COURSE 3

Doc, I’m Seeing Double

COPE Course 40262-NO

SCO HOMECOMING / FALL CE WEEKEND • OCTOBER 9-12, 2014

Promoting Lifelong Learning

SOUTHERN COLLEGE OF OPTOMETRY
Levels of disease

- Peripheral: neurogenic, myogenic, neuromuscular juncture, restrictive
- Nuclear: brainstem
- Supranuclear palsies: brainstem, cerebellum
- Conjugate palsies: cerebrum

Binocular diplopia patient

- Evaluate 3 cranial nerves responsible for eye movements
- Forced duction test with evaluation of orbital signs
- Evaluate other cranial nerves to determine brainstem signs
- Always consider myasthenia gravis with appropriate workup if positive signs

Oculomotor nerve palsy

- Oculomotor nerve innervates medial rectus, superior rectus, inferior rectus, and inferior oblique, with ptosis from levator muscle involvement
- Can be painful on onset, greater pain associated with aneurysm
- Presents with eye down and out with ptosis
- Pupil dilated indicates aneurysm of posterior communicating artery until proven otherwise

Oculomotor nerve palsy etiology

- Microvascular disease
- MS
- Posterior communicator artery aneurysm
- MG
- Polyneuritis
- Tumor
- Trauma
- Transtentorial compression

75 year old black female
- History high blood pressure
- No diabetes
- Binocular diplopia for 3 months
- History of cancer with intracranial metastasis
- MRI and MRA negative for vascular disease
3rd nerve palsy associated with metastatic brain cancer and surgery

Treatment of oculomotor nerve palsy
- Microvascular workup
- Orbital imaging
- Brain imaging
- Arteriogram if blown pupil
- Patch for diplopia
- May consider prism or surgery after spread of commitment several months

Isolated trochlear nerve palsy
- Remember nerves decussate above facial nerve nucleus
- Park’s three step test isolates muscles involved in cases of vertical palsies
- Must remember skew deviation during evaluation

3-Step Test Vertical Deviations
- Superior Toward
- Opposite Oblique
- Lower Away
- Same Rectus

Trochlear nerve palsy etiology
- Microvascular
- Trauma
- MS
- MG
- Aneurysm
- Tumor

Abducens nerve palsy- isolated
- Lateral rectus involved
- Abductional deficit
- Microvascular
- MS
- MG
- Brain stem tumor
- Trauma
- Polyneuritis
- Increased ICP
Congenital bilateral abducens nerve palsies
- No brainstem lesions
- No Mobius Syndrome with facial nerve palsies

Gradenigo’s Syndrome
- 6th nerve palsy
- Middle-ear infection
- Mastoiditis
- Pain on ipsilateral side of face
- Neurogenic hearing loss

Mobius Syndrome
- 6th nerve palsy
- 7th nerve palsy
- Congenital, bilateral

Differential Diagnosis
- Duane’s retraction syndrome

Duane’s syndrome
- Type 1- no abduction, retraction and narrowing of palpebral opening upon adduction and nystagmus of contralateral abducted eye
- Type 2- exotropia, reduced adduction and abduction, retraction of globe on adduction
- Type 3- poor ad and abduction, esotropic or exotropic, retraction on adduction

35 year old female
- History optomotor abnormality since childhood
- No diplopia
Bilateral Duane’s Syndrome

- 60 year old male
- History optomotor abnormality since childhood
- No diplopia

6th Cranial Nerve Palsy

- If pediatric case, with acquired ET (6th cranial nerve palsy), pontine glioma until proven otherwise!

- 72 year old male
- Recent onset horizontal diplopia
- No other symptoms
- History high blood pressure
6th nerve palsy, unknown etiology
72 year old male, no hx of diabetes, hx of HBP
All lab and MRI of brain neg

4 weeks later with no treatment

55 year old male
History HBP
6th nerve palsy right eye

47 year old male
History progressive 6th nerve palsy
Medical history negative otherwise
No other neurologic symptoms

Brainstem syndromes
- Medulla- rostral (posterior inferior cerebellar artery syndrome)
- Pons- caudal ventral, (Millard-Gubler syndrome), caudal tegmental (syndrome of Foville)
- Mid-brain- ventral (Weber), medial (Benedikt), dorsal (Parinaud’s)

