IS THIS A Cranial Nerve III Palsy?

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Kelly A. Malloy has no financial interests to disclose.

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ABSTRACT: Clinical cases are used to demonstrate varied presentations with combinations of anisocoria, pain, diplopia, and ptosis. A stepwise approach is used to determine when the presentation may indicate an emergent cranial nerve III palsy versus another less urgent etiology.

Course Learning Objectives:
1. To emphasize the importance of the optometrist’s role in identifying signs and symptoms which suggest a neuro-ophthalmic disease process, such as a cranial nerve III palsy.
2. To understand how to conduct an examination oriented to the detection of neuro-ophthalmic disease, and to differentiate this from other conditions.
3. To discern the differential diagnoses for a variety of clinical presentations, and determine when the presentation is indeed a manifestation of a neuro-ophthalmic disease process.
4. To become familiar with the necessary diagnostic testing for a variety of neuro-ophthalmic presentations related to anisocoria, pain, diplopia, and ptosis.
5. To emphasize the need to promptly identify and refer patients presenting with emergent neuro-ophthalmic disease conditions.
6. To have a better understanding of the work-up, management, and treatment of neuro-ophthalmic disease conditions.

OUTLINE:

Conditions that may be demonstrated through actual clinical cases are included in the outline below.

CRANIAL NERVE III PALSY - ANEURYSM

Pattern of CN III Palsy:
- Hyper Deviation Which Increases In Upgaze and Reverses In Downgaze
  - Exo Which Increases Across From the Vertically Limited Eye
  - Ptosis, limited levator function
- Possible pupil involvement (may be first and only manifestation)
  - Anisocoria greatest in bright illumination
  - Larger pupil is abnormal pupil
  - Pupil may be fixed and dilated, or just somewhat larger
DEATH FROM SUBARACHNOID HEMORRHAGE
20% Of Patients With SAH Die Within 48 Hours  ****need to consider aneurysm in all CN III palsy

ANSWER BY PUPIL  INVOLVED=ANEURYSM  SPARED=VASCULOPATHIC

DOES NOT APPLY IF:
- Complicated CNIII
- Incomplete CNIII
- Relative Sparing
- 20-50 Years Of Age

ABERRANT REGENERATION OF CN III
- Pseudo Graefe Sign
- Eyelid Synkinesia
- Light-Near Dissociated Pupils

ABERRANT REGENERATION OF CN III - CAUSES
- COMMON CAUSES: Aneurysm, Tumor, Trauma
- UNUSUAL: Inflammation
- NEVER: Diabetes Mellitus

ISOLATED CN III PALSY IN ADULTS
- Undetermined 24%
- Aneurysm 21%
- Ischemia 18%
- Trauma 13%
- Neoplasm 12%

CN III PALSY Work-up ADULTS
- 20-50 YEARS - CT, MRI, MRA, Arteriogram (R/O aneurysm immediately)
- 50+ YEARS (pupil, palsy, pain) – Neuroimaging, Vasculopathic Evaluation

If Aneurysm is ruled out, other etiologies of CN III considered:
- Vasculopathic (should resolve in 3-6 months)
- Metastatic lesions to midbrain
- Carcinomatous meningitis
- Viral etiology

TONIC PUPIL
Damage to ciliary ganglion

FEATURES OF TONIC PUPIL
- “FUNNY LOOKING PUPIL”
- MID-DILATED
- LIGHT NEAR DISASSOCIATION
- “3 S’s”
  - Sector paralysis
  - Stromal spread
  - Stromal steaming
- “Flat” edges
- “Vermiform” iris movement
- Poor response to light & near or LND
• “Dilation lag” following prolonged near effort
• “Paradoxical Pupil” - aniso greater in light & dim

• Ciliary ganglion
  • 90% CB
  • 3% iris
• Aberrant regeneration of CB fibers to iris sphincter (light-near/gaze pupil dissociation)

Pharmacologic Testing for Adie's Tonic Pupil
• Weak (1/8 or 1/10) pilocarpine
• Miosis owing to “denervation supersensitivity”
• Acquired phenomenon

Tonic Pupil Imposters
• POSTERIOR SYNECHIA
• ACUTE ANGLE CLOSURE
• BITEMPORAL SPHINCTER PALSIES
• TADPOLE SHAPED PUPILS

Causes of Tonic Pupil
• local (orbit)
• infection
• inflammation
• ischemia
• tumor
• anesthesia (R-B block)
• s/p surgery
• toxicity (quinine)
• s/p laser
• trauma
• neuropathic (diabetes)
• s/p CN III palsy
• Adie’s syndrome

Types of Tonic Pupil
• LOCAL
  • VARICELLA
  • RETROBULBAR
  • ORBITAL TUMOR
  • ORBITAL SURGERY
• NEUROPATHIC
  • DIABETES
  • SYPHILIS
  • SARCOID
• IDIOPATHIC (Adie’s) – unknown etiology

Symptoms:
• 20% ASYMPTOMATIC
• 72% PUPIL RELATED
• 35% CILIARY MUSCLE RELATED
Accommodation effects
- NORMAL
- PARETIC
- TONIC
- INDUCED ASTIGMATISM @ NEAR

Features of Adie’s Tonic Pupil
- FEMALE: MALE 2.6:1
- AGE: 20-40
- 90% UNILATERAL
- FELLOW EYE @ 4% year
- DIMINISHED CORNEAL SENSATION
- DECREASED DEEP TENDON REFLEXES

Management
- PUPIL: LEAVE IT ALONE
- ACCOMMODATION
  - (SUPERSENSITIVITY CRAMP)
  - TONICITY = TROPINE
  - PARESIS = ESERINE
  - OCCLUSION

ANGLE CLOSURE GLAUCOMA
most common in people of Asian descent and people who are far-sighted

- The iris may be pushed forward up against the trabecular meshwork.
- The iris may be pulled up against the trabecular meshwork
- Resulting in increased intraocular pressure

SYMPTOMS:
- Severe eye pain
- Nausea and vomiting
- Headache
- Blurred vision and/or seeing haloes around lights (Haloes and blurred vision occur because the cornea is swollen.)
- Profuse tearing

Peripheral anterior synechiae (scarring) and adhesions may be visible between the cornea and the iris. Peripheral anterior synechiae may destroy the trabecular meshwork, and adhesions may cause permanent dilation of the iris.

