

CLINICAL VIGNETTE

Parsonage Turner Syndrome in a Geriatric Patient

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Case Report

A 72-year-old female with osteoarthritis and a history of staphylococcus capitis bacteremia (on indefinite dicloxacillin prophylaxis) presented to the hospital with sudden onset left shoulder and upper extremity pain and weakness after breaking down cardboard boxes. The pain radiated from her left shoulder to her elbow and was associated with paresthesias along her forearm and lateral hand. She had a similar episode two weeks prior that was suspected to be musculoskeletal related and had self-resolved. This time, however, her pain and weakness were severe and prompted her to visit her local emergency room. She was seen by neurology and MRI / MRA of her brain and neck was negative for acute stroke. MRI showed cervical spinal stenosis most severe at C4-5 with cervical spondylolisthesis and left-sided C4-5 neuroforaminal narrowing. MRI shoulder also revealed severe glenohumeral osteoarthritis, joint effusion, as well as total thickness rotator cuff tear, with complete tear of the long head biceps tendon. Orthopedics recommended outpatient follow-up and physical and occupational therapies. Neurology suspected possible Parsonage-Turner syndrome. She was initially started on high-dose steroids in the hospital, which were discontinued due to intolerance and intermittent fevers. Patient was seen by infectious disease and extensive testing was negative except for an incidental urinary tract infection.

She was discharged to a skilled nursing facility (SNF) for rehabilitation, with a prolonged stay. At the SNF, she continued to have severe pain of her left shoulder and arm that lasted many weeks. She was treated with several short courses of low dose steroids, opiates, and topical analgesics. She required a shoulder immobilizer to help with pain and weakness. As her pain subsided over a few weeks, she developed complete flaccid paralysis of her left upper extremity. Neurology performed outpatient EEG / NCS which showed active denervation with diffuse sensory and motor axon loss of the left upper extremity nerves, and early chronic reinnervation in all upper arm and distal muscles with relative sparing the deltoid, and periscapular muscles. The pattern and clinical presentation were consistent with an acute brachial plexopathy (Parsonage-Turner syndrome). By SNF discharge, her left shoulder pain had improved but she had ongoing severe weakness and paralysis of her arm and forearm. Left-hand grip strength improved to 2/5, but she continued to require use of a sling and nighttime splint to prevent contractures of her elbow, wrist and fingers. At an orthopedic-spine follow-up five months after initial presenta-

tion, she had supple neck range of motion and variable muscle strength.

Discussion

Parsonage-Turner syndrome (PTS) is a serious, underdiagnosed, disabling peripheral nervous system disorder. There is sudden onset, severe pain involving the shoulder girdle and upper extremity typically lasting several weeks, followed by progressive muscle weakness and paresis of the upper limb that usually lasts several months.¹ The recovery period can be variable. PTS can be difficult to diagnose because symptoms mimic other conditions. It has frequently been misdiagnosed as cervical radiculopathy or cervical spondylosis.¹ Other common misdiagnoses that mimic PTS include rotator cuff and shoulder capsule disorders such as rotator cuff tendinopathy, adhesive capsulitis, and bicipital tenosynovitis.² Accurate early identification of the syndrome is important to ensure prompt intervention to reduce the severity of the clinical course.

Parsonage-Turner syndrome is named after the two neurologists who described the condition in 1948.¹ They termed the disorder as “brachial neuralgic amyotrophy” to highlight the neuropathic pain and muscle atrophy that is seen.² The syndrome is also commonly known as brachial plexus neuritis or acute brachial plexus neuropathy. The pattern of nerve involvement can be highly variable but typically affects the upper and middle trunks of the brachial plexus, with a tendency to affect the supra-scapular and long thoracic nerves.^{1,2} Either a single or multiple nerve palsy can occur. The shoulder and the scapular regions are the common sites of pain with one upper limb typically affected.² The incidence of Parsonage-Turner syndrome in the general population was previously reported to be rare at 2-3 cases per 100,000 per year.¹ However, it is now believed the incidence is higher since PTS is commonly misdiagnosed or underdiagnosed. A retrospective study by Milner et al 2016 reported an incidence of 32 cases per 100,000 referred patients. However, the authors speculated that their higher incidence may be due to selection bias with their hand surgical practice with a large patient referral base. A prospective primary care cohort study in the Netherlands reported incidence of 100 cases per 100,000 persons per year (or 1 in 1,000). This involved primary care physicians trained to identify Parsonage-Turner syndrome.^{2,3}

Etiology of the Parsonage-Turner syndrome can be either idiopathic or hereditary. The hereditary form is also called hereditary brachial plexus neuropathy (HBPN). Attacks are difficult to distinguish from the idiopathic form.² For the hereditary form, dominant mutations of SEPT9 are the only known current mutations associated with PTS.¹ The idiopathic form is believed to have some sort of inflammatory or immune-mediated trigger that targets specific components of the peripheral nervous system, increasing the risk of developing PTS.^{1,2} These triggering events include a preceding infection, immunization, trauma, surgery, and childbirth, with infection being the most common triggering event.^{2,4}

