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EMG Case No. 87, May 2007

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Presenting Symptom(s): Right shoulder pain and weakness.

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Disclosure: BJ Fuller, None; L DiPonio, None.

CME Credit Information: CME credit is available for three (3) years from the date of publication. CME credit for completing this case study must be claimed online through your Portfolio via the Academē Web site (www.me.aapmr.org).

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Appropriate Audience: Residents and practicing physicians.

Learning Objectives: After completing this educational activity, participant will be able to: (1) formulate a differential diagnosis of post-operative shoulder pain and weakness; (2) summarize etiologies of spinal accessory nerve palsy; and (3) electrophysiologic evaluation of the spinal accessory nerve.

Level of Difficulty: Advanced

History

The patient is a 60 year old male with right shoulder pain and weakness presenting for electrophysiologic evaluation 26 days after a 14 hour thoraco-abdominal aortic aneurysm repair.

1. Prior to continuing, please develop a differential diagnosis and list each possible diagnosis in order of likelihood.
2. 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

The differential diagnosis of a patient presenting with post-operative shoulder pain and weakness is protean. Consideration of a neurologic cause must be the primary focus. Neurologic conditions that may present with shoulder pain and/or weakness include compression mononeuropathy, brachial plexopathy, cervical radiculopathy, neuralgic amyotrophy and mononeuritis multiplex. Other conditions that may be considered include adhesive capsulitis, heterotopic ossification, rotator cuff pathology, subacromial bursitis, venous thrombus, glenohumeral or acromioclavicular joint pathology and vascular thoracic outlet syndrome.

A detailed history should be obtained focusing on the presentation and acuity of symptoms as well as a thorough review of the operative record.

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History, continued

The patient's past medical history is significant for hypertension, morbid obesity, gastroesophageal reflux disease, dyslipidemia, chronic obstructive pulmonary disease and tobacco abuse. Previous functional level was independent.

Review of the operative report indicates that the patient was placed in the right lateral decubitus position. The left internal jugular vein was cannulated to provide vascular access. Under general anesthesia a sternotomy was performed and incision extended inferiorly to the aortic bifurcation. Replacement of the entire thoraco-abdominal aorta from the left subclavian to the aortic bifurcation as well as a splenectomy was completed.

Post-operatively multiple areas of right neck, upper back and arm bullous skin breakdown were noted. Further post operative history was significant for atrial fibrillation, type two diabetes mellitus and two right hip stage two pressure wounds. Right shoulder weakness and pain was noted by post-operative day five. Multiple post-operative chest X-rays were significant only for minimal bilateral pleural effusions and minor atelectasis.

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

Commentary II

The peri-operative time course makes neuralgic amyotrophy and subacromial bursitis less likely. Mononeuritis multiplex is unlikely as it is classically considered a painless weakness. Lack of radiographic evidence of a first rib fracture or hemidiaphragm elevation decreases the probability of a brachial plexopathy or upper cervical nerve root avulsion respectively.

The physical exam should focus on testing range of motion, strength and reflexes of the extremities to assess for asymmetries.

Physical Examination

The patient's physical exam from the fifth postoperative day revealed limited right shoulder active range of motion in flexion and abduction to 80 degrees due to pain. The patient's cervical spine had full active range of motion in all planes. Shoulder passive range of motion was painless and to 180 degrees for flexion and abduction. The neurologic exam revealed symmetric 2+/4+ reflexes of the bilateral biceps, brachioradialis and triceps. The skin reveals a 10 cm by 4 cm right sided neck stage 2 pressure wound and pitting edema of the right upper extremity.

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

Commentary III

Adhesive capsulitis and heterotopic ossification may be post-surgical sequelae but are more often present during a subacute to chronic time course and without full passive joint range of motion. The lack of pain with passive range of motion makes the glenohumeral and acromioclavicular joints less likely causes of the patients symptoms.

A thorough shoulder exam testing the strength of all rotator cuff muscles and for shoulder impingement signs is warranted. A possible dermatomal pattern to the shoulder pain should be assessed.

Physical Examination, continued

Further physical examination reveals full strength of the bilateral shoulder internal and external rotators though significant scapular winging is noted on the right with resisted shoulder external rotation. Hawkins test is negative

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bilaterally. No venous cords are palpated in the right upper extremity. Adson's test was negative on the right. Sensation is intact to light touch and pinprick throughout.

