American Academy of Physical Medicine and Rehabilitation

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EMG CASE No. 60, February 2003

Presenting Symptom(s): Painful droopy shoulder and right arm weakness

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Appropriate Audience: Residents and practicing physicians

Learning Objectives: After completing this educational activity, participant will be able to: 1) Recognize patients with spinal accessory nerve lesions and resultant trapezius muscle weakness, and differentiate how this type of weakness may differ from other neurogenic or myopathic causes of weakness; 2) Explain electrodiagnostic correlates of a spinal accessory nerve lesion; and 3) Identify the anatomy of the spinal accessory nerve and be able to describe how lesions of this nerve may occur and result in limb impairment, and predict potential for recovery.

This case is no longer available for CME credit.

History

Chief Complaint: painful drooping right shoulder and right limb weakness
Presentation: A 56-year-old right-handed man presents with a history of sudden onset of right shoulder pain and drooping of his right shoulder15 months ago. With the onset of pain, he developed weakness of his right upper limb, as he is unable to raise his arm over his head, but no weakness in his hand. He describes atrophy of his right shoulder and neck muscles, but not of his arm, forearm, or hand. He reports no progression of weakness, but no improvement either. There is no history of numbness and no symptoms of weakness or pain in his other three extremities or his back. He had no slurred speech, double vision, or difficulty swallowing.

The patient had an initial consultation with a previous physician including an EMG. The EMG included nerve conduction studies with conduction velocities of the right ulnar and median nerves being normal. Concentric needle examination included testing of the first dorsal interosseous muscle, biceps, triceps, deltoid, and infraspinatus muscles, and were all recorded as normal. Needle examination of the supraspinatus muscle was reported as abnormal with fibrillation potentials, positive waves, and large amplitude, polyphasic voluntary motor units. The examiner diagnosed the patient as having a possible "C4 radiculopathy versus a suprascapular mononeuropathy."

DIFFERENTIAL DIAGNOSIS

Neuropathic causes with primarily lower motor neuron symptoms of weakness with atrophy and pain:

1. Cervical radiculopathy - compressive or inflammatory causes

2. Mononeuropathy - nerves susceptible to injury involving proximal arm weakness: long thoracic, spinal accessory, suprascapular, dorsal scapular; degenerative causes such as motor neuron disease

3. Brachial plexopathy - trauma including stretch injury, compressive from tumor invasion, idiopathic/inflammatory (Parsonage-Turner syndrome)

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Musculoskeletal causes with shoulder joint disruption and resultant disuse muscle atrophy and weakness:

4. Trauma to glenoid joint: rotator cuff tear/rupture

5. Inflammatory joint disease resulting in impairment of glenoid joint mobility

Myopathic causes with pain and focal atrophy:

6. Focal inflammatory myopathy

7. Facioscapulohumeral muscular dystrophy - often presents with asymmetry

8. Metabolic myopathy with pain and weakness: carnitine deficiency, mitochondrial deficiency, glycogen storage defect

Commentary I

Given the sudden onset of pain associated with weakness and muscle atrophy, a neuropathic lesion affecting lower motor neuron(s) is listed first in the differential diagnosis. Plexopathy may be less likely as one would expect the patient to complain of numbness. Asymmetry is classic for motor neuron diseases, but pain is atypical. Additional history surrounding the onset of symptoms will help clarify where the lesion may reside, i.e. if the onset occurred with neck movements this may represent a compressive root lesion from a disk; trauma to his neck or chest may suggest a spinal accessory or long thoracic nerve lesion respectively, or traction on his arm a lesion to the brachial plexus. Additional history regarding cancer risk and diabetes may give addition reason to suspect tumor invasion or inflammatory process involving the plexus or nerve roots.
A type of muscular dystrophy seems less likely given the patient's age and focality of muscle atrophy. However, more details of his family history and prior symptoms need to be obtained.

History, continued

The symptoms arose immediately after a lymph node biopsy to the right side of his neck. The biopsy came back as benign; he has no history of smoking, previous cancers, or diabetes.

DIFFERENTIAL DIAGNOSIS REVISED

Now with knowledge that the patient's symptoms occurred after a neck lymph node biopsy on the same side as his weak limb, trauma to nerves including the spinal accessory nerve in the neck, compression of the long thoracic nerve, or traction of the brachial plexus during surgical positioning become definite considerations. A sudden disk protrusion and compression of a cervical nerve root is also still a possibility.

Physical examination details will help define where the lesion is located by determining which muscles are atrophied and/or weak. Sensory impairment is more likely in a brachial plexus lesion. Loss of reflexes may connote a plexus or radicular lesion.

