CLINICAL VIGNETTE

Neuralgic Amyotrophy: Parsonage-Turner Syndrome

Angela Ruman, MD

A 48-year-old male who presented with sudden onset of right upper extremity tingling and weakness over 24 hours. He specifically complained of right periscapular pain and weakness and numbness in his right hand. He initially developed pain behind his right shoulder, which persisted for 5 days and was followed with numbness in the medial forearm down to the 4th and 5th fingers and weakness involving arm and hand.

He described awakening and noting numbness and tingling involving his right shoulder and arm. In addition he reported generalized weakness of his right arm. At the time of the visit the numbness and tingling had lessened and was more localized to the ulnar aspect of his hand and arm. His weakness had improved but was still present.

He had no previous episodes and denied any recent trauma to the right arm. The pain radiated up the right side of his neck. He denied fevers, chills, headaches, visual changes, or dizziness. He also denied chest pain or numbness or tingling in any other extremity. His past medical history was significant for type one diabetes mellitus diagnosed at age 21 that was well controlled on an insulin pump. He had no known sequelae including proteinuria, eye disease or atherosclerotic heart disease. He had no significant surgical history and had no known drug allergies. Medications included daily aspirin 81 mg, ramipril 10 mg, and atorvastatin 20 mg. In addition he received insulin via an insulin pump. His family history was significant for an older brother with type 2 diabetes. He denied tobacco, alcohol or recreational drug use.

- Vital signs: BP: 109/71 pulse: 62 resp: 16 height 5”8” weight: 130 pounds. Physical exam was remarkable for normal ROM of the shoulder. Normal cranial nerves and mental status.
- Gait: normal based gait, normal heel toe and tandem walking, normal rhomberg.
- Motor: There was atrophy of the right intrinsic hand muscles and extensor forearm with normal left hand.
- Strength is 5/5 in all muscles of legs and proximal arms; with slight weak right wrist extension and finger flexors of 5th digit: right 4/5
- Sensory: intact touch, position, and vibration. Pinprick intact in all 4 extremities except for slight subjective decrease in touch and pin prick in 4th and 5th right ring fingers and hypothenar eminence.
- DTRs symmetric in all 4 extremities
- Coordination: normal rapid alternating movements bilaterally. No ataxia

The patient was referred to neurology for further evaluation and was diagnosed with Parsonage Tuner syndrome.

Parsonage-Turner syndrome or neuralgic amyotrophy was initially described in 1887 and clinically defined in 1948 by Parsonage and Turner.[1] This condition is also described as: acute brachial neuropathy, acute brachial plexitis, idiopathic brachial neuritis, and brachial plexus neuropathy, paralytic brachial neuritis and brachial radiculitis.[1,2]

Etiology

There are two different forms of brachial neuropathy. One is the idiopathic form and the other a hereditary form.[2] Hereditary neuralgic amyotrophy is about ten times less common the idiopathic neuralgic amyotrophy.[3] It is autosomal dominant with a high but not complete penetrance estimated at 80%.[4] The etiology of the idiopathic form is unknown. The current hypothesis is that the attacks are caused by an immune mediated response to one’s own peripheral nerves.[4] Immune mediated mechanisms may explain the occurrence of attacks but do not explain why certain individuals are susceptible to neuralgic amyotrophy.[4]

Evidence suggests that neuralgic amyotrophy is a disorder with a complex pathophysiological mechanism in which autoimmune, genetic and external factors all seem to play an interwoven role.[4]

It has been associated with recent viral infection such as an upper respiratory tract infection as well as recent vaccination.[2] Recent infection precedes the development of the disease in 25% to 55% of patients, while recent vaccination has been associated in 15% of patients.[2] Other causes include strenuous exercise, pregnancy and post-surgical plexopathy.[2]

