Case 1: 78 yo Woman with Head Drop Syndrome

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History at Time of EMG

- 78 yo spanish-speaking woman with inability to hold up her head voluntarily
- Sent to EMG 7/13 to search for a neuromuscular etiology

78 yo Woman: Further History

- She was ambulating independently in 2/13
- Insidiously progressive generalized weakness ultimately leading to use of a wheelchair for the past two months
- Remote history of neck pain, not in 7/13
- Requires assistance to accomplish ADLs

Disclosures

None
Pre-EMG Neuro Exam

- CN normal, including normal tongue power and bulk bilaterally
- Sensory-identifies light touch in all 4 limbs
- Motor exam
  - SOB supine, better with partial sitting up
  - Arm >> leg weakness
  - Unable to walk or stand
  - Unable to hold neck in extension; not painful

Q! - Which of the following is the least likely cause of a head drop?

1. ALS
2. Myopathy
3. Myasthenia gravis
4. CIDP
5. An upper motor neuron process

Head Drop Syndrome: DDx

- Kyphotic deformity of the cervical spine
  - Orthopedic
    - Listhesis with or without endogenous fusion
    - Anterior wedging vert bodies
  - Dystonic-antecollis, Parkinsonian syndromes
- Neuromuscular-weakness of the cervical paraspinal muscles

Head Drop Syndrome: Neuromuscular Etiologies

- Anterior horn cell-ALS
- Nerve-CIDP, GBS
- Neuromuscular junction-MG
- Myopathy-inflammatory, mitochondrial, dystrophy, other

Isolated head drop uncommon-most assoc with other findings on neurologic exam
Head Drop and Camptocormia

- Classic: Flexion of the thoracolumbar spine
- Greek Kamptos (bend) and Kormos (trunk)
- 1818-Brodie suggests destructive spine disorders or hysterical reaction
- Consider psychogenic until latter 20th century
- May be associated with head drop, but should consider other causes

Azher SN, Jankovic J. Neurology 2005;65:355-59

Q2-Which choice below is not a limitation to the clinical assessment of paraspinal muscles?

- Cannot test muscle power attributable to a specific segmental level
- Inaccessible for muscle biopsy
- There are no norms for the degree of fatty muscle replacement on MRI with age
- Pathologists do not know the range of normal anatomy for paraspinal muscles

Neurologic Assessment of the Paraspinal Muscles

- Neurologic exam is insensitive to assess paraspinal muscles-neck ext, T/L posture
- MRI can detect fatty replacement of muscle
  - Fatty replacement increases with age
  - Lack of control groups corrected for age
- Little muscle biopsy normal data
- Ultrasound-future use to direct muscle biopsy in the OR

Electromyographic Assessment of the Paraspinal Muscles

- Can directly assess muscle function with needle EMG recording and activation of muscles by extension of the neck
- Detects short dur motor units (myopathy or nmj) or long duration units (nerve/AHC)
- Limitations: overlapping innervation at a given level, incomplete muscle relaxation
78 yo woman: EMG findings

- NCS right CTS; no other segmental nerve conduction slowing or conduction block
- Sural 11 microvolts-normal
- Needle EMG: absence of fibrillations, myopathic or neurogenic motor units
- **Needle EMG: diminished motor unit firing frequency in all muscles-no pain, no breakaway weakness, constant effort**

Q3-In general, what is the Least Likely Cause Diminished Firing Frequency of MUAPs?

1. Lack of effort
2. An upper motor neuron process
3. Pain associated breakaway weakness
4. Inattentiveness

EMG: Diminished Firing Frequency

- Needle EMG notable for diminished firing frequency (“diminished suprasegmental activation of motor units”) is due to:
  - Poor effort-not apparent in this patient; effort constant in all muscles
  - Pain-not present in this patient during the test
  - An upper motor neuron process

Additional Neurologic Exam Findings

- Normal tone in the arms and legs
- Power: Arms-barely antigrav prox, weaker distally; Legs 4- quadriceps bilat 2s distally
- Slowed FFM and foot taps-but very weak
- Hyperactive leg and arm reflexes-normal jaw jerk
- Bilateral Babinski signs
- No sensory level
Abnl Cervical MRI Findings