Cranial nerve location in brainstem
- Rule of 4
- Nerves 12, 11, 10, 9 in medulla
- Nerves 8, 7, 6, 5 in pons
- Nerves 4, 3 in midbrain
- Nerve 2 in thalamus
- Nerve 1 in limbic lobe
Medulla lesions
- Because of location of pyramids and fact that fibers are pre-decussated within the corticospinal tracts, lesions may cause ipsilateral cranial nerve palsy at level of lesion with contralateral palsy of lower cranial nerves and hemiparesis
- Middle lesions may involve spinal thalamic tract and nucleus ambiguus with contralateral paresthesias and ipsilateral motor involvement of nerves 9, 10 and 11

Medulla
- Upper lesions may result from occlusion of the posterior inferior cerebellar artery (pica) with solitary nucleus, inferior cerebral peduncle, and vestibular nuclei involved with taste defect, and nonpyramidal motor insufficiency with nystagmus
- Medial lemniscus involvement yields fine motor and proprioception defects post decusation after nucleus gracilis and cuneatus
- Trigeminal sensory nucleus into medulla, ipsilateral facial paresthesia

Medulla
- Summary
- Causes contralateral paresis, paresthesias, taste defects, nystagmus, proprioception defects, with ipsilateral cranial nerve involvement at the level of lesion

Pons lesions
- Inferior lesions yield abducens palsy with contralateral hemiparesis with bulbar involvement
- If facial nerve is involved with mid pons lesions, is Millard-Gubler syndrome
- If trigeminal nucleus involved, mastication and facial sensory involved on ipsilateral side
- If MLF involved, contralateral INO called Foville syndrome

Pons lesions
- Pontocerebellar angle- trigeminal ganglion involvement, peripheral facial nerve palsy with intact brow and lower face involved, and acoustic-vestibular nerve involvement with hearing loss, vertigo, nystagmus- caused from acoustic- neuroma, also called Schwann cell neuroma

Pons lesion summary
- 6th and 7th nerve palsy
- Contralateral paresis
- INO possible
Midbrain lesions-summary

- Weber’s syndrome- oculomotor palsy with contralateral hemiparesis
- Benedikt’s syndrome- oculomotor palsy with contralateral hemiparesis and hemiparesthesia
- Parinaud’s syndrome- near light dissociation with superior gaze palsy, upbeat nystagmus, ptosis

- 63 year old male
- History intracranial aneurysm with surgery of midbrain
- 10 months before this exam
- Diplopia since surgery
- Left side paresis of leg and arm
- Right 3rd nerve palsy, left INO with vertical down beating nystagmus

Parinaud’s Midbrain Syndrome

- Dorsal midbrain compression
- Superior gaze palsy with up-beating nystagmus and near-light dissociation of pupils
- Most common cause is pineal gland tumor

Brown’s syndrome

- Inability to raise adducted eye above mid horizontal plane with adduction
- May have downshoot of adducting eye
- Exodeviation increased with upward movement in midline
- Positive forced duction test
- Attachment of tendon of superior oblique to Tenon’s capsule

WE COULD CERTAINLY SLOW THE AGING PROCESS DOWN IF IT HAD TO WORK ITS WAY THROUGH CONGRESS……..

- Will Rogers

MAYBE IT IS TRUE THAT LIFE BEGINS AT FIFTY. BUT EVERYTHING ELSE STARTS TO WEAR OUT, FALL OUT, OR SPREAD OUT….

- Phyllis Diller
MY LUCK IS SO BAD THAT IF I BOUGHT A CEMETERY, PEOPLE WOULD STOP DYING…

Rodney Dangerfield

ONLY IRISH COFFEE PROVIDES IN A SINGLE GLASS ALL FOUR ESSENTIAL FOOD GROUPS: ALCOHOL, CAFFEINE, SUGAR, AND FAT…

Alex Levine

BY THE TIME A MAN IS WISE ENOUGH TO WATCH HIS STEP, HE IS TOO OLD TO GO ANYWHERE…

Billy Crystal

Progressive supranuclear palsy

- Down gaze firstly restricted, then horizontal and up gaze affected
- Early saccadic defects, slowed and cog-wheel pursuits
- Degeneration of basal ganglia, midbrain tegmentum, and periaqueductal gray