In the idiopathic form patients usually experience only one attack in their lifetime. In the hereditary form attacks occur more frequently in almost 75% of patients.[4] The hereditary form is due to various type of mutations in the Septin (gene, chromosome 17q25. Septin 9 is a guanosine 5” triphosphate-binding protein highly expressed in glial cells.[2] It is involved in cytoskeleton regulation and function.[2]

Epidemiology

The incidence of neuralgic amyotropy is 1.64 cases per 100,000 person years.[2] Although neuralgic amyotropy is considered a rare disease the disorder appears to be as common as Guillain – Barre syndrome.[4] The hereditary form is much rarer with about 200 families known worldwide.[4] Neuralgic amyotrophy is more common in men than women.[4] Cases have been reported in patients three months to 75 years old with peak age of onset during the 3rd and 7th decades.[2]

Parsonage-Turner syndrome typically presents acutely with abrupt onset neurogenic pain involving the shoulder and arm followed by muscle weakness, muscle atrophy, and sensory loss.[1,5] The pain is usually severe involving he neck, shoulder and arm region.[4] The initial pain can last up to four weeks, but in 5% of individuals it disappears within 24 hours.[6] The paresis can take up to 2 weeks to develop.[4]

Clinical Presentation

Neuralgic amyotrophy often goes unrecognized by physicians with an average delay of 3 to 9 months before diagnosis is made.[7] It is often misdiagnosed as rotator cuff disease or cervical radiculopathy.[7]

In 96% of patients neuralgic amyotrophy presents with acute severe pain in the upper extremities, neck, or trunk.[6] It often wakes patients in the middle of the night and usually increases to maximum severity in a few hours.[4] The pain is usually described as unlike anything the patient experienced before.[4] The numerical rating scale for pain in these patients is usually a score of 7 or more.[6] It is described as a constant pain with variable quality.[2] Movement of the shoulder or arm often exacerbates it.[2] It can be somewhat relieved by elbow flexion and shoulder adduction.[2] On average, the pain may last from 2-3 hours to more than 8 weeks.[2] Once the acute stage of an attack is over, the initial pain has subsided a patchy paresis and atrophy have become evident.[4] The onset of weakness (paresis) is sudden in 80% of patients.[2] It can coincide with the pain or develop later.[2] Histologic studies have shown that the pathologic, presumably inflammatory process can cause very focal damage to one or a few of the fascicles that make up the brachial plexus trunk or chord or a peripheral nerve, while simultaneously affecting several scattered parts of the plexus as a whole. Any part of the brachial plexus, and clinically, any muscle or skin area can be involved in multiple combinations.[4] Sensory symptoms frequently do not correlate with the location of the paresis.[4,6]

Upper brachial plexus involvement is the most common presentation and both motor and sensory nerve symptoms are usually present.[1,4] This classic presentation occurs in 71% of attacks either with (50%) or without (21%) involvement of the long thoracic nerve that leads to an unstable winged scapula.[4] Women have symptoms in a middle or lower brachial plexus distribution twice as often (23%) as men (11%).[4,6]

The patchiness of symptoms-that is- motor symptoms that do not correspond to areas of sensory deficits or pain is a helpful clue aiding in clinical diagnosis.[6] The most commonly affected muscles are the spinati, serratus anterior, deltoid, biceps and triceps.[2] Sixty-six percent of cases are unilateral and 34% are bilateral.[2] Of the cases that are unilateral, 54% involve the right side.[2] No statistically significant relationship has been described between unilaterality of the disease and the patient's dominant side.[2]

One pitfall in diagnosing Neuralgic amyotrophy is that patients will complain about that part of their shoulder or arm that is most impaired by either pain or paresis but hardly notice or not give attention to other lesser impairments in strength or sensory loss.[2] About 1/3 of attacks or bilateral but usually asymmetrical in terms of severity.[2] It is important to examine the areas that the patient does not mention.[2]