The diagnosis of Parsonage-Turner syndrome involves conducting a thorough history, examination, and testing. Evaluation should include assessing for risk factors such as a recent preceding infection, immunization, certain medications, trauma, recent surgery, and childbirth / postpartum status. Viral infections that have been associated with Parsonage-Turner syndrome include Epstein-Barr, Varicella Zoster, Dengue, Hepatitis E, Influenza A, and Coronavirus 2019.^{1,2,5} Antecedent immunizations that have been associated with PTS include tetanus toxoid, influenza,¹ and COVID-19 vaccinations.⁴ Potential culprit medications include antiepileptics, antibiotics, immunosuppressants such as nivolumab,⁴ antivirals and botulinum toxin.^{1,4} Some patients have reported prior sudden physical exertion and trauma. The trauma can be minor such as a fall without any visible injury.⁴ The latency period between the inciting event and the onset of PTS is typically four to six weeks.⁴

PTS presents clinically as sudden onset, severe pain (sharp, stabbing, constant) of the shoulder and arm. Typically, a unilateral upper limb is affected^{2,5} but, in one third of cases, both upper limbs may be affected.² The forearm and hand may also be involved. The severe pain is debilitating and usually lasts for several weeks, eventually turning into a dull ache.² Significant weakness to complete flaccid paralysis of the affected limb subsequently occurs.² Some patients may experience acute onset dyspnea and orthopnea if there is phrenic nerve and diaphragmatic involvement. A painful winged scapula may be seen in 30 to 70% of attacks due to weakness of serratus anterior or trapezius muscles from long thoracic and spinal accessory neuropathies.² In the hereditary form of PTS, age of onset is typically around 20 years whereas, in idiopathic PTS, the average age of onset is 40 years. There is 2.5:1 male predominance in both idiopathic and hereditary PTS.² There are some clues on history and exam that can help distinguish cervical radiculopathy from PTS. Although patients with PTS and cervical radiculopathy may both have pain in the cervical spine, shoulder and upper extremity, the pain in cervical radiculopathy is usually insidious in onset while the pain in PTS is rapid in onset.⁶ Furthermore, symptoms of cervical radiculopathy tend to be exacerbated with neck movements, whereas symptoms related to PTS should not be exacerbated with neck movements.⁵

Magnetic resonance imaging (MRI), electrodiagnostic tests, and neuromuscular ultrasound are useful to distinguish Parsonage-Turner syndrome from other disorders. MRI can rule out other potential etiologies of upper limb weakness such as shoulder pathology or extrinsic nerve compression.¹ MRI findings such as multifocal T2 nerve and muscle hyperintensities with nerve hourglass-like constrictions, pre-and post-lesion dilations, and bullseye changes are supportive of a PTS diagnosis,^{2,4} as well as findings of edema and atrophy of affected shoulder-girdle muscle.¹ More than 50% of PTS patients will also have abnormalities on cervical spine MRI without correlation with clinical presentation of PTS.² Nerve conduction study (NCS) and needle electromyography (EMG) are useful in pinpointing the affected nerves,^{1,2} as well as improving the diagnostic accuracy of PTS.² EMG findings include fibrillation potentials and positive waves.^{5,6} Ultrasound can determine if there is phrenic nerve (and, hence, diaphragmatic) involvement.² Nerve biopsies are typically not performed for diagnosis of PTS. However, prior nerve biopsies have shown severe axonal degeneration associated with lymphocytic inflammatory infiltrates,^{2,4} as well as presence of infiltrates in microvessels.²

Management of Parsonage-Turner syndrome is typically conservative. Analgesics like NSAIDs, opiates, and oral corticosteroids can help reduce pain. Gabapentin, pregabalin, amitriptyline, and carbamazepine can be useful for neuropathic pain.¹ High dose steroids within one month of symptom onset may be associated with earlier pain resolution and faster recovery in some patients.^{1,7} Immunomodulators may be considered but typically are not used due to high cost and insurance reimbursement issues.¹ In patients with the hereditary form of PTS, recurrent attacks of pain and muscular weakness can be induced by surgery or childbirth. They may be treated prophylactically with steroids or intravenous immunoglobulin.²

Physical and occupational therapies are important for functional recovery in PTS. They include range of motion and strengthening exercises of the rotator cuff and shoulder girdle musculature.^{1,2} Strategies to restore scapular function and prevent contractures are also important. Immobilizers / splints for the shoulder and/or arm can help prevent contractures, and TENS (transcutaneous electrical nerve stimulation) units can be used to keep motor end plates viable while awaiting recovery from muscular paralysis.¹ Therapy should be continued until the patient has either fully recovered or reached a plateau in improvement. In PTS patients without recovery, there are a few case reports of surgical exploration of the affected peripheral nerves, and neurolysis or resection with interposition grafting with possible benefit.¹ However, this is controversial and needs further study.²

