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Differential Diagnosis for a Vietnam Veteran with Complex Neurological Signs and Symptoms: A Case Report

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Abstract

Background: Neck pain has an annual incidence of 10.4 to 21.3% affected in the United States. Clinical practice guidelines developed by the American Physical Therapy Association have classified neck pain into four categories that guide treatment: neck pain with mobility deficits, neck pain with movement coordination impairments, neck pain with headaches, or neck pain with radicular symptoms. **Case Description:** A patient presented to an outpatient physical therapy clinic with severe chronic neck pain, dropped head syndrome, and neurological symptoms including bowel incontinence and split hand sign. His chief complaint was neck pain. The patient transported Agent Orange during the Vietnam War and had comorbidities linked to his military history. **Differential Diagnosis:** Several diagnoses were considered for this patient due to the risk factors and signs and symptoms identified during the evaluation. Agent Orange exposure has been linked to a number of disease states. Dropped head syndrome can be a sign of certain neurological and autoimmune conditions. Epidemiological studies identified a 60% greater risk of ALS in military men during the last 100 years compared to the general population. The purpose of this case report is to describe the clinical decision-making process for the differential diagnosis of a patient with complex neurological signs and symptoms. **Discussion:** Physical therapists are direct access providers in many states, so it is imperative to screen for underlying conditions that would not be appropriately treated with physical therapy alone. This patient is an example of someone who did not realize the potential implications of his additional neurological signs and symptoms. Several physical therapy interventions were used with this patient including manual therapy, dry needling, and movement coordination training.

Keywords: Physical Therapy; rehabilitation; neurology; Vietnam veteran; split hand sign; dropped head syndrome; Agent Orange

Introduction

Neck pain is a common source of disability in the United States with 10.4% to 21.3% of the population experiencing it each year. Office staff and people with computer-based jobs have an increased proportion of neck pain, likely due to the static sitting posture often required of these careers. Additionally, there is a high reoccurrence rate of neck pain, with 50% to 75% of people experiencing it again within 1 to 5 years of the initial onset.[1] Clinical practice guidelines (CPGs) synthesized by the American Physical Therapy Association (APTA) classify interventions recommended for neck pain as well as risk factors based on the strength of evidence. Risk factors for developing neck pain include age greater than 40 years, high physical job demands, limited social support system, concurrent low back pain, cycling for exercise, loss of hand strength, worried or anxious attitude, low quality of life, and poor overall health. Highly predictive risk factors for acute neck pain include female sex and history of prior neck pain.[2]

Prognostic factors for neck pain with moderate and high level evidence include pain rating, self-rated disability level, pain catastrophizing, posttraumatic stress, and cold hyperalgesia. Prior health status, age, and history of musculoskeletal conditions are particularly important to consider as prognostic factors for nonspecific neck pain. Further information about tools that can be used to develop a prognosis is summarized in **Table 1.** The neck pain CPG also stressed the importance of checking for red flags and signs or symptoms that would indicate underlying pathology such as cancer, infection, or arterial insufficiency. Imaging might also be considered at this point based on guidelines that have been established previously such as recommendations from the Canadian C-Spine Rule (CCR), National Emergency X-Radiography Utilization Study (NEXUS), and American College of Radiology (ACR).[2]

Table 1. Tools to Develop Prognosis for Neck Pain[2]

Construct	Recommended Tool and Significant Score
Pain Intensity	Numeric rating scale from 1-10, score of ≥6 is valuable for prognosis
Self-reported disability	Neck Disability Index, >30% is useful for prognosis
Pain catastrophizing	Pain Catastrophizing Scale, score of ≥20 useful for prognosis
Acute posttraumatic stress symptoms	Impact of Events Scale (revised), score of ≥33 is significant for prognosis, scale used in this context to predict symptoms chronicity
Cold hyperalgesia	TSA-II – NeuroSensory Analyzer is the gold standard, but expensive. Alternative tools include cold pressor task for cold endurance or assess cold tolerance with ice cube.

This case report is based on a patient who presented with a chief complaint of chronic idiopathic neck pain for the past 3 years. He presented with a pain level of 7 out of 10 and was unable to significantly change the pain level on his own. The 2017 CPG update recommends categorizing a patient with neck pain into one of four categories: neck pain with mobility deficits, neck pain with movement coordination impairments (including whiplash associated disorder), neck pain with headaches (cervicogenic headache), or neck pain with radicular pain.[2] Based on this classification model, the patient was in the “neck pain with mobility deficits” category. However, this patient had further symptoms of concern that required close screening by a physical therapist and referral to additional health care providers to rule out possible underlying neurological diseases. The purpose of this case report is to describe the clinical decision-making process for the differential diagnosis of a patient with complex neurological signs and symptoms.

