Chronic Progressive External Ophthalmoplegia: Diagnosis and Management

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CPEO - Purpose

- Present clinical and diagnostic aspects of 3 patients with chronic progressive external ophthalmoplegia
- Describe the results of strabismus surgery in 2 of these patients
CPEO - Introduction

- First described by Von Graefe in 1868
- Mitochondrial encephalomyopathy
- Onset: age 11-82 years
- Progressive bilateral ptosis
- Progressive paresis of motility with exotropia
  - Diplopia rare
CPEO – Introduction con’t:
Systemic Involvement – CPEO plus

- Facial, limb muscles (60-90%)
- Endocrine (67%)
- Cardiac conduction disorders (26%)
- Ataxia and tremor (39%)
- Polyneuropathy (23%)
- Dementia/ other CNS abnormalities (13%)
- Vestibular dysfunction / hearing loss
Methods

- 3 patients with chronic, progressive, EOM weakness and strabismus referred for possible strabismus surgery
- Complete ophthalmologic, neuroophthalmic and neurologic examinations were done
- Neuro-imaging, cardiograms, audiograms, DNA studies and muscle biopsy were performed when possible
- Surgery was performed in 2 patients to reduce the exotropia and correct the head turn
Case #1 (JD) – 32 yr old woman
Ophthalmologic exam

- Marked bilateral ptosis
- Frontalis overaction
- Chin-up position
- Poor levator function
- Pigmentary retinopathy
Case #1 - EOM’s and Alignment

- **Adduction:** -5 OU
- **Abduction & Vertical gaze:** -3 OU
- **Saccadic velocity:** Poor OU
- **Alignment:** XT/XT’60, mild hypotropia OU. *No diplopia*
Case #1 Video
Case #1 – Systemic findings

- Heart block
- Mild sensory-neural hearing loss
- Proximal muscle weakness
- Ragged red fibers
- Decreased COX activity
- Elevated CSF protein

Kearns-Sayre Syndrome
Case #2: (JF) – 66 yr old woman

- Marked bilateral ptosis
- Frontalis overaction
- Chin-up position
- Exposure keratitis
Case #2: EOM’s & Alignment Pre-Op

- **Adduction:** OD -6, OS -5
- **Abduction:** OD -3, OS -2
- **Vertical gaze:** Elevation & Depression -4 OU
- **Saccadic velocities:** Poor OU
- **Alignment:** XT/XT’ 50, mild hypotropia OU
Case #2: Surgery

- Bilateral Lateral Rectus Recession
  - OD 8mm
  - OS 7mm
- Bilateral Medial Rectus Resection 7mm
- Adjustable x4
Case #2: Post-op

- **Alignment:** XT/T’ 5-10
- **Motility:** poor vertical and horizontal ductions and versions
Improved head position
Case #3 (GM) – 49 yr old man

- Previous strabismus surgery age 12 yrs
- Amblyopia OS
- Progressive LXT since age 25 yrs
- Constant diplopia
Case #3 - EOM’s & Alignment Pre-Op

- **Adduction**: OD -4, OS -2
- **Abduction**: OD -1.5, OS -2.5
- **Vertical gaze**: Elevation -3 OU, Depressions Normal
- **Saccadic velocities**: Poor OU
- **Alignment**: XT/XT’ 20, mild Hypotropia OU
Case #3 - Video
Case #3: Surgery #1

- Bilateral Medial Rectus Recession
  - OD: 5.5mm
  - OS: 6.5mm
- Left Lateral Recession from 13 to 16 mm from limbus
- Post-op
  - Head turn to left
  - LET/T’ 15
  - Abduction: OS -4
Case #3: Surgery #2

(L) Med Recess from 7 to 9 mm from limbus

Post-op #2

- **Adduction:** OD -4, OS -2
- **Abduction:** OD -1, OS -4
- **Vertical gaze:** Elevation -3 OU, Depressions Normal
- **Saccadic velocities:** Poor OU
- **Alignment:** Ortho, mild Hypotropia OU
Discussion

- CPEO is a common presenting sign of a potential multi-system disorder
  - Ptosis and XT are the most common ocular manifestations
  - Cardic workup critical
- Surgery possible for strabismus
  - Recess/Resect most common procedure
  - Large corrections usually necessary
Discussion cont. – Surgery

- Case 2
  - For cosmesis
  - Required large corrections
  - Ptosis still an issue

- Case 3
  - Complicated by previous surgery
  - Had diplopia

- Both used recess/resect procedures on adjustable sutures
References