Prognosis of Parsonage-Turner is variable. Many patients will have good functional recovery.² However, some patients have lifetime residual pain and weakness without complete motor recovery.¹ Upper trunk plexus injuries (shoulder and arm) are associated with near complete functional recovery by one year in most patients.² However, middle and lower trunk plexus

involvement (forearm and hand) are associated with a slower recovery and make take up to 1.5 to 3 years longer.² If the etiology is hereditary, recurrence is more common.² Milner et al's nine year retrospective study reported 65% of patients regained muscle strength grade 4 or higher at a mean period of 10 months after diagnosis.

Conclusion

Parsonage-Turner syndrome (brachial plexus neuritis) is an underrecognized disorder characterized by sudden onset, severe pain of the shoulder and upper limb followed by progressive muscle weakness and paralysis of the affected muscles. The pain typically lasts weeks and the weakness / paresis typically lasts months. Most cases are idiopathic, although an antecedent trigger may be identified, and the exact etiology is often unclear. In our patient, her left shoulder and upper extremity pain lasted a few months but eventually subsided. It was followed by persistent, left upper extremity weakness and flaccid paralysis. She was seen by specialists including neurologist, neurosurgeon, orthopedist, and physiatrist for her pain and monoparesis. Despite her known diagnosis of Parsonage-Turner syndrome, this patient also had other contributing factors to her left shoulder pain and upper extremity weakness: cervical myelopathy and severe rotator cuff arthropathy. The diagnosis of Parsonage-Turner syndrome might have been overlooked if it were not for her EMG/NCS results. Her recovery from Parsonage-Turner syndrome is anticipated to take longer due to involvement of all the trunks of the brachial plexus (upper, middle and lower) as manifested by her shoulder, arm, forearm, and hand paralysis. Her neurologist prognosticated that functional recovery may take about one year. Patients presenting with sudden onset upper extremity pain and associated weakness / paralysis, should include Parsonage-Turner Syndrome in the differential diagnosis, and consider EMG / NCS for diagnostic accuracy.

REFERENCES

1. **Milner CS, Kannan K, Iyer VG, Thirkannad SM.** Parsonage-Turner Syndrome: Clinical and Epidemiological Features From a Hand Surgeon's Perspective. *Hand (N Y)*. 2016 Jun;11(2):227-31. doi: 10.1177/1558944715627246. Epub 2016 Mar 8. PMID: 27390568; PMCID: PMC4920531.
2. **Meiling JB, Boon AJ, Niu Z, Howe BM, Hoskote SS, Spinner RJ, Klein CJ.** Parsonage-Turner Syndrome and Hereditary Brachial Plexus Neuropathy. *Mayo Clin Proc*. 2024 Jan;99(1):124-140. doi: 10.1016/j.mayocp.2023.06.011. PMID: 38176820.
3. **van Alfen N, van Eijk JJ, Ennik T, Flynn SO, Nobacht IE, Groothuis JT, Pillen S, van de Laar FA.** Incidence of neuralgic amyotrophy (Parsonage Turner syndrome) in a primary care setting--a prospective cohort study. *PLoS One*. 2015 May 27;10(5):e0128361. doi: 10.1371/journal.pone.0128361. PMID: 26016482; PMCID: PMC4445915.
4. **Rosca EC, Al-Qiami A, Cornea A, Simu M.** Parsonage-Turner Syndrome following COVID-19 Vaccination: A Systematic Review. *Vaccines (Basel)*. 2024 Mar 14;12(3):306. doi: 10.3390/vaccines12030306. PMID: 38543940; PMCID: PMC10975425.
5. **Ameer MZ, Haiy AU, Bajwa MH, Abeer H, Mustafa B, Ameer F, Amjad Z, Rehman AU.** Association of Parsonage-Turner syndrome with COVID-19 infection and vaccination: a systematic review. *J Int Med Res*. 2023 Jul;51(7):3000605231187939. doi: 10.1177/03000605231187939. PMID: 37523491; PMCID: PMC10392513.
6. **Mamula CJ, Erhard RE, Piva SR.** Cervical radiculopathy or Parsonage-Turner syndrome: differential diagnosis of a patient with neck and upper extremity symptoms. *J Orthop Sports Phys Ther*. 2005 Oct;35(10):659-64. doi: 10.2519/jospt.2005.35.10.659. PMID: 16294987.
7. **Malik K, Giberson C, Ballard M, Camp N.** Multimodal Pain Management for Parsonage-Turner Syndrome in the Acute Rehabilitation Setting: A Case Report. *Cureus*. 2023 Aug 9;15(8):e43216. doi: 10.7759/cureus.43216. PMID: 37692705; PMCID: PMC10487277.