Case Description

Patient History

A 74-year-old male was referred to an outpatient orthopedic physical therapy clinic for neck pain by his primary care physician. His neck pain was more severe on the left side and had been constantly present with gradually increasing severity for the last 3 years. The patient reported he woke up one morning in his recliner in 2016 and had intense neck pain and weakness that prevented him from actively extending his neck. He had been sleeping in his recliner for several months prior to this episode due to mild neck pain when he was positioned in supine. However, he was particularly when he experienced the intense symptoms that morning because it was the first time he could not actively extend his neck and never resolved after that. Since then, he had to prop his head up with his fist to obtain a neutral head posture. The patient said he could occasionally actively extend his neck, but this only happened about one time per day early in the morning when he was not fatigued. His main goal in therapy was to reduce his chronic neck pain. He had never received physical therapy services in the past.

The patient served in the Vietnam War and had two deployments to Vietnam in the late 1960s that totaled 24 months. His primary responsibility was loading and transporting Agent Orange on a truck between army bases. Additionally, he experienced a land mine explosion in Vietnam as he was riding in a truck and had several pieces of shrapnel lodged in his gluteal region. Some fragments were surgically removed shortly after the injury, but he reported the surgeon said there were at least four pieces of metal that were too deep to remove safely. The patient said he did not have the rest of the shrapnel removed due to the risks involved with the surgery.

Outpatient Examination and Evaluation

The patient's medical history included a cardiac bypass in 2014, chronic low back pain and bilateral sciatica in the lower extremities for more than 10 years, and type 2 diabetes mellitus. His bilateral lower extremities distal to the knees had dark purple, firm skin due to venous insufficiency for more than 20 years. In 2014, the patient had a left transtibial amputation due to an infection lasting 15 years in the ankle and foot; the infection had been treated by three different large hospital systems with minimal to no improvement. He wore a prosthetic leg on the left and ambulated without an assistive device. He had difficulty standing from a low chair and had to utilize upper extremity support to initiate standing.

Exaggerated thoracic kyphosis was identified at his evaluation. Supine and sitting positions provoked the patient's neck pain, so he was positioned on a reclined plinth with two pillows behind his head for support and comfort during the initial evaluation. His neck pain level was 7/10 during the initial evaluation, which he reported was generally his average pain level each day. Relieving factors included heat and use of a massaging pillow that wrapped around the posterior side of his neck; these modalities only decreased his pain during the application to a 5/10, and the pain returned to the 7/10 baseline when he removed the heat pack or massage pillow from his neck. During range of motion testing, he was unable to lift his chin from his chest, however full passive cervical extension was available when it was assessed by the physical therapist, indicating a strength level of 1/5 on the manual muscle testing scale. A manual evaluation of the neck caused reproduction of reported symptoms in the cervical region and into the left shoulder during left first rib depression (grade II) and during palpation of the left upper trapezius. He had gross hypertonicity of the cervical region bilaterally and in the superior portion of the posterior thoracic region. The patient described that the most debilitating symptom was the constant neck pain as it interfered with his ability to drive, sleep, sit comfortably, and eat.

Neurologic Examination

During the patient's initial evaluation, bilateral thenar atrophy was noted and prompted a full physical therapy neurological exam. The C1-C2 (neck extension), C8 (wrist flexion) and T1 (finger abduction) myotomes were impaired upon manual muscle testing, and dermatomes were grossly unaffected except for some sensory impairment in his calf likely due to his peripheral neuropathy. The

patient noted he had been experiencing bowel and bladder incontinence almost daily for the past three years, to the extent that he decreased his food and drink intake to prevent more severe incontinence. He usually only ate two meals per day because he was nervous about having bowel incontinence while he was completing his daily tasks in the community.

The patient reported he was unable to button shirts and was having difficulty with grip strength for the last two to three years. This would not be expected given his job history, as the patient had retired from factory work about 7 years prior to this evaluation date. He reported that he performed heavy manual work as part of his responsibilities at work every day and had never noticed strength deficits until the last few years. Significant bilateral thenar wasting was present and affected his grip strength extensively as measured by a grip dynamometer. The right hand had 64.3 pounds of grip strength, and the left hand had 56.9 lbs of grip strength. Based on age-adjusted norms, he was in the 25th percentile for grip strength with his right hand compared to healthy controls, and the 15th percentile for grip strength in the left hand, taking into account that was his non-dominant hand; there is a 10 percent expected difference between the dominant and non-dominant hands. The average grip strength for a right-handed male aged 70-74 years is 76 pounds on the left and 81 pounds on the right[6]. He also had decreased pinch grip strength bilaterally with a positive Froment's Sign, and the left hand was more significantly impaired than the right.