Parkinsonism

- 73 year old male
- Slow development of poor eye movements
- No complaints of diplopia
- Progressive supranuclear ophthalmoplegia
- Cog-wheel movement of skeletal muscles and pursuits, glabellar reflex hyperreflexia, mask face, tremor at rest, running type walk
- Oculogyric crisis- irritative lesions causing conjugate ocular deviations which can last for several hours
- Abnormal substantia nigra in midbrain
### Cerebellar disease
- Un-monitored VOR
- Un-monitored voluntary or involuntary movements
- No pursuits
- Intention tremor
- Nystagmus

### Skew deviation
- Lesion in brainstem, cerebellum
- Seen with INO
- No cycloversional defect with hyper deviation which helps to differentiate from 4 nerve palsy
- Usually committant deviation

### Internuclear ophthalmoplegia
- MLF lesion
- Adduction defect contralateral to lesion unless lesion at 6th nerve nucleus
- PPRF lesion causes conjugate gaze palsy toward side of lesion with no adduction on ipsilateral side- 1 ½ syndrome
- Anterior INO- convergence involved
- Bilateral-MS, unilateral-basilar artery branch infarct- skew deviation due to nucleus of Cajal

### Internuclear ophthalmoplegia
- 75 year old female
- Sudden onset of binocular diplopia
- History of HBP
- No jaw claudication, temporal head tenderness, weight loss, or fever

### Brain stem MRI, intracranial MRA negative
- Sed rate, c-reactive protein, CBC, lyme titer, FTA-ABS, Hem A1-c, FANA all negative
- ETIOPATHIC INO, possibly associated with HBP

### 32 year old female
- Diabetes
- INO
Chronic Progressive External Ophthalmoplegia

- Bilateral ptosis
- Motility defects, starting with superior gaze
- No diplopia
- Kearns-Sayre disease is included
- Ragged red fibers in EOM’s
- Associated with cardiac muscle disease
- Associated with mitochondrial disease

Bell’s Palsy

- Differentiate from Ramsey-Hunt disease
- Differentiate from CNS 7th nerve palsy
- Associated with inflammation of nerve in canal in mastoid bone
- Treat with steroids
- Controversy on use of Acyclovir routinely
- Watch for exposure keratopathy and lacrimal gland insufficiency

- 48 year old white male
- Poor ocular motilities for 10 years
- No other health problems, including no heart disease
- CPEO

- 80 year old black female
- Medical history of HBP and diabetes
- Longstanding history of facial twitching
- Previous MRI of the brain negative

- Hemi-facial spasm
- Botox injections for treatment

- Blepharospasm
Myasthenia Gravis

- Always a consideration with any optomotor defect
- Acetylcholine receptor antibody titer and tension test would help to rule out
- Ice test
- Scan thymus gland

- 65 year old male
- History binocular, transient ptosis
- Transient diplopia also noted
- Acetylcholine receptor antibodies, blocking and binding 10 times normal

Eaton-Lambert Syndrome

- Paraneoplastic syndrome
- Looks and smells like MG
- Differential by EMG
- Screen for lung cancer

- 70 year old male
- Vision loss and diplopia for 6 years
- Pigmentary retinopathy, vision of counting fingers both eyes
- All test neg. except high FTA-ABS

