Case in Point

Parsonage-Turner Syndrome: Diagnosis and Rehabilitation Strategies

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Knowing that the diagnosis of Parsonage-Turner syndrome is one of exclusion, the author presents the signs, symptoms, and rehabilitation therapies that helped a patient return to a rewarding way of life.

Patients with acute and nonacute cervical and shoulder pain often seek assistance from medical professionals for pain relief. In most cases, these patients can be easily diagnosed and treated. However, in some cases, no definitive common diagnosis can be determined. This inability to readily identify the etiology or provide adequate treatment results in frustration for the patient and the medical professional. Parsonage-Turner syndrome (PTS) is an infrequently seen diagnosis that mimics similar characteristics of other common cervical and shoulder pathologies.

Parsonage-Turner syndrome is a rare disease of unknown etiology that mainly affects the lower motor neurons of the upper trunk of the brachial plexus. The brachial plexus is a group of nerves that conduct signals from the dorsal roots of the spinal column to the peripheral nerves of the shoulder, arm, and hand. Parsonage-Turner syndrome is characterized by the sudden onset of excruciating unilateral or bilateral shoulder pain followed by flaccid paralysis of the shoulder and lack of motor control within the upper extremities several days later.[1-5] Although a rare disease, PTS has been referred to in the literature as acute brachial neuritis, brachial plexus neuropathy, neuralgic amyotrophy, neuritis of the shoulder girdle, idiopathic brachial plexopathy, and shoulder girdle syndrome.[2,3,6,7]

Parsonage-Turner syndrome is reported to occur in 1.64 cases per 1,000,000 annually.[7-9] Men have a higher incidence rate, between 2:1 and 4:1.[2,6,10] There is no specific age group that PTS affects; however, it is most likely seen between the third and seventh decades (aged 30-70 years).[3,6,11,12]

As previously reported, the demographics are unique for PTS, which can also be said for the pathophysiology and etiology of the disease. Parsonage-Turner syndrome exists in 2 forms: idiopathic and hereditary. In the idiopathic form, the cause of the disease remains unknown, but it can have ties with an inflammatory response against nerve fibers of the brachial plexus.[1 6,13] In the hereditary form, PTS is autosomal dominant characterized by recurrent attacks of pain in a brachial plexus distribution.[1,6,14] Although the cause of the disease remains unknown, PTS has been linked to prior events or diseases, such as trauma, heavy exercise, surgery, childbirth, lumbar puncture, systemic diseases, parasitic infestation, vaccinations, and radiation therapy. In the hereditary form of PTS, a mutation on the SEPT9 gene on chromosome 17 has been identified.[1,5,8,11,15]

Clinical presentation of PTS can mislead medical professionals in accurately diagnosing and medically managing these patients. As mentioned earlier, the symptoms of PTS can mimic various cervical and upper-extremity diagnoses. Patients with PTS will typically present with acute onset of severe pain that progressively worsens over 2 days with no apparent cause. Patients usually describe the pain as a constant severe ache associated with tenderness of the affected muscles. The patient's pain is usually not affected by coughing or cervical range of motion (ROM). However, the patient's symptoms increase with upper extremity movement and pressure applied to the affected muscles. It is not uncommon for the patient to experience pain on the numeric scale of 9 to 10 within the shoulder girdle.[3,12] Although the disease commonly affects the right upper extremity, 30% of the cases involve bilateral upper extremities.[3,6] Once the acute stage of the disease is over, the pain is followed

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Table 1. Patient's physical therapy assessment