The Babinski sign was negative on the right side, although there was no movement of the toes despite the patient stating that it was an uncomfortable sensation. Because he did not demonstrate the signs for a positive test including great toe extension and abduction of his toes, it still could not be considered a positive sign even though there was an abnormal reaction. The Babinski test was performed two times on the right foot with the same result each time—the patient appropriately acknowledged the noxious stimulus but did not have a positive test. It was not possible to test this reflex bilaterally due to the transtibial amputation on the left side. Inverted supinator sign, suprapatellar sign, clonus reflex, and Hoffman's reflex were negative. Reflexes were all hyporeflexive (1+ rating) with biceps, brachioradialis, triceps, and quadriceps tested bilaterally and Achilles tested on the right only due to the amputation. He reported that bilateral sciatica was present at times, but he denied saddle paresthesia.

Clinical Impressions

Although the initial referral was for neck pain, several concerning signs and symptoms appeared during the patient's evaluation. The insidious onset of neck weakness and dropped head syndrome were concerning from a neuromuscular standpoint. Bowel and bladder incontinence can be a sign of conus medullaris syndrome and can require immediate referral to the emergency department if there is a sudden onset. Thenar wasting is another yellow flag that will be discussed later. The history of transporting Agent Orange was noteworthy as that is a toxic chemical linked to many health conditions. It was deemed appropriate to contact the patient's primary care physician to inquire about any other pertinent health history and imaging to gain a more comprehensive clinical picture and assess the need for further testing or consultations.

Communication with Physicians

Further medical history was collected by calling the patient's primary care physician. She said the patient had visited an orthopedic surgeon in 2017 for a consultation about his neck pain. The surgeon ordered an MRI and radiographs of the cervical spine, but the only abnormal finding was degenerative disc disease and forward flexed cervical posturing. The physician informed the patient that he was not a surgical candidate and that there was nothing he could do to help him.

The patient also visited a physician assistant in neurology for a consultation regarding his neck pain about six months prior to presenting in physical therapy (early 2019). The physician assistant was contacted for additional information since the patient's records were not available through the online documentation system used at the outpatient physical therapy clinic. The neurologist had performed an additional cervical MRI at the appointment and said there were no apparent neurological findings from

the imaging or examination with the conclusion that this was a musculoskeletal condition not requiring intervention from a neurologist. Of note, the patient reported having several of his neurological signs and symptoms at this time including the dropped head syndrome, bilateral thenar atrophy, and bowel and bladder incontinence, but the physician assistant said he had not reported the incontinence and the thenar atrophy was not identified during that visit since the referral was for neck pain. It is also interesting that the patient had two cervical MRIs in a year and a half period although he had not experienced many changes in symptoms or chief complaint in that timeframe.

Differential Diagnosis

Risks of Agent Orange Exposure

Agent Orange exposure can have many negative effects on a person's health. Since the patient transported this hazardous chemical during his two deployments to Vietnam, it was important to consider the potential implications of this work history. Agent Orange is a mix of two compounds: butoxyethanol ester or 2.4-dichlorophenoxyacetic acid (2,4-D) and 2,4,5-trichlorophenoxyacetic acid (2,4,5-T). The nicknames of the herbicides used during the Vietnam War come from the color of stripe on 55-gallon drums used to identify each substance. Dioxin, or TCDD, is found in Agent Orange and is a known teratogen that was banned in the United States from domestic use starting in the early 1970s. Higher levels of TCDD were found in Agent Purple, an early form of Agent Orange. The United States and Republic of Vietnam sprayed over 20.2 million gallons of military herbicides between 1961 and 1971. The purpose of these herbicides were to deforest portions of South Vietnam for reduced cover of enemy troops, thus causing bombing targets to be more visible.[7]

Several studies have been performed to determine the long-term impact of Agent Orange exposure for Vietnam veterans. The problem with many of the studies is that they use varying criteria to classify "exposure" to Agent Orange. Some classify "exposure" as having served in Vietnam, and others have a graded exposure classifications depending on which of the four military zones the person was serving in, as the amount of herbicide use varied greatly by geographical location. **Table 2** summarizes diseases with a known link to Agent Orange exposure that qualify for treatment and compensation through the VA Hospital.[7]

Table 2. Medical Conditions Linked to Agent Orange Exposure[7]

Medical Conditions	
AL Amyloidosis	Non-Hodgkin's Lymphoma
Chronic B-cell Leukemia	Parkinson's Disease
Chloracne	Peripheral Neuropathy (early onset)
Diabetes Mellitus Type 2	Porphyria Cutanea Tarda
Hodgkin's Disease	Prostate Cancer
Ischemic Heart Disease	Respiratory Cancers
Multiple Myeloma	Soft Tissue Sarcomas