HORNER SYNDROME
Anisocoria greater in dim illumination – damage to sympathetic pathway
Smaller pupil is abnormal pupil
Associated ptosis – muscle of Mueller – few mm ptosis at most

MIOSIS, PTOSIS, ANHYDROSIS
Anisocoria > Dim “Lazy Dilator” (Dilation Lag) Anisocoria > 5 Sec Than 12 Sec
Measure in bright AND dim - Greater anisocoria in dim illumination
Pancoast’s Tumor (Apical lung tumor)
TRIAD: Ptosis, Miosis, Arm Pain

CAROTID ARTERY DISSECTION
CLASSIC TRIAD: Pain On Side Of Face, Head Or Neck, Oculosympathetic Paresis Without Anhydrosis, Delayed Retinal Or Cerebral Ischemia (50-95% Of Patients)

SYMPTOMS
Exploding, Ipsilat Headache
Transient Monocular Blindness
Diplopia
Orbital, Facial, Neck, Jaw Pain
Dysguesia
Facial Numbness
Neck Swelling

SIGNS
Horner’s Syndrome
Neck Bruit Or Swelling
CN VI, IX-XII
CRAO
Cerebral Ischemia

Need to consider CAROTID DISSECTION in EVERY PAINFUL HORNER SYNDROME
Can occur with or without trauma
Medical Emergency
Horner’s with eye, head, neck pain - Pt to hospital (MRI, MRA, CTA, angiogram)

MYASTHENIA GRAVIS
NEUROMUSCULAR DISORDER
WEAKNESS & FATIGABILITY OF VOLUNTARY MUSCLE
DECREASE OF Ach RECEPTORS
AUTOIMMUNE ATTACK

PEAK INCIDENCE
YOUNGER WOMEN (15-20)
OLDER MEN (50-60)
OVERALL F:M 2:1 UNDER 30: F:M 4.5:1

23% HAVE AN ASSOCIATED IMMUNOLOGIC DISORDER

60% INITIAL PRESENTING SIGN IS AN OCULAR MANIFESTATION
90% WILL DEVELOP EYE SIGNS
15% WILL DEVELOP ONLY EYE SIGNS

MG WORK-UP
AChR ANTIBODY ASSAY (binding, blocking & modulating)
MuSK Antibody
TSH, T4, T3, thyroid antibodies (to r/o associated thyroid dysfunction)
EMG (SINGLE FIBER)
CHEST CT (to r/o thymoma in MG)

PROGNOSIS IN 5 YEARS
40% STAY OCULAR
40% CONVERT TO GENERALIZED
11% SPONTANEOUS REMISSION
85% CONVERT TO GENERALIZED

SYMPTOMATIC THERAPIES
ACH ESTERASE INHIBITORS (MESTINON (pyridostigmine) / PROSTIGMIN (neostigmine))
IMMUNOTHERAPIES
ANTICYTOKINE AGENTS
CORTICOSTEROIDS, CYCLOSPORIN
CYTOTOXIC: IMMURAN (azathioprine)
HUMORAL THERAPY
PLASMAPHERESIS, INTRAVENOUS GAMMA GLOBULIN

SURGICAL THERAPY
THYMECTOMY

DRUGS TO AVOID IN MG
IODINATED CONTRAST AGENTS
CALCIUM CHANNEL BLOCKERS
BETA-BLOCKERS: PROPRANOLOL, TIMOPTIC
NEUROMUSCULAR BLOCKING AGENTS
SUCCINLYCHOLINE, VECURONIUM
QUININE, QUINIDINE, PROCAINAMIDE
SELECTED ANTIBIOTICS
AMINOLYPOSIDES, CIPROFLOXACIN

**INTERNUCLEAR OPHTHALMOPLEGIA**
ADDUCTION DEFICIT WITH CONTRALATERAL ABDUCTING NYSTAGMUS

An exo deviation that increases across from the MLF lesion, and therefore, across from the horizontally (medially) limited eye

Localizes to the brainstem (Medial longitudinal fasciculus)
Further localize by testing convergence
  - Convergence spared = pons
  - Convergence affected = midbrain

FEATURES:
**UNILATERAL** (INO)
  • VASCULAR
  • OLDER
  • MALES
  • APOPLECTIC
  • SKEW (43%)
  • UPGAZE NYSTAGMUS

**BILATERAL** (BINO)
  • DEMYELINATING
  • YOUNGER
  • MALES = FEMALES
• PROGRESSIVE
• SKEW (13%)
• UPGAZE NYSTAGMUS

ADDITIONAL CAUSES:
• HYDROCEPHALUS
• TUBERCULOSIS MENINGITIS
• PARANEOPLASTIC ENCEPHALOMYELITIS
• HIV-CMV ENCEPHALITIS
• HEAD TRAUMA
• SLE
• MIGRAINE
• SUPRATENTORIAL AVM
• INTRACRANIAL TUMOR