Peripapillary Nerve Fiber Layer

a) Determined anatomically by the disc size and shape

b) Pathologic disorders can increase the thickness and opacify the appearance due to fluid accumulation. They can also reduce the thickness, resulting in a better ability to view underlying vasculature

c) Pigmentary changes may reflect an anatomical variation or disease process

Variations in optic disc appearance are caused by anatomical variations and disease processes that may be very difficult to distinguish clinically.

IV. Differentiating Glaucomatous from Non-Glaucomatous Optic Neuropathy

A. Compressive optic neuropathy

1. Non-glaucomatous features

a) Predilection for the papillomacular bundle

b) Early impact on visual acuity

c) Neuro-retinal rim pallor

2. "Glaucomatous" features

a) Cupping: Neuro-retinal rim pallor distinguishes it from glaucomatous cupping

b) Slowly progressive

c) Arcuate visual field changes can be seen but not without papillomacular involvement

B. Ischemic optic neuropathy

1. Non-glaucomatous features

a) Disc edema

b) Acute onset

c) Neuro-retinal rim pallor

d) Should not progress

2. "Glaucomatous" features

a) Cupping: Neuro-retinal rim pallor distinguishes it from glaucomatous cupping

b) Arcuate defects with respect for the horizontal hemianopic line can result in nasal steps

C. Papilledema

1. Non-glaucomatous features
    a) Disc edema
    b) Cup obliterated rather than enlarged
    c) Associated symptomatology
2. “Glaucomatous” features
    a) In the atrophic phase, the visual field defects precisely mimic glaucoma; visual field changes are progressive
    b) Transient visual obscurations can mimic the obscurations seen in angle closure

D. Traumatic optic neuropathy

1. Non-glaucomatous features
    a) Acute onset
    b) Stable; no visual field progression
2. “Glaucomatous” features
    a) Arcuate visual field loss with respect for horizontal hemianopic line
    b) Milder cases can be asymptomatic
    c) Can have overlying traumatic glaucoma that confounds the presentation

E. Optic Disc Drusen

1. Non-glaucomatous features
    a) Minimal to no cupping
    b) Disc often appears edematous
    c) Scalloped contour to neuro-retinal rim
2. “Glaucomatous” features
    a) Visual field loss precisely mimics glaucomatous visual field loss
    b) Spares visual acuity

F. Megalopapilla

1. Non-glaucomatous features

a) Visual fields essentially normal (enlarged blind spot)

b) Stable when observed over time

2. "Glaucomatous" features – large cup

G. Optic Pit

1. Non-glaucomatous features

a) No progression over time

b) Evidence of dysplasia

c) Often associated with decreased visual acuity

2. "Glaucomatous" features

a) Disc often appears cupped

b) Visual field defects can mimic glaucomatous visual field loss