It was important consider all of the diseases linked to Agent Orange exposure in this case as the patient had prolonged exposure and presents with a broad range of neurological and musculoskeletal signs and symptoms. A systematic review by Stellman et al. asserts: "there is a need for further research in this area because the at-risk veteran population is not at an age at which chronic diseases become manifest, so the time is optimal for conducting such studies, crafting health programs for veterans to better meet their needs, and truly assessing, addressing, and ameliorating health conditions and continuing exposures to lingering traces of Agent Orange in Vietnam."[7] Therefore, it is important to

ask veterans about if they had any chemical exposure during their service years, especially when they are presenting with symptoms that do not fit a normal diagnosis.

Split Hand Sign

The split hand sign is a term used to describe wasting of the thenar eminence and sparing of the hypothenar eminence. Patients' hands will often appear hollow or "scooped out" on the lateral dorsal portion of the hand. There is wasting of the abductor pollicis brevis (APB) and first dorsal interosseous (FDI) but sparing of the abductor digiti minimi (ADM). It was first described in 1996, and the clinical manifestation of this is decreased pincer grip strength and commonly occurs with ALS. Motor wasting of all the intrinsic hand muscles can occur with other neurological diagnoses including spinal muscular atrophy, cervical spondylotic amyotrophy, a component of peripheral neuropathy, spinocerebellar ataxia type 3, and could even occur in elderly individuals without neurological pathology.[8] Figures 1-3 illustrate the significance of thenar wasting in this patient.

Figure 1: Split hand sign in the case study patient

A study by Kuwabara et al. established that the split hand sign is fairly specific for amyotrophic lateral sclerosis (ALS). They studied compound muscle action potential (CMAP) amplitudes of the abductor pollicis brevis, first dorsal interosseous, and abductor digiti minimi. There was a significant reduction in APB/ADM and FDI/ADM ratios for 20% of patients with ALS compared to none of the controls and 0.5% of the neurological disease controls.[8] The split-hand index (SI) was also calculated from these values by multiplying the CMAP amplitude over the APB by the CMAP amplitude over the FDI and dividing the resultant number by the CMAP amplitude from the ADM.[9] A reduction in split-hand index was present in 64% of ALS patients who were categorized as "possible ALS" initially, indicating possible use for it in a clinical context.[9] Additionally, the thenar muscles were found to have a larger cortical representation by Lemon and Griffiths, which could suggest an initial cortical onset of ALS.[10] Therefore, it is important as a clinician to consider ALS as a possible diagnosis when split hand sign is present.

Figure 2: Sparing of hypothenar eminence

Figure 3: Significant thenar atrophy

Dropped Head Syndrome

Patients with dropped head syndrome usually present with neck pain, difficulty eating due to extreme kyphosis of the cervical spine, and impaired horizontal gaze. This forward flexed posture is typically passively correctable and is most prevalent in females, typically appearing in the seventh decade of life. Dropped head syndrome can occur with Parkinson's disease (PD), myasthenia gravis (MG), and autoimmune conditions affecting connective tissue and muscle such as systematic sclerosis or polymyositis.[3] There are not clear treatment guidelines for treating dropped head syndrome since the underlying cause varies and not much research has been completed in this area. The general guideline clinicians follow is using steroids, bracing, and strengthening as an initial intervention, with surgery considered later on if the patient does not respond to the conservative treatments.

A systematic review by Drain et al that included 74 studies totaling 129 patients found that dropped head syndrome is most often related to isolated neck extensor myopathy (31.8%), followed by PD (20.2%), MG (12.4%), and ALS (7.0%). The 28.6% of patients who did not fit into any of these categories were split between 17 individual etiologies.[3] In this review, there was a positive outcome for 73.5% of patients who were treated with primary medical treatment (the typical intervention used for an underlying diagnosis contributing to dropped head syndrome). If primary medical interventions did not work, 87.5% had symptom relief through immune-modulating therapy, and surgery resulted in a positive outcome for 92.9% of patients. It is important to note that proper selection of surgical candidates is critical for the success of surgery to correct dropped head syndrome, and it is still considered a second-line treatment. A surgery was considered beneficial if positive results lasted for at least three years to improve an individual's quality of life. Surgery performed for dropped head syndrome is commonly a fusion from the mid-cervical spine to the upper thoracic spine to allow preservation of some head motion. The fusion crosses the cervicothoracic junction so normal thoracic kyphosis does not contribute to fatiguing of cervical extensors by shifting the head's center of gravity anteriorly. The decreased neck mobility from this procedure could cause an increased fall risk, however, since it would limit the ability to look down at one's feet while walking.[3]