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Commentary II

REVISED DIFFERENTIAL DIAGNOSIS

1.Mononeuropathy- nerves susceptible to direct injury from lymph node biopsy include the spinal accessory nerve, indirect injury from placement during surgery from compression: long thoracic nerve, or calcium deposits at the scapular notch compressing the suprascapular nerve

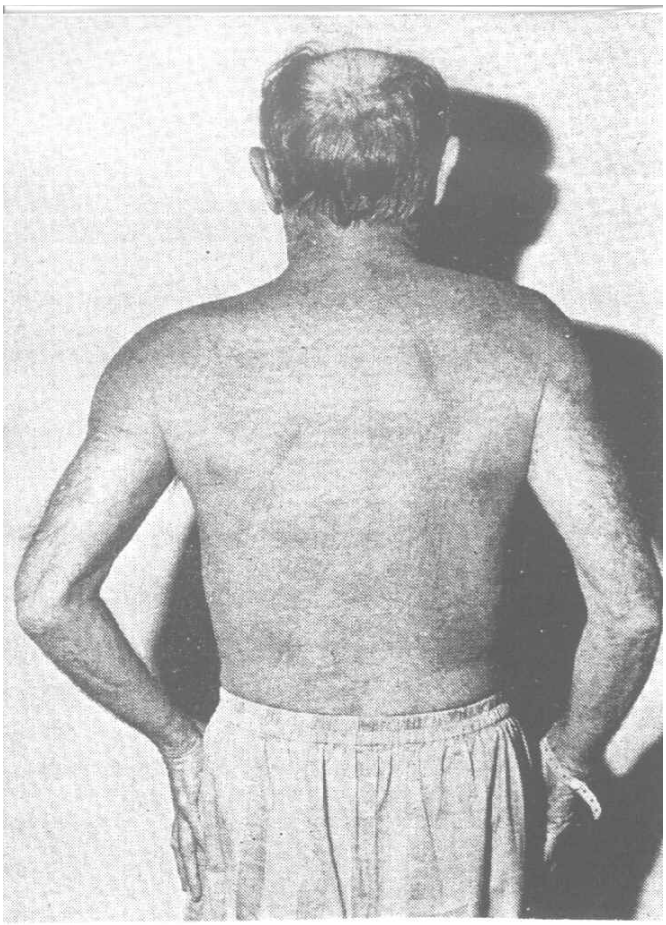
2.Brachial plexopathy from stretch injury during lymph node biopsy

3.Cervical radiculopathy- from sudden disk protrusion and compression

Physical Examination

Below is a picture of a patient with lesions causing muscle atrophy and weakness similar to the patient in our case description.

#1) 67 year old man with drooping left shoulder after removal of left neck lipoma.1
Olarte M, Adams D. Accessory nerve palsy. J Neurol Neurosurg Psychiatry 1977;40:1113-6. Reprinted with permission from the BMJ Publishing Group.



Note the patient's left shoulder is lower than the right, the left arm is externally rotated,

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and the scapular spine is more prominent on the left. (For another excellent image, see Bibliography entry #3.)

DIFFERENTIAL DIAGNOSIS

This patient had atrophy of the right trapezius muscle, with atrophy of the superior fibers causing an accentuated supraclavicular fossa, and atrophy of the middle and lower fibers resulting in prominence of the right scapular spine. There is evidence of trapezius muscle weakness in that the patient is unable to elevate his right shoulder. There is further evidence of trapezius muscle wasting in that the entire shoulder girdle is lower on the affected side.
This now narrows our differential to a spinal accessory nerve lesion coinciding with the right neck lymph node biopsy, resulting in atrophy and weakness of the right trapezius muscle.

Commentary III

Trapezius muscle function

i. The trapezius muscle supports the shoulder girdle; with weakness the scapula descends and is displaced laterally so there is slight winging of the scapula.

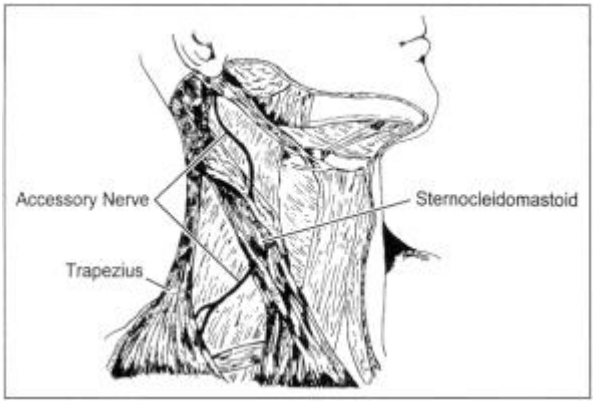
ii. The trapezius is needed to stabilize the scapula with arm movements such as shoulder abduction. Our patient was unable to actively abduct his arm above 90 degrees.

iii. Shoulder abduction can be compensated by rhomboids and levator scapula stabilizing and elevating the scapula in place of the trapezius so shoulder abduction can be fully strong.

Commentary IV

Spinal Accessory nerve anatomy

Reprinted from The American Journal of Surgery, Vol. 180, Nason R et al, "The Anatomoy of the Accessory Nerve and Cervical Lymph Node Biopsy," 241-3, 2000 with permission from Excerpta Media Inc.