	Time		
	Initial	Discharge	Change
Pain	7/10	4/10	N/A
UE sensation	Right: diminished from mid-humeral area distally; left: median nerve distribution numbness	No change	N/A
UE deep-tendon reflexes	Right: 2 + triceps, bicep, 0 brachioradialis; left: 2 + triceps, bicep, brachioradialis	No change	N/A
Shoulder strength	3 +/5 bilaterally in all planes	Right: 4/5 all planes; left: 4/5 all planes	+ 1 + 2
Biceps	Right: 1/5; left 3 +/5	Right: 3 +/5, left 4/5	+ 6, + 8
Triceps	Right: 2/5; left 3/5	Right 3 +/5; left 4/5	+ 2, + 4
LE	Within functional limits bilaterally	No change	No change
Shoulder flexion	Right: 0-90	0-125	+ 35
Shoulder abduction	Right: 0-90	0-90	0
Shoulder internal rotation	Right: 0-40	0-80	+ 40
Shoulder external rotation	Right: 0-20	0-70	+ 40
Elbow	Right: 0-140	0-140	0

LE = lower extremity; UE = upper extremity.

by rapid muscle atrophy, flaccidity, sensory impairments, and decreased deep-tendon reflexes within the affected arm.[10-12,16] In addition, it is very common for the patient to experience discrepancies within the muscles that have atrophied and have denervation between muscles by the same nerve.[7,10]

Since the clinical presentation mimics various other pathologies, PTS is difficult for medical professionals to diagnose. The diagnosis of PTS is achieved only through exclusion.[10,11] Some of the similar diagnoses include stroke; brachial plexus trauma; cervical radiculopathy; shoulder pathology (eg, rotator cuff tears, tendonitis, subacromial bursitis, and impingement syndrome); osteoarthritis of the neck and shoulder; somatic dysfunction; myofascial pain; thoracic outlet syndrome; and Paget-Schroetter disease. Therefore, for the medical professional it is critical that these diagnoses are differentiated by taking the patient's medical history and performing a physical examination and diagnostic tests.[5,17]

In addition to ruling out these pathologies, the medical professional must use diagnostic studies to assist in the confirmation of the disease. Since patients often report of an upper extremity weakness and paresis, an electromyography (EMG) study is an accurate and useful diagnostic test to assist in diagnosing PTS. The limitation of an EMG is that it may take 2 to 3 weeks before a decrease in nerve conduction velocity in the patient's affected nerves is revealed.[8,11] Other diagnostic tests also include magnetic resonance imaging (MRI) and a computer tomography scan to rule out cervical radiculopathies and blood tests for any potential broader autoimmune diseases.[7,11,12]

One of the most important clinical neurologic skills is the ability to localize the motor or sensory lesion to a particular nerve, nerve root, trunk, or branch within the brachial plexus, because this will assist with the differential diagnosis. Once the medical professional determines that the brachial plexus is affected in multiple areas, "patchwork localization" is confirmed, resulting in differential diagnosis restrictions.

Because the diagnosis of PTS is made through exclusion, the medical professional must look for 3 distinct signs of the disease. These signs and the accompanying patient symptoms will allow the medical professional

to make an efficient and cost-effective diagnosis of PTS. The first sign is that the patient's muscles will tend to have a discrepancy for muscle atrophy and denervation between the muscles that are innervated by the same nerve. Second, there will be patchwork distribution of muscles that are denervated and innervated by several nerves or a nerve trunk arising from the brachial plexus. Third, there will be dissociation between sparing of the sensory nerve action potentials and muscles denervation, depending from the same mixed nerve.[7]

Treating PTS can be challenging. To date, there are no specific treatments that have been known to be the most effective. For this reason, a curative treatment for PTS may be difficult to achieve. Palliative and supportive care for pain control, ROM, and strengthening activities are critical components toward the patient's long-term recovery.[1,7]