The recommended course of treatment for patients who have dropped head syndrome with underlying PD or MG is to try pharmacologic regimens before surgery. The Drain et al systematic review also recommends that if a patient with this condition reports to a spine surgery clinic, they should first see a neurologist or rheumatologist before surgery is considered in order to rule out possible underlying neuromuscular or autoimmune conditions. If no diagnosis is made in rheumatology or neurology, the patient likely has isolated neck extensor myopathy and would require immune-modulating therapy, typically in the form of corticosteroids. Unfortunately, patients who have dropped head syndrome are the most debilitated of the cervical spine deformity populations based on quality of life ratings, so there can be great benefits through treatment, whether pharmacologic, therapeutic, or surgical. Overall, the previous study demonstrated a positive response to medical and immune therapy in more than 70% of patients. They did not find a significant improvement with bracing or physical therapy although there was a large amount of variation between studies as to what was included in these more conservative treatment methods.[3]

Signs and Symptoms of ALS

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease characterized by painless progressive muscle weakness as the usual first symptom. Other symptoms might include dropping objects, tripping, difficulty grasping a pen, muscle cramping, uncontrollable periods of laughing or crying, and slurred speech. Only the motor neurons are affected, so sensation remains intact. Eventually the diaphragm and accessory breathing muscles are affected, which is fatal unless the person decides to use a ventilator.[11] The prognosis is poor, with most people surviving about 3 to 5 years after the diagnosis is made, and only 4% living more than 10 years.[12]

Because the symptoms can be highly variable, it is difficult to diagnose ALS. Several presentations may occur, including limb-onset, bulbar-onset, primary lateral sclerosis, and progressive muscular atrophy. The hallmark sign to look for with ALS is a combination of upper and lower motor neuron

signs. Limb-onset presents with both upper and lower motor neuron symptoms in the arms and legs, bulbar-onset initially has speech and swallowing impairments at the beginning and limb symptoms later, primary lateral sclerosis only has upper motor neuron symptoms and is less common, and progressive muscular atrophy has only lower motor neuron symptoms. Limb-onset of symptoms occurs in about 70% of people with ALS, bulbar-onset for 25%, and initial trunk or respiratory involvement for the remaining 5%.[13] Upper motor neuron signs include hyperreflexia and spasticity. Lower motor neuron involvement can include muscle wasting, weakness, and fasciculations. Bulbar signs may include dysarthria, dysphagia, and sialorrhea. Occasionally people with ALS present with the pseudobulbar effect which is excessive laughter or crying without a stimulus to warrant that type of emotional reaction.[14]

There are several tests that are usually performed to rule out all other possible diseases that might mimic ALS. This workup typically includes: electrodiagnostic tests (electromyography and nerve conduction velocity), blood and urine studies (high resolution serum protein electrophoresis, thyroid and parathyroid hormone levels, urine collection for 24 hours to identify presence of heavy metals), spinal tap, radiographs, MRI, myelogram of cervical spine, muscle or nerve biopsy, and a thorough neurological exam.[11] There is a great need to develop biomarkers that can definitively identify the presence of this disease and its progression. Research is currently being conducted to identify susceptibility genes, but the results are not significant yet due to a limited sample size.[13] Differential diagnoses clinicians must consider to avoid an incorrect diagnosis of ALS are summarized in **Table 3.**[13] Because ALS has such a poor prognosis, it is especially important to ensure that the diagnosis is correct to protect patients from unwarranted emotional distress.

Table 3. Differential diagnosis when considering ALS[13]

Category	Disorder/Pathology
Disorders of motor neurons	Spinal muscle atrophy, X-linked spinobulbar muscular atrophy, poliomyelitis, post-polio syndrome, hexosaminidase A deficiency
Disorders of motor nerves	Multifocal motor neuropathy, chronic inflammatory demyelinating neuropathy, cramp-fasciculation syndrome, neuromyotonia, hereditary spastic paraparesis plus, hereditary motor neuropathy with pyramidal features, radiculoplexopathy, paraneoplastic syndrome, heavy metal poisoning, mononeuritis multiplex
Disorders of neuromuscular junction	Myasthenia gravis, Lambert-Eaton myasthenic syndrome
Structural CNS and spinal lesions	Syringomyelia, syringobulbia, tabes sorsalis, multiple sclerosis, monomelic spinal muscular atrophy, Lyme disease, human T-lymphotropic virus-1
Myopathy	Inclusion body myositis, polymyositis, dermatomyositis, polyglucosan body disease
Endocrine	Thyrotoxicosis, hyperparathyroidism, subacute combined degeneration, celiac disease

