



## EMG Case No. 68, June 2004

### Presenting Symptom(s):

Weakness of the neck extensor with drooping of the head

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**Case-specific Diagnosis:** Isolated neck extensor myopathy “dropped head syndrome”.

**Appropriate Audience:** Residents and practicing physicians

**Learning Objectives:** After completing this educational activity, participant will be able to: (1) formulate a differential diagnosis of a case presenting with a dropped head as the chief complaint, (2) evaluate and recognize the clinical findings in a patient with focal myopathy of neck extensors, and (3) analyze the electrodiagnostic findings in a patient with focal myopathy of neck extensors.

**This case is no longer available for CME credit.**

### History

A 73-year-old, right handed woman with history of hypertension and hyperlipidemia presented to our Electrodiagnostic Medicine Laboratory with chief complaint of weakness of the neck. The patient was not able to hold her head in an upright position. Her symptoms started 13 months before we were consulted, there is no history of inciting event or trauma. Weakness of her neck is progressively worsening. She has no complaint of neck or shoulder pain, upper extremities pain or parasthesias.

- Prior to continuing, please develop a differential diagnosis and list each possible diagnosis in order of likelihood.
- Is there any additional information regarding the clinical history that might be helpful in clarifying your differential list or changing its order of priority?

### Commentary I

Differential diagnosis of a 73-year-old patient presenting with neck extensor weakness includes: Myasthenia gravis, Amyotrophic lateral sclerosis, Polymyositis, Dermatomyositis, Inclusion body myositis, Chronic Inflammatory Demyelinating Polyneuropathy, hyperparathyroidism, Carnitine deficiency and the rare condition of Isolated neck extensor myopathy also known as Dropped head syndrome.

History taking in such a case should be directed toward symptoms associated with the disorders included in the differential diagnosis. Myasthenia gravis is associated with bulbar symptoms such as diplopia, dysarthria and drooping of eyelids, and fatigue with exercise.

Amyotrophic lateral sclerosis is definitely a possibility in elderly patients although it typically presents with distal weakness, fasciculations and cramps in the absence of sensory symptoms or bowel and bladder dysfunction.



Polymyositis and dermatomyositis are inflammatory conditions that might be idiopathic, or associated with other connective tissue diseases or malignancy. It is usually associated with proximal muscle weakness with difficulty raising from chairs or with overhead activities. In dermatomyositis a skin rash is also noted. Inclusion body myositis affects males three times more frequently than females. It often affects the limbs in an asymmetric fashion.

Chronic inflammatory demyelinating polyneuropathy is a multifocal disorder that presents with more widespread weakness.

Carnitine deficiency and hyperparathyroidism are two disorders that lead to metabolic myopathies. Carnitine deficiency is associated with history of alcohol use and impaired renal or hepatic functions. Neurological changes in Hyperparathyroidism is secondary to hypercalcemia, patients usually report generalized fatigue, proximal weakness and history of renal stones.

Isolated neck extensor myopathy is a rare disorder with myopathic changes limited to the neck extensor muscles. The disease course is not progressive and weakness usually stays confined to the neck extensors.

### **History, continued**

There is no history suggestive of bulbar or limb muscles weakness or generalized fatigue. There is no history of muscle cramps or bowel and bladder dysfunction. Patient denies any complaints of weight loss, decreased appetite, skin rash, joint swelling, renal stones or bone pain. She has no history of alcohol use.

- If necessary, revise your differential diagnosis based on the additional clinical history.
- On which details of the physical examination should you focus at this point?

### **Commentary II**

Given the fact that the patient denies symptoms suggestive of a generalized disorder, the possibility of an isolated weakness of neck extensor is high in the differential diagnosis. Amyotrophic lateral sclerosis is a consideration in this elderly patient who has no sensory symptoms or sphincter dysfunction. Other possibilities are still in the list of differential diagnosis. Physical examination should stress the distribution of weakness and the presence or absence of upper motor neuron findings.

### **Physical Examination**

The patient is in no apparent distress. She is awake, alert and oriented to person, time, place and situation. She has normal speech. She has good balance. She was able to assume the standing position without difficulty, she does not have a waddling gait. She is noted to have flexed head with her chin over her chest with inability to raise her head upright. When her head position is corrected passively she is not able to maintain normal position and her head falls on her chest. The patient uses a soft collar to keep her head in an upright position. No skin rash or joint swelling noted.