1. If necessary, revise your differential diagnosis based on the additional physical findings.
2. Design your approach to the electrophysiologic examination based on the existing data.

Commentary IV

The normal strength of the shoulder internal and external rotators coupled with the lack of shoulder impingement signs point away from a rotator cuff pathology. The winging of the right scapula is suspicious for a long thoracic nerve or spinal accessory nerve injury. The winged scapula and shoulder pain raises the specter of neuralgic amyotrophy despite the peri-operative time course. A sensory exam without abnormality in a specific dermatome makes an acute radiculopathy less likely. The lack of venous cords and the negative Adson's test make a venous thrombus or thoracic outlet syndrome less likely.

A focused electrophysiologic exam of the right upper extremity should include: nerve conduction studies of the right median sensory, radial sensory as well as the bilateral lateral antebrachial cutaneous and spinal accessory nerves. A needle examination of the right trapezius, sternocleidomastoid, cervical paraspinals, rhomboids, supraspinatus, serratus anterior, and deltoid muscles should also be performed.

Electrophysiologic Data

SENSORY NERVE CONDUCTION STUDIES							
NERVE	**SIDE**	**STIM SITE**	**RECORD**	**cm**	**AMPL**	**LAT**	**CV**
Median	R	wrist	2nd digit	14	22	3.2	
Radial	R	forearm	1st digit	10	35	2.9	
Lateral antebrachial cutaneous	R	Antecubital fossa	Lateral forearm	12	21	2.2	
Lateral ante brachial cutaneous	L	Antecubital fossa	Lateral forearm	12	18	2.1	

MOTOR NERVE CONDUCTION STUDIES							
NERVE	**SIDE**	**STIM SITE**	**RECORD**	**cm**	**AMPL**	**LAT**	**CV**
Sp Acc	R	Post triangle	Trapezius	9	NR		
Sp Acc	L	Post triangle	Trapezius	9	6.9	2.2	56
3 month follow up							
Sp Acc	R	Post triangle	Trapezius	9	2.7	3.4	55
Sp Acc	L	Post triangle	Trapezius	9	7.1	2.3	53

The morphology of the right spinal accessory motor response recorded at the trapezius muscle demonstrated marked temporal dispersion consistent with incomplete re-myelination.

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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	sust	3+	0	N	0			
R	Lower Trapezius	sust	3+	0	N	0			
R	Deltoid	N	0	0	N	N	N	N	N
R	Supraspinatus	N	0	0	N	N	N	N	N
R	Serratius Anterior	N	0	0	N	N	N	N	N
R	Sternocleidomastoid	N	0	0	N	N	N	N	N
R	Rhomboids	N	0	0	N	N	N	N	N
R	Cervical Paraspinals	N	0	0					
	3 month follow up								
R	Upper Trapezius	sust	1+	0	N	dec1+	dec1+	inc3+	inc3+
R	Lower Trapezius	sust	1+	0	N	dec1+	dec1+	inc3+	inc3+

The electrophysiologic testing of the spinal accessory nerve is fairly straight forward. Motor conduction studies are done with the active recording electrode placed over the muscle belly of the Trapezius and the recording electrode placed distally on the ipsilateral acromion. Stimulation is performed posterior to the mid-sternocleidomastoid muscle. During the needle exam of the Trapezius muscle care must be taken to avoid passing through a severely atrophied muscle to sample supraspinatus and therefore seeing normal motor unit action potentials or even worse piercing the apices of the lungs and causing a pneumothorax. 1

1. 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.
2. 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

No further data.

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

Diagnostic Impression

The clinical and electrodiagnostic examination indicate a severe injury to the right spinal accessory nerve in the right posterior cervical triangle.

The normal needle exam of the supraspinatus and serratius anterior muscles make a mononeuropathy or neuralgic amyotrophy involving the suprascapular or long thoracic nerves unlikely. The normal needle exam of the deltoid and cervical paraspinals point away from a radiculopathy. Finally, the needle exam findings of denervation only in the Trapezius muscle and the normal sensory studies of the median sensory, radial sensory, and lateral antebrachial cutaneous nerves make a brachial plexopathy less probable.