ALS and Military Connection

Although it is not known what causes ALS, there are some clinical and non-clinical risk factors for ALS that researchers have identified. Clinical risk factors related to faster progression of symptoms include bulbar onset, older age at time of diagnosis, and shorter delay in diagnosing the patient.[12]

Beard et al. conducted research with data from the NIH-funded Genes and Environmental Exposures in Veterans with Amyotrophic Lateral Sclerosis (GENEVA) case-control study to develop hazard ratios (HRs) for associations between military-related factors and ALS survival statistics of affected U.S. veterans.[12] Survival was significantly shorter for veterans who served prior to 1950 (HR: 1.74), were deployed during World War II (HR: 1.97), and who mixed or applied burning agents (HR: 1.57). A longer expected survival was associated with exposure to: locally sourced food not provided by the Armed forces (HR: 0.68), burning agents in the field (HR: 0.56), Agent Orange in the field (HR: 0.66), and paints, solvents, or petrochemical substances (HR: 0.73). Of importance, low hazard ratios for exposures to chemicals like Agent Orange in the field or local food might be because those who were affected by these exposures died before enrollment in this study. Factors relating to the patient in this report include age 70-79 (HR: 1.45), with the highest hazard ratio at 1.45 for age >79. He was deployed to Vietnam (HR: 1.02), and the highest hazard ratio was for deployment to World War II (HR: 1.97). He was involved in an IED explosion (HR: 1.27) and had exposure to Agent Orange (HR: 0.66). The hazard ratio for Agent Orange is surprisingly low, but that could be due to the roughly 50 years that have passed since the Vietnam War and the resultant aging of involved veterans.[12]

Occupations that are associated with developing ALS include construction workers, athletics, veterinarians, manufacturing of precision tools, medical work, working in a power plant, and military.[15-17] Traumatic brain injuries (TBIs) that occur early in life might be connected to higher rates of ALS in military populations as many people are young when they join the military and are deployed.[18] The patient in this case report experienced a blast injury while driving past an IED that resulted in embedded shrapnel in his gluteal region, thus it is probable that he suffered from a traumatic brain injury due to the energy released by the IED. He did not report a concussion, but the nature of his job history suggests that it is possible.

Persian Gulf War veterans are at a particularly high risk for ALS compared to veterans who served in other wars. A study by Dr. Horner from the National Institute of Health performed a study on 2.5 million military veterans who actively served during the Gulf War and discovered those who were in the Gulf nearly double the risk of developing ALS compared to military serving in other locations in the same time frame. Additionally, Army and Air Force branches have a higher risk than other branches of the military.[19] Another study about Gulf War veterans conducted by Haley et al. determined ALS occurred at two times the rate in young veterans under age 45 compared to the general population in the same age range.[20] Because of the growing amount of evidence supporting the link between ALS and military service, a Veterans Administration regulation passed in 2008 that makes veterans with ALS eligible for full service-connected benefits. By passing this regulation, the VA assumes the ALS development was provoked by military service and allows any veteran who served at any time period to be eligible for these services.[21] Epidemiological studies performed at Harvard found a 60% greater risk of ALS in men who were in the military in the last 100 years compared to men in the general population throughout that same time period.[4,5] A study by Walt et al. that was recently published by the VA showed a similar rate of chronic traumatic encephalopathy (CTE) and ALS as the rate with other neurodegenerative conditions and CTE. However, the patients who had CTE and ALS were at an increased risk for presenting with bulbar onset, mood changes, and dementia signs connected with the accumulation of tau protein.[22] Due to the many risk factors associated with military service, this diagnosis was considered for the patient in this case study.

Discussion

Interventions

The first two weeks of physical therapy focused on decreasing the patient's high pain rating (rated 7/10) and improving neuromuscular control. He had two 40-minute sessions each week. He had to be positioned in a reclined position because supine increased his symptoms. A plinth was used with the back propped to about a 60-degree angle with two pillows behind his head to facilitate better cervical positioning. It was important to start out with manual and low intensity neuromuscular reeducation since the patient's symptoms were severe and irritable, and to determine which interventions would be best

tolerated. Manual therapy was performed on the posterior cervical region, as this was where most of the pain was located, with the left side more painful than the right. Soft tissue mobilization of the anterior cervical region was also performed as these muscles were functionally shortened due to the patient's near-constant forward flexed posture and exaggerated kyphosis of the thoracic spine. He was able to tolerate moderate pressure during manual therapy, but occasionally a shooting pain would spread towards his shoulder when pressure was applied on the left cervical paraspinal muscles.