Cranial nerve II – XII are intact. There are no muscle atrophy or deformities noted in the



upper extremities or both shoulder. There is no winging of the scapulae. Range of motion of the upper extremities is full and painless. Strength in upper limbs are 5/ 5, (including the shoulder girdle). Neck extensors are 2-/5. Neck flexors are 5 /5. Pinprick and light touch sensation are intact over all dermatomes. Reflexes are 1+ in the biceps, triceps , brachioradialis and knees in both sides, ankle reflexes are absent. Cerebellar function is normal. Babinski are downgoing .There is no clonus. No fasciculations are noted.

- At this point, review your differential diagnosis and revise as appropriate.
- Are there additional observations on physical examination that might be helpful in narrowing your differential list?

**Commentary III**

Based on the findings in physical examination, Amyotrophic lateral sclerosis is less likely in the absence of upper motor neuron signs and fasciculations, chronic inflammatory demyelinating polyneuropathy is less likely in the presence of symmetric reflexes and absence of multifocal weakness. Polymyositis and dermatomyositis are not high in the list with absence of shoulders and hip muscles weakness, absence of skin rash and other manifestation of connective tissue disorders. Carnitine deficiency is usually accompanied by generalized features of myopathy including proximal limb weakness, scapular winging and waddling gait.

Myasthenia gravis still a consideration as well as isolated myopathy of neck extensor muscles.

**Electrophysiologic Data**

| MOTOR NERVE CONDUCTION STUDIES |       |                  |                    |     |           |            |          |
|--------------------------------|-------|------------------|--------------------|-----|-----------|------------|----------|
| NERVE                          | SIDE  | STIM SITE        | RECORD             | cm  | AMPL (mV) | LAT (msec) | CV (m/s) |
| median                         | right | Wrist/<br>elbow  | Thenar<br>eminence | 8cm | 9<br>9    | 3.8        | 61       |
| ulnar                          | right | Wrist /<br>elbow | ADM                | 8cm | 8<br>8    | 3.0        | 62       |

| SENSORY NERVE CONDUCTION STUDIES |       |           |           |    |           |            |          |
|----------------------------------|-------|-----------|-----------|----|-----------|------------|----------|
| NERVE                            | SIDE  | STIM SITE | RECORD    | cm | AMPL (µV) | LAT (msec) | CV (m/s) |
| median                           | right | wrist     | Digit III | 14 | 44        | 2.9        | 54       |
| ulnar                            | right | wrist     | Digit V   | 14 | 41        | 3.0        | 55       |



| <b>NEEDLE ELECTROMYOGRAPHY</b>            |                           |       |     |     |     |       |       |        |       |
|---|---------------------------|-------|-----|-----|-----|-------|-------|--------|-------|
| INSERTional activity: N, sust, unsust     |                           |       |     |     |     |       |       |        |       |
| FIB: 0, 1+, 2+, 3+, 4+                    |                           |       |     |     |     |       |       |        |       |
| OTHer: 0 or fascic, myotonia, myokymia    |                           |       |     |     |     |       |       |        |       |
| EFFort: N, decr                           |                           |       |     |     |     |       |       |        |       |
| RECRuitment: N, inc or dec 1+, 2+, 3+, 4+ |                           |       |     |     |     |       |       |        |       |
| AMPliitude: N, inc or dec 1+, 2+, 3+, 4+  |                           |       |     |     |     |       |       |        |       |
| DURation: N, inc or dec 1+, 2+, 3+, 4+    |                           |       |     |     |     |       |       |        |       |
| POLyphasia: N, inc or dec 1+, 2+, 3+, 4+  |                           |       |     |     |     |       |       |        |       |
| R/L                                       | MUSCLE                    | INSER | FIB | OTH | EFF | REC   | AMP   | DUR    | POL   |
| R   | Deltoid                   | N     | 0   | 0   | N   | N     | N     | N      |       |
| R   | Biceps                    | N     | 0   | 0   | N   | N     | N     | N      |       |
| R   | Triceps                   | N     | 0   | 0   | N   | N     | N     | N      |       |
| R   | Extensor carpi radialis   | N     | 0   | 0   | N   | N     | N     | N      |       |
| R   | First dorsal interosseous | N     | 0   | 0   | N   | N     | N     | N      |       |
| R   | Cervical Paraspinals      | N     | 3+  | 0   | dec | inc3+ | dec2+ | dec 2+ | inc3+ |