Cranial nerve XI, the spinal accessory nerve, leaves the skull through the jugular foramen, passes across the dens, and descends obliquely to pass through

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the sternocleidomastoid muscle to then enter the posterior triangle of the neck.

Here the nerve is very susceptible to trauma as it covered only by skin and subcutaneous tissue before it innervates the trapezius muscle.

Electrophysiologic Data

SENSORY NERVE CONDUCTION STUDIES							
NERVE	**SIDE**	**STIM SITE**	**RECORD**	**cm**	**AMPL**	**LAT**	**CV**
Median	Right	Wrist	Index	14	22.4	3.4	51.9
Ulnar	Right	Wrist	5th	14	19	3.2	56.0

MOTOR NERVE CONDUCTION STUDIES							
NERVE	**SIDE**	**STIM SITE**	**RECORD**	**cm**	**AMPL**	**LAT**	**CV**
Median	Right	Wrist	Thenar	7	6.1	4.2	
Median	Right	Elbow	Thenar	203	5.9	8.9	53.6
Ulnar	Right	Wrist	Hypothenar	7	14.7	3.6	
Ulnar	Right	Below elbow	Hypothenar	195	13.2	8.4	52.1
Spinal accessory	Right	Neck	Trapezius	8	6.1	4.5	
Spinal accessory	Left	Neck	Trapezius	8	7.4	2.8	

NEEDLE ELECTROMYOGRAPHY
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+
AMPlitude: 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	Upper trapezius	N	0	0	Decr	D 2+	I 2+	I 1+	I 2+
L	Upper trapezius	N	0	0	N	N	N	N	N

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R	Supraspinatus	N	0	0	N	N	N	N	N
R	Infraspinatus	N	0	0	N	N	N	N	N
R	Biceps brachii	N	0	0	N	N	N	N	

The electrophysiologic data support a right spinal accessory nerve lesion with a significantly prolonged spinal accessory motor evoked distal latency relative to the left, and a decrease in amplitude relative to the left. Concentric needle examination demonstrates old denervation with reinnervation of the superior fibers of the right trapezius muscle, but normal needle examination of the supraspinatus and infraspinatus muscles, and of the contralateral trapezius.

Additional needle examination could have been performed on the cervical paraspinal muscles to rule out a superimposed radiculopathy, and deltoid muscles as the patient had some impairment of abduction beyond 90 degrees. Needle study could also have been done on the middle and lower trapezius fibers on the affected side.

Diagnostic Impression

The diagnosis of a right spinal accessory nerve lesion is made by history and physical examination demonstrating right trapezius muscle atrophy and weakness. The electrophyisologic data are supportive of this diagnosis. Given that the patient's symptoms coincided with a lymph node biopsy of his neck on the same side of his nerve lesion, an iatrogenic injury to the nerve appears to be the cause of the spinal accessory neuropathy.

The patient's previous EMG study reported abnormalities of the right supraspinatus muscle (as described above), although there was no mention of trapezius muscle testing. The anatomy of these two muscles is such that a portion of the trapezius muscle lies superficial to the supraspinatus muscle. Therefore, the EMG examiner may actually have placed the needle in the right trapezius muscle and not the deeper supraspinatus. Supraspinatus and infraspinatus muscle testing requires the examiner to penetrate muscles either superior or inferior to the palpable scapular spine until the needle reaches resistance against the scapula. The examiner then pulls the needle back slightly to ascertain that the electrode is in the deeper spinati muscles rather than the superficial trapezius muscle.

Commentary V

Trauma to the spinal accessory nerve has been reported from war wounds, lymph node biopsies, neck resections, compression by local tumors, carotid endarterectomies, neck biting, bee stings, and face lifts, among others. These arise because of the nerve's superficial location in the posterior triangle of the neck[4]. However, the nerve is also susceptible to stretch injury. Since the nerve passes through the sternocleidomastoid muscle, it has traction placed upon it when that muscle contracts to turn the head away from the affected side and the shoulder is passively pushed downward. The nerve is then stretched between the posterior border of the sternocleidomastoid and anterior border of the trapezius muscles (see figure 2). Tractive forces are suspected in sporting injuries that forcefully push downward on the shoulder or prolonged heavy lifting, depressing the shoulder, as greater than a 10% increase in the nerve resting length can result in neurapraxia (conduction defect without structural degeneration)[5].

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Friedenberg et al reviewed 56 cases of spinal accessory neuropathy, demonstrating no EMG correlation to outcome. However, three aspects of clinical presentation did predict a poor outcome: injury to the dominant arm, impaired arm abduction, and scapular winging[6].

Treatment includes aggressive pain control, as these patients often complain of significant pain that can impair shoulder range of motion and result in frozen shoulder syndrome. Muscle strengthening of deltoid, levator scapula, and rhomboids can compensate for weakened trapezius muscle function. Surgical exploration and/or grafting are recommended if there is evidence of complete nerve palsy from trauma[4].

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