Light cervical distraction was also used in the first few treatments to relax the suboccipital muscles that often contribute to headaches and cervical pain. He tolerated the light distraction well, and it did not increase his pain. Between manual interventions, the patient was instructed about how to perform chin tucks to introduce cervical neuromuscular control training. Tactile cueing was utilized, as the physical therapist placed her hands behind his head to facilitate the correct movement pattern. He performed these in the semi-reclined position and was able to produce a very small movement for about 8 repetitions before moderate fatigue occurred. Seated scapular retraction was the other neuromuscular reeducation exercise utilized during the initial treatments, and he did not need cueing to perform this correctly after the initial education and demonstration. Similar to the chin tucks, he was able to complete about 8-10 repetitions before fatigue. The patient subjectively reported that he had a significant reduction in neck pain after these sessions, with an average pain rating of 3/10 for at least 24 hours after an intervention. He stated that he was able to actively hold his head up for about 48 hours after a physical therapy session since his neck was not as painful and fatigued. He reported after the 48 hours post-intervention, his neck pain started increasing and the dropped head positioning happened almost immediately when fatigue occurred. Prior to having some resolution of symptoms from physical therapy, he would prop his head up with his fist under the chin in order to drive and when he had to look up from the forward flexed position; he would also have to use this strategy at approximately 48 hour post-intervention as the fatigue intensified. This was a great outcome as the patient had been unable to facilitate prolonged symptom reduction in the previous three years with heat, ice, massage pillows, and medications and had never tried physical therapy. The response where the patient was able to actively maintain a neutral cervical spine indicated that there was a musculoskeletal component to his neck dysfunction and physical therapy would likely be able to produce a long-term positive effect.

After the positive result from the first two physical therapy sessions, grade II mobilizations of the first rib were introduced because the patient's first rib was elevated and hypomobile. The patient reported an increase in his neck and shoulder symptoms with first rib depression on the left side but only a minor increase in neck pain with first rib depression on the right. However, when pressure was released from the first rib, the increase in symptoms immediately subsided to the initial pain level. This was an important manual treatment to incorporate because some of his pain was likely sourcing from the first rib due to the reproduction of his symptoms. First rib mobilizations (2 to 3 repetitions at a time) were gradually introduced into physical therapy interventions in order for the patient to minimize the spike in pain, and he was able to eventually tolerate this manual intervention for two sets of 6 repetitions of grade II mobilizations. The rib became less hypomobile over a few sessions of treatment. Standing exercise was also trialed with the patient's forearms exerting pressure onto the wall to facilitate serratus anterior activation. His cervical spine was in a maximally flexed posture since he was not in the reclined position anymore to facilitate improved posture, so the physical therapist helped the patient achieve active assisted neutral head positioning. He became fatigued after only a couple seconds of neutral head positioning and was unable to maintain this position despite the external assistance. This exercise was discontinued at the time as it was too challenging for the patient and was introduced a week later when the patient had improved neck extensor control.

Because the patient had extensive cervical pain and was often forward flexed, collar orthoses were considered to help the patient passively maintain neutral cervical posture. The Headmaster Collar, which is commonly used with patients who have cervical weakness, was issued and fit to the patient because it is flexible and can be adjusted to fit the patient's needs. It has an open front on the collar, as many patients do not like having the anterior portion of the neck covered by collars. He used the

Headmaster most of the time, but he also had a regular soft collar that he utilized the rest of the time. This helped reduce his neck pain and abnormal posture that exacerbated his symptoms.

Interventions utilized in sessions after the first two weeks of physical therapy included neuromuscular reeducation of various muscle groups including scapular stabilizers, cervical extensors, and deep neck flexors. Scapular stabilization exercises included standing and seated rows, shoulder horizontal abduction with an elastic band, bilateral shoulder external rotation with an elastic band, and scapular protraction against the wall to increase serratus anterior activation. A study by Moezy et al. researched the effect of scapular stabilization training on pain and posture for patients with shoulder pain and impingement. They found that there was a significant reduction in forward head and shoulder posture, increase in pectoralis minor extensibility, and an increase in active range of motion for shoulder abduction and external rotation.[23] Although this patient did not present with strictly shoulder impingement syndrome characteristics, it was still beneficial to address the weakness for postural improvements and reduced impingement that could be exacerbating his symptoms.