Signs of orbital disease

- Pain
- Progression
- Proptosis
- Palpation
- Pulsation
- Periorbital changes

- Docholiectasis internal carotid artery, transient diplopia
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<tr>
<th>Orbital pseudotumor</th>
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<tr>
<td>● Idiopathic orbital inflammation</td>
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<td>● Mass effect with exophthalmos</td>
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<td>● Histopathology- cellular infiltration into orbit to include muscles, fat, lacrimal gland-eosinophilia noted, particularly in children</td>
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<td>● Optic nerve, cranial nerve involvement, muscle inflammation</td>
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<td>● Unilateral involvement in adults, can be bilateral in children</td>
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<tr>
<td>● Sudden onset of binocular diplopia</td>
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<td>● Red right eye with pain</td>
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<tr>
<td>● No other symptoms</td>
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<tr>
<td>● Medical history negative</td>
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<tr>
<td>● No vision loss</td>
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<td>● Visual fields normal both eyes</td>
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<td>● Extraocular muscle inflammation</td>
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<td>● Note orbital aching with diplopia</td>
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<tr>
<td>● Sectoral episcleral injection</td>
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<tr>
<td>● Possible positive forced duction</td>
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<td>● Differential includes collagen vascular disease, sarcoidosis, muscle tumor, Grave’s orbitopathy, polymyositis</td>
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<td>● Infectious disease of orbit</td>
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<td>● Bacterial, viral, fungal possible</td>
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<td>● With diabetes, consider mucor infection</td>
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<td>● Painful rapid exophthalmos with fever, ophthalmoplegia, vision loss</td>
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<td>● Can lead to cavernous sinus thrombosis or cranial abscess</td>
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<td>● Treat aggressively, may need orbital culture</td>
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<td>● History pain and swelling around OS for 4 days</td>
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<td>● History of liver transplant with anti-immune treatment</td>
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<tr>
<td>● White cell count 500/cubic mm</td>
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<td>● Sent to ER, admitted, started on IV vancomycin and rocephin</td>
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<th>Superior orbital fissure syndrome</th>
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<td>● Sphenocavernous syndrome</td>
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<td>● Inflammation of superior orbital fissure area</td>
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<td>● Cranial nerves II, III, IV, VI and ophthalmic V involved</td>
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<td>● Oculo-sympathetic denervation</td>
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<td>● Conjunctival congestion</td>
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<tr>
<td>● Pathology includes cavernous sinus thrombosis, Tolosa-Hunt syndrome</td>
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66 year old white male
- Diplopia for 4 days with lid shut
- History of lymphoma, in remission
- Off chemotherapy

Orbital mass extending from sphenoid wing and cavernous sinus, meningioma

Grave’s orbitopathy-thyroid related orbitopathy (TRO)
- Most common cause of exophthalmos in adults, lid retraction, lid lag, inflammation
- Usually bilateral
- Age 30-50, women 4 times more common than men
- Systemic signs of Grave’s disease-diffuse hyperplastic goiter, tachycardia, sweating, weight loss, pre-tibial myxedema

Grave’s orbitopathy
- 20% have normal chemical endocrine tests (called euthyroid)
- Immune etiology- an anti-thyroid immunoglobulin is produced called TSI(thyroid-stimulating Ig), previously called LATS for long-acting thyroid stimulator
  - This antibody binds to the TSH site on the thyroid follicular cell causing the gland to function autonomous
  - Possible decrease in T-cell suppressor activity allowing B-cells to produce more antibody

37 yo black female, hx slowly developing ptosis OD that is transient
- VA 20/20 OD, OS, fields WNL OD, OS
- Ductional restrictions superior and right gaze OD
- Bilateral exophthalmos of 30 mm OD and 26 mm OS

30 year old female
- Gradual onset of binocular diplopia
- Had no history of dysthyroid
Medicine work-up revealed abnormal thyroid function
Was also screened for M/G, negative

Orbital tumors
- Metastatic- from carcinoma of breast, lung, colon, prostate
- May expand quickly with mass effect and inflammation
- Nasopharyngeal carcinoma may easily spread due to close proximity
- Paranasal sinuses also important location of metastatic location

Primary orbital tumors
- Hamartomas- glioma and meningioma of the optic nerve- gradual vision loss with optic neuropathy, opticociliary shunt vessels
- Choristomas- dermoid cysts or tumors
- Lacrimal gland tumors

61 YEAR OLD FEMALE
- HX DIPLOPIA FOR 3 MONTHS
- PRESENTED FIRSTLY AS 6TH NERVE PALSY
- MEDICAL HISTORY NEG.
- FIRST MRI AND MRA OF HEAD NEGATIVE

75 year old female, exophthalmos for 2 years
- Sent for imaging, refused
- Sent to oculoplastics, refused
- Probable mucocele

Reviewed cranial nerve palsies
Reviewed brainstem lesions
Reviewed orbital disease
Hope you are still awake!

THANK YOU