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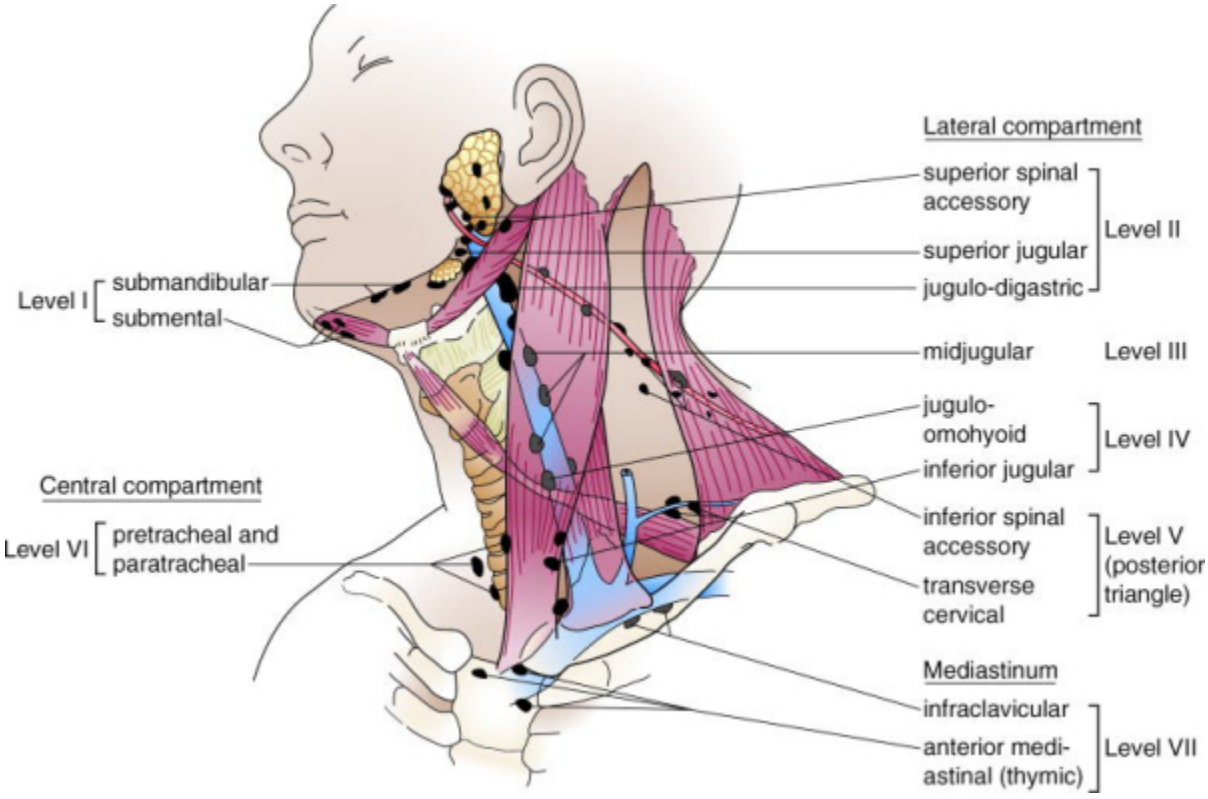
1. What other diagnostic procedures (laboratory tests, etc.), if any, are needed?
2. What treatment would you recommend?

Commentary V

The electrophysiologic data likely represents a severe injury to the right spinal accessory nerve. This is supported by the initial study revealing complete denervation and unobtainable spinal accessory motor conduction studies followed by a subsequent exam, three months later, that revealed evidence of significant reinnervation and re-myelination. These findings are consistent with axonotmesis, likely a Sunderland type two nerve injury. This type of nerve injury may be secondary to focal ischemia as might be caused by a compressive lesion. The rapid time course of recovery may be explained by the faster axonal regeneration found in more proximal peripheral nerve lesions. The increased duration of the compound motor action potential is likely due to the incomplete myelination of the immature axons.

The etiology of this patient's injury is likely due to a compressive mononeuropathy of the spinal accessory nerve, at the right posterior triangle of the neck that occurred during his prolonged surgery. This is supported by the initial electrophysiologic study revealing active denervation of the right Trapezius muscle and the unobtainable spinal accessory conduction studies. This is coupled with the absence of denervation in the right sternocleidomastoid and the clinical findings of a large stage two pressure wound covering the right side of the neck. The winging of the right scapula with resisted external shoulder rotation and/or abduction of the shoulder is almost pathognomonic for an injury to the spinal accessory nerve. The lack of the expected atrophy of the right trapezius on physical exam is easily explained by the patient's morbid obesity.