Training of the deep neck flexors and cervical extensors were an aspect of his rehabilitation as well. Neck pain can reduce the activation of deep neck flexors and increase muscle fatigue and co-contraction of neck muscles, all further exacerbating the neck pain and postural deficits. People with neck pain usually present with increased sternocleidomastoid and anterior scalene contraction and decreased activity of the deep neck flexors.[24] Deep neck flexor training utilizes low load holds typically in a supine position to increase endurance of these postural muscles. A recent systematic review by Blomgren et al. found that training of the deep neck flexors can increase neuromuscular coordination but does not have a direct correlation with cervical flexor strength or endurance at higher intensities. To address endurance at a higher intensity, multimodal treatment is required instead of just deep neck flexor training alone.[24]

Functional dry needling was used with this patient to reduce his neck pain. It has been performed in his shoulder musculature as well as cervical paraspinals. Dry needling uses a small needle that can be pushed into a myofascial trigger point in the muscle to facilitate decreased pain, increased function, and reduced hypertonia. These trigger points are irritable, tight adhesions that feel like “knots” in a muscle which can cause pain and radiating symptoms. Poor posture is often accompanied by myofascial trigger points. A localized twitch response may happen with trigger point dry needling that disrupts activity at the motor end-plate, facilitating a reduction in pain. Researchers believe that the gate control theory of pain might be involved in this pain reduction with dry needling, and research has shown that there might be greater pain relief when a twitch response is elicited.[25] There is currently a limited number of quality randomized controlled trials published, so this is an important area for further research about functional dry needling.

LSVT BIG training was also incorporated in this patient’s physical therapy sessions due to the presence of movement impairments during ambulation, decreased neuromuscular coordination, and difficulty standing from a standard height chair. By training the patient to use large amplitude, exaggerated movements that were specifically selected for his impairments, he was able to start recognizing the motor mismatch between how he was actually moving compared to how his body perceived his movement.[26] This treatment approach is typically used for patients with Parkinson’s disease, but it was included in his therapy sessions to see if it would improve movement coordination, and because his neurological condition had not yet been diagnosed.

A Physical Therapist’s Role in Differential Diagnosis

Physical therapists have a unique perspective in the healthcare environment. With extensive knowledge in movement analysis, musculoskeletal conditions, and neuromuscular conditions, physical therapists are trained to take the “whole picture” into account when developing a treatment plan. This includes thorough screening through interviewing and testing to identify possible yellow or red flags that would warrant referral to another healthcare provider. Many states allow patients to have direct access to physical therapists without a referral from their physician, which makes it even more important for physical therapists to take all the patient’s symptoms into account and ask appropriate screening

questions. An interesting aspect of this case is that if the patient was just treated for neck pain, many components affecting his health would have been missed. He needed a full neurological examination by the physical therapist to identify several red and yellow flag findings that would have otherwise gone unnoticed. Although this was in an outpatient orthopedic physical therapy clinic, it was still critical to have knowledge of various neurological testing techniques, including tests for myotomes, dermatomes, reflexes, and other special tests. This can impact the patient's speed to proper diagnosis and impact quality of life and outcomes.

As the patient shared more of his personal history during the second appointment in physical therapy (such as mentioning the bowel and bladder incontinence), it was clear that he needed more testing from his neurologist. The physician assistant (PA) who saw the patient for a neurology neck pain consultation was contacted by phone to discuss findings in physical therapy. The physical therapist shared that several yellow and red flags were identified including bowel and bladder incontinence, bilateral thenar atrophy, and insidious onset of neck pain and weakness. The PA reported that these symptoms were not reported at his previous neurology appointment and the thenar atrophy was not identified since the referral was for neck pain. The PT suggested testing to rule out diagnoses such as ALS due to the patient's history of 24 months in Vietnam, thenar atrophy, and weakness starting distally in the limbs. The PA agreed and said she wanted to see the patient again to perform further testing based on these clinical findings and would consult her colleagues about this case.

The patient was contacted by neurology to set up an appointment at their clinic. Upon evaluation, the patient was diagnosed with amyotrophic lateral sclerosis, and the neurologist recommended a second opinion to confirm this diagnosis. The patient was referred to a large hospital system for a consultation, and his case was not accepted there. At the last follow up with this patient, he was waiting for an appointment at a different hospital system with a neuromuscular neurologist who specializes in ALS and other neurodegenerative diseases to confirm his diagnosis and prognosis.

Through the patient's physical therapy interventions, he reported that he did not have any neck pain (0/10 on a pain rating scale), and this was consistently reported for the past month. He continued to have movement impairments and decreased activity tolerance, but the reduction in neck pain from a 7/10 to 0/10 was a significant outcome. Although the diagnosis of ALS had not been confirmed through a second opinion of another neurologist at the time this case report was presented, he continued to experience ongoing improvements in pain level and increased function during 5 months of physical therapy. His NDI score started at 23 points on 7/3/19 and he had a score of 16 on the NDI on 9/16/19, indicating a significant increase in function after two months of physical therapy interventions. Physical therapists play an integral role in recognizing impairments that lead to earlier diagnosis and treatment of many simple and complex health conditions. Additionally, if other healthcare professionals should be included in the patient's care, physical therapy is highly valuable to provide unique interventions that may improve a patient's function and quality of life.

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