- On the basis of both the clinical and electrophysiologic evaluations, formulate your diagnostic impression. List the most likely diagnosis first and follow in order with the other possibilities that are not excluded by the data. Eliminate those diagnoses not supported by the data.
- Are there additional electrophysiologic data that you feel would further delineate the diagnosis? (Remember, collecting data that are not needed for the diagnosis is costly and uncomfortable for the patient.)



**Electrophysiologic Data, continued**

| REPETITIVE STIMULATION ( 2 Hz) |       |           |                        |            |           |                  |  |
|--------------------------------|-------|-----------|------------------------|------------|-----------|------------------|--|
| NERVE                          | SIDE  | STIM SITE | RECORD                 | LAT (msec) | AMPL (µV) | TIME             | DECREMENT<br>5 <sup>th</sup> potential |
| ulnar                          | right | Wrist     | Abductor digiti minimi | 3.0        | 8         | Rest             | 0%                                     |
|                                |       |           |                        |            |           | Exercise (0 min) | 0%                                     |
|                                |       |           |                        |            |           | 1min post ex     | 0%                                     |
|                                |       |           |                        |            |           | 2 min post ex    | 0%                                     |
|                                |       |           |                        |            |           | 3 min post ex    | 0%                                     |

| REPETITIVE STIMULATION ( 2 Hz) |       |                    |                 |              |           |              |  |
|--------------------------------|-------|--------------------|-----------------|--------------|-----------|--------------|--|
| NERVE                          | SIDE  | STIM SITE          | RECORD          | LAT 9 (msec) | AMPL (µV) | TIME         | DECREMENT<br>5 <sup>th</sup> potential |
| Spinal accessory               | right | Posterior triangle | Upper trapezius | 3.1          |           | Rest         | 0%                                     |
|                                |       |                    |                 |              |           | Exercise (0) | 0%                                     |



|  |  |  |  |  |  |         |    |
|--|--|--|--|--|--|---------|----|
|  |  |  |  |  |  | 1 min   | 0% |
|  |  |  |  |  |  | post ex |    |
|  |  |  |  |  |  | 2 min   | 0% |
|  |  |  |  |  |  | post ex |    |
|  |  |  |  |  |  | 3 min   | 0% |
|  |  |  |  |  |  | post ex |    |

- Make the final revisions of your diagnostic impression(s).

### Diagnostic Impression

Based on the clinical findings and electrodiagnostic testing, our diagnostic impression is that the patient has an isolated neck extensor myopathy or the so called dropped head syndrome.

- What other diagnostic procedures (laboratory tests, etc.), if any, are needed?
- What treatment would you recommend?

Laboratory work included CPK level, acetylcholine receptor antibody titer, calcium level and serum electrolyte. All values are within normal limits.

### Commentary V

Diagnosis of isolated neck extensor myopathy, also known as “Dropped Head Syndrome” is diagnosed based on history, physical examination, electrodiagnostic studies, and histological studies. Muscle biopsy in these cases shows variability in myofibers diameter with both atrophic and hypertrophic fibers, and a decrease in central staining on NADH-TR reaction. This syndrome usually affects older patients who are 65 years of age or older.

Differential diagnosis of an elderly patient who is presenting with neck extensor weakness includes: Myasthenia Gravis, Amyotrophic lateral sclerosis, polymyositis, dermatomyositis, inclusion body myositis, chronic inflammatory demyelinating polyneuropathy, Carnitine deficiency and hyperparathyroidism. In a younger patient with the same presentation, facioscapulohumeral dystrophy, myotonic dystrophy, congenital myopathy should be considered. .

In cases of isolated neck extensor myopathy, nerve conduction studies are usually normal unless a coexisting pathology is present. The important part of the electrodiagnostic evaluation is the needle examination which shows abnormality consistent with a myopathic process mainly limited to the neck extensor muscles.

The disease process does not progress to affect other muscle groups. Weakness of the neck extensors persists and patients need to use a cervical collar to achieve normal posture.



## **Bibliography**

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