Reprinted from Townsend: Sabiston Textbook of Surgery, 17th Ed., Courtney M. Townsend Jr, MD, "Right Spinal Accessory Nerve Palsy", © 2004, with permission from Elsevier.

The Spinal Accessory nerve is conventionally thought to be purely motor, though histological evidence of some sensory nerve fibers has been reported. 21 It is derived from the C1-C4 cervical ventral rami which combine and

ascend through the foramen magnum to join the 11th cranial root and return through the jugular foramen with the Vagus nerve. It then accompanies the internal jugular vein before piercing the sternocleidomastoid muscle. 1, 3, 6 There it supplies a branch to the sternocleidomastoid muscle before running superficially through the posterior cervical triangle, covered only by skin, subcutaneous tissue and cervical fascia, to innervate the trapezius muscle. 1, 3 At this point it is approximately 1 mm in diameter. 17

The trapezius muscle is a shoulder stabilizer, contributing to scapula-thoracic rhythm by elevating; rotating and retracting the scapula, therefore injury to the spinal accessory nerve can lead to apparent weakness of the shoulder abductors and internal rotators. 10

There are several classic physical exam findings. One is the inability to actively abduct the arm greater than 80 degrees, but retain full painless passive range of motion. 3, 5 Scapular winging that is worse with resisted shoulder external rotation or abduction is a cardinal sign. 11 Of course, atrophy of the trapezius and dropped shoulder is often evident. Finally, shoulder pain is present which is likely due to loss of shoulder girdle support and traction on the brachial plexus or suprascapular nerve. 3, 5 Rarely, this dropped shoulder might also result in a neurovascular thoracic outlet syndrome from compression of the axillary artery. 2

Injury to the spinal accessory nerve is well documented in the medical literature. A myriad of causes have been documented but most commonly the nerve is injured iatrogenically in the posterior cervical triangle during cervical lymph node biopsy. 1, 3, 6 Most surgical injuries are due to division of the nerve but stretch injury from retraction, though rare, has also been reported. 4 Other surgical causes have been reported including carotid endartectomy, radical neck dissection and even a previous case of a spinal accessory nerve palsy following a thoracotomy. 3, 5, 6

Trauma is reported as the second most common cause and this has occurred in a variety of ways including: carrying a carpet, a blow from a hockey stick, shoulder dislocations, and even doing push-ups. 5, 6, 7, 14, 19 Other reported causes are penetrating neck injuries, tuberculosis, cervical lymphoma, radiation procedures, internal jugular vein catheterization, hemi-thyroidectomy, neuralgic amyotrophy and vascular compression at the medulla oblongata. 3, 5, 6, 15, 16, 18

The many possible causes of shoulder pain and weakness make a spinal accessory nerve injury easy to overlook and delayed or missed diagnosis is common. 12, 20 Injury due nerve transection as often occurs in radical neck dissection may have the worst prognosis. 13 Partial lesions on electrophysiologic examination such as those cased by blunt trauma, stretch and of spontaneous injury have a better prognosis 8, 19 Electrophysiologic testing is the gold standard for diagnosis but other diagnostic tests have been utilized including ultrasonography and motor evoked potential measurements from magnetic coil stimulation at the base of the skull. 16, 17 If a late diagnosis is made the electromyographer may want to needle the levator scapula and rhomboids to determine their suitability for possible muscle transfer surgery. 13

This patient continued to improve clinically, and physical therapy was discontinued after his second electrophysiologic exam showed continued reinnervation. He was completely pain free and had full active range of motion of the shoulder in all planes at his last exam six months post-injury. If this patient's physical therapy had failed and serial electrophysiologic exams did not reveal reinnervation, surgical intervention may have been warranted. The timing of these surgeries is controversial and success maybe unpredictable but successful nerve repair surgeries have been reported up to 20 months post-injury. 5, 10 12, 13 A surgical intervention for spinal accessory nerve injuries diagnosed even later is the Eden-Lange procedure, a lateral tendon transfer of the levator scapulae and rhomboids, to stabilize the scapula. 6, 10, 13

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