- Anterolisthesis of C2 on C3 and C4 on C5
- Loss of normal cervical lordosis (*kyphosis*)
- Marked narrowing of the cervical spinal canal due to soft tissue or bone
- Abnormal signal at level of the compression

“Normal” Cervical Spine MRI

78 yo woman: Cervical spine MRI

“Normal” Cervical Spine CT
78 yo woman: Cervical CT Findings

• Confirms bony canal narrowing rather than soft tissue
• C1-C2: The anterior atlantodental interval is increased and measures 5.5 mm
• Autofusion of C1 to C2
• Anterolisthesis of C2 on C3 by 4 mm

Sensitivity of abnormal signal for compressive myelopathy

• “Approximately 75%”
• If neuro exam shows progressive myelopathy and radiology severe spinal stenosis (but no increased signal) then decompress
• How distinguish cortical vs. upper cervical - possible use of DTI or other imaging

Operative report

• Pre-Op diagnosis: C1-C2 subluxation and severe stenosis with cervical kyphosis and spinal cord contusion at C1-C2
• Procedure: suboccipital craniectomy; C1 and C2 laminectomy; occiput to C5 posterior cervical fixation and posterolateral fusion;
Clinical Course

- Able to stand with assistance at discharge
- 10/13 using a walker, no SOB, more independent for ADLs
- 12/13 using a cane
- Continues to improve

Case 2: 61 yo man with acute painful neuropathies

- 4/05-painful left radial neuropathy-resolved over months
- 6/06-C3-4 spinal fusion; awake 1 mo later with SOB-bilateral phrenic nerve palsies
- 11/11 numbness and shocks over the toes of L foot; VA EMG-mononeuropathy multiplex

Q4-In general, what is the least likely cause of mononeuropathy multiplex?

1. Leprosy
2. Diabetes
3. Vasculitis
4. Brachial neuritis

Mononeuropathy multiplex

- Axonal injury to multiple single nerves
- Worldwide-leprosy, diabetes
- Other-vasculitis, sarcoid, AIDP/CIDP, mult entrapments, lyme, leukemia, lymphoma
- Only 20% with 2+ specific nerves by exam
Mononeuropathy Multiplex

- 2/3 appear by clinical exam to have distal polyneuropathy; asymmetry on EMG
- Diagnosis-Search for systemic illness assoc with vasculitis; consider nerve biopsy
- Exclude focal neuropathies at sites prone to compression

Case 2: 61 yo man with acute painful neuropathies

- 3/12-Decr light touch, pin entire left foot
- EMG-mild L peroneal/sciatic axonal neurop
- 10/12 Right upper arm pain that tapered over several months; new R arm weakness
- Recalled similar event L arm 20 yrs ago
- Motor Ex-power R arm deltoid 0, SS 3, IS 3, biceps 5; biceps reflex R 1, L 2
- Sens-Decr light touch over R lateral deltoid

Q5-What would your predict was the next diagnostic step?

1. Another EMG
2. Blood tests to search for vasculitis
3. Genetic testing
4. Joint psychiatry referral for a frustrated patient and neurologist
5. Refer to the Mayo Clinic

Hereditary Brachial Neuritis: Similarities to Classic Form

- Triggers-infections, immunizations, surgery, parturition
- Symptoms at onset-severe pain followed by focal weakness and muscle atrophy
- Spontan, slow recovery between attacks
- Management-persistent pain, rehab for focal motor loss of function
Hereditary Brachial Neuritis

- SEPT 9 gene mutation-autosomal dominant; chromosome 17 (17q25)
- Unusual Clinical Features- episodes that are pain only, weakness only, painless attack (5%), CN involvement, or lumbar plexus
- Phrenic nerve in 14%
- Diagnosis: 1) Recurrent attacks 2) positive family history 3) positive genetic testing (Athena)

Hereditary Brachial Neuritis

- Genetic variations:
  - Whole gene duplication, partial gene duplication
  - Mapped to other loci on chromosome 17
  - 15% of US families do not map to SEPT9
- Genetic Counseling
  - Penetrance 80-90%
  - Identify who at risk does not have the mutation
  - Cannot predict when symptoms will occur

Moral of the Story

The smartest neurologist is the last one to see the patient!