During the acute phase of the disease, pain management is critical. Patients will often be prescribed long-acting nonsteroidal antiinflammatory drugs and opiates to help with pain control.[1,7] In addition to medication therapy, patients may be referred for physical therapy (PT) and occupational therapy (OT). During PT, the therapist focuses on patient education, upper-extremity ROM, strengthening exercises to maintain periscapular motion, and modalities for adjunct pain relief.[1,7,10] An occupational therapist is consulted to treat and assist the patient with functional conditioning of the affected upper extremity, use of assistive devices, such as splints, education of activities of daily living (ADL) equipment, and ROM and strengthening activities if the hand or wrist is involved. In addition to assisting the patient with sensory motor deficits, OT may play an imperative role in assisting the patient with the psychological aspects of disability and lifestyle adjustment.[1]

Once the patient's pain is controlled, rehabilitation services should begin immediately. Although the patient's prognosis is variable, the severity and duration of the pain will tend to have a direct correlation to the patient's overall recovery.[11] Symptoms seem to quickly stabilize. A gradual recovery period follows over 6 months to 5 years. On average, 75% of all patients will have their deficits completely resolved within 2 years. The other 25% of patients will experience permanent upper-extremity deficits.[11,12]

CASE STUDY

The patient is an incarcerated 63-year-old male under federal custody. Overall, the patient's past medical history is unremarkable. In December 2008, the patient injured his right thumb by scraping it against an outside door frame, which resulted in a superficial wound. The minor wound required cleansing, and the patient was prescribed an antibiotic as a precautionary antiinfective measure. Eight days following the incident, the patient reported symptoms of a rash developing throughout his body, multijoint pain and stiffness within the bilateral shoulders, right wrists and right fingers, and increased edema within the right hand and fingers. The patient also exhibited sensory and deep-tendon reflex deficits. These deficits were greater on the right side than on the left.

The patient underwent diagnostic tests, including a complete panel of blood work, cervical MRI, X-ray of the shoulder and right hand, and an EMG study. The blood work revealed a negative rheumatoid factor, the cervical MRI was unremarkable, and X-rays revealed negative findings within the shoulder but severe periarticular deossification of the right wrist. The EMG test results provided the greatest information. The needle examination of the right upper extremity showed evidence of fairly diffuse acute and chronic reinnervation affecting most muscles in the arm, forearm, and hands. The needle examination of the right deltoid and supraspinatus muscles was normal. Needle examination of the right C6-C7 cervical paraspinal muscles was normal. Needle examination of the left upper extremity showed patchy involvement of the extensor digitorum communis, triceps, and infraspinatus muscles with evidence of acute and chronic reinnervation. The patient's right upper-extremity nerve-conduction velocity test revealed median nerve sensory responses were absent, ulnar nerve sensory responses were reduced, radial nerve sensory responses were absent, and the musculocutaneous nerve sensory response was absent. The patient's left upper-extremity nerve-conduction velocity test revealed the median nerve sensory response was reduced with prolonged latency, ulnar nerve sensory response was normal, radial nerve sensory response was absent, and the musculocutaneous nerve sensory response was absent. The nerve conduction motor test for the right upper extremity revealed that the median nerve had normal conduction latencies and reduced motor response. In the left upper extremity, the median nerve was normal, and the ulnar nerve was normal with slow conduction above the elbow.

The analysis of the diagnostic studies, especially the abnormal nerve conduction and EMG tests that revealed patchy involvement within the left upper extremity, resulted in the diagnosis of PTS. The patient continued with his regimen of medica-

Table 2. Patient's occupational therapy assessment

	Time		
	Initial	Discharge	Change
Right wrist			
Flexion	0-30	0-30	0
Extension	0	0-35	+ 35
Supination	0-45	0-45	0
Pronation	0-60	0-75	+ 15
Right index finger			
MCP	35-50	5-65	+ 45
PIP	35-65	10-45	+ 5
DIP	10-35	5-40	+ 10
Right middle finger			
MCP	35-50	0-40	+ 25
PIP	40-65	10-55	+ 30
DIP	10-50	5-60	+ 15
Right ring finger			
MCP	35-50	0-40	+ 25
PIP	40-65	10-55	+ 30
DIP	0-50	5-60	+ 5
Right little finger			
MCP	45-65	10-55	+ 25
PIP	35-70	10-35	– 10
DIP	5-60	5-55	– 5

DIP = distal interphalangeal joint; MCP = metacarpophalangeal joint; PIP = proximal interphalangeal joint.

tion for the pain reduction and was referred to PT and OT for initial evaluation and treatment due to the unresolved initial symptoms and deficits.

In the federal prison environment, the management of prescription pain medications can be very challenging for medical professionals. The patient's prescription list consisted of 800 mg of ibuprofen every 8 hours. The patient was also prescribed 25 mg of topiramate, used off label, for his neuropathic pain. In the 6 months following the initial injury, the patient was referred to PT and OT. The rehabilitation team consisted of a physical therapist who is a commissioned officer within the U.S. Public Health Service and a civilian contract occupational therapist who is employed by the Federal Bureau of Prisons.

During the initial PT evaluation, the patient indicated that he continued to have a significant amount of pain with a significant decrease in overall function. He reported he led a sedentary lifestyle with no participation in any type of exercise or work program due to his medical condition. Vital signs were stable, and the patient reported his bilateral upper-extremity pain to be a constant 7 out of 10 on the numeric pain scale.

The physical examination showed decreased bilateral shoulder strength, decreased bilateral bicep strength, decreased bilateral triceps strength, and normal strength within the lower extremities bilaterally (Table 1). Decreased bilateral shoulder active ROM deficits were found in all planes. The patient could generate slightly less active ROM of the left elbow compared with the right, but both were within functional limits. Sensation testing showed a diminished sensation within the right upper extremity from the mid-humeral area distally to the tips of all fingers; left upper-extremity testing revealed numbness within the median nerve distribution.

The initial OT evaluation findings were similar to the physical therapist's findings. The patient continued to report increased pain and decreased bilateral upper-extremity function. The OT evaluation revealed numbness on the anterior and posterior surfaces of the right upper extremity from the mid-humeral area radiating distally. The patient's greatest ROM deficit was seen within the right fingers (Table 2). Abduction and adduction of the patient's right fingers were weak, but he was able to oppose to the index. On visual examination and touch of the patient's right hand, the skin showed pallor but felt normal to touch. Minimal right-hand edema was present at the initial evaluation. Coban wrapping and a compression glove were used to assist in decreasing the edema. These techniques were found to be more beneficial than hand massage

and elevation. The patient's edema within his hand subsided within normal limits, and there was no evidence of edema at discharge.

The patient received rehabilitation services for 9 months. The PT and OT treatments were guided by the protocols described earlier. The PT treatment consisted of education, pain management, active and passive ROM of the shoulder and other affected joints in all planes, and strengthening exercises. Strengthening exercises with cuff weights, progressing to dumbbells; reaching and grasping activities; and active and passive ROM stretches through all planes, performed supine and standing, resulted in the most significant changes. The OT provided similar treatment strategies with treatment focused on regaining fine motor control and functional activities. Treatments consisted of right hand and finger edema control via a coban wrap, progressing to a compression glove and active and passive ROM of the right hand. In addition, the patient received education on the use of adaptive equipment and sensory awareness during ADL.

After 9 months, the patient was discharged from rehabilitation services. At that time the patient's active ROM in all affected joints had improved compared with the initial evaluation. The patient showed improvement within his upper-extremity strength and overall ADL. At discharge, the patient was able to use both upper extremities to assist himself in dressing and feeding. He was also able to return to work as a dishwasher and participate in recreational activities such as gardening. Being able to return to these activities was very rewarding for the patient.

CONCLUSION

Various common pathologies, such as shoulder pathology, cervical radiculopathy, and myofascial pain, must be differentiated when diagnosing PTS. Medical professionals must be aware of the similar adverse effects of PTS when patients seek medical attention due to their concerns and presentation, as well as the poor correlation between musculocutaneous findings and diagnostic imaging results. ●

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