Physical Examination and Diagnosis

The diagnosis involves clinical suspicion based on history and exam findings, including winging of the scapula or scapula alta.[2] The most important element for the correct diagnosis of neuralgic amyotrophy is the chronological development of signs and symptoms.[1] Winging can be observed at rest but becomes more prominent with arm movement.[2] As sensory deficits in neuralgic amyotrophy are often restricted to small skin areas it is useful to compare pinprick sensation of both sides in shoulders, arms and hands.[2]

There are no current available tests that can confirm the Parsonage-Turner diagnosis. In typical cases laboratory investigations are unnecessary.[4] CT, MRI, EMG are used to try to exclude other disease processes.[6] Other disorders that can mimic the clinical picture of neuralgic amyotrophy include cervical radiculopathy, degenerative cervical radiculopathy, disc rupture, mononeuritis multiplex and shoulder or elbow joint pathology. These other disease processes can be distinguished based on onset of symptoms, acute vs. progressive or fluctuating course, sensory and motor symptoms being in the same dermatome, symptoms involving other parts of the body such as the legs and distal arms.[4]

In the acute phase of the disease MRI of the shoulder can reveal diffuse T2 signal hyperintensities as a consequence of edema secondary to nerve demyelination.[2] The role of sensory nerve conduction studies (NCS) is unclear. One study confirmed the clinical impression that SNAP amplitudes of individual nerves in sensory NCS of the brachial plexus are rarely abnormal.[3] No correlation was found between motor deficits and sensory NCS abnormalities.[3]

EMG can evaluate the demyelination of the brachial plexus. It can show acute denervation.[1] It has to be performed 3 weeks after onset of symptoms to show any significant finding.[2]

Treatment of Parsonage-Turner/neuralgic amyotrophy is unknown. However, the severe pain associated with this

condition can be managed through the use of NSAID's and opiates.[1] The short-term use of steroids anecdotally may decrease pain and speed recovery but there has been no systematic review.[1,7] Physical therapy can help treat the musculoskeletal pain that is caused by altered biomechanics of the affected extremity. The therapy focuses on proper posture and joint mobility. Strength training is contraindicated due to muscle denervation of the affected extremity.[6] Most individuals will have complete recovery over time, but observational studies suggest that ½ to 2/3 of patients will have chronic pain and functional deficits.[6] Sixty percent of upper plexus lesions may recover in less than one year.[2]

Conclusion

The diagnosis of Parsonage-Turner /amyotrophic neuralgia can be overlooked and mistaken for neck or shoulder pathology. A three-step approach to diagnose Parsonage-Turner involves asking three questions.[4]

1. Is the pain acute, very severe and unlike anything the patient had before? If yes, neuralgic amyotrophy is likely.

2. Is there a limitation of passive arm movement including external rotation and abduction on exam? If not, then neuralgic amyotrophy is likely. If no, then shoulder joint pathology is more likely.

3. Are the symptoms of pain, paresis, and sensory disturbances in the same nerve root distribution? If not, neuralgic amyotrophy is likely. If yes, cervical radiculopathy is more likely.

If yes to these three questions, then the diagnosis of Parsonage-Turner is likely.[4]

REFERENCES

1. **Stutz CM**. Neuralgic amyotrophy: Parsonage-Turner Syndrome. *J Hand Surg Am*. 2010 Dec;35(12):2104-6. doi: 10.1016/j.jhsa.2010.09.010. Epub 2010 Oct 30. Review. PubMed PMID: 21035964.

2. **Ortiz Torres M, Mesfin FB**. Brachial Plexitis (Parsonage Turner Syndrome, Brachial Neuropathy, Brachial Radiculitis). 2017 Oct 9. StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2017 Jun-. Available from http://www.ncbi.nlm.nih.gov/books/NBK448114/ PubMed PMID: 28846332.

3. **van Alfen N, Huisman WJ, Overeem S, van Engelen BG, Zwarts MJ**. Sensory nerve conduction studies in neuralgic amyotrophy. *Am J Phys Med Rehabil*. 2009 Nov;88(11):941-6. doi: 10.1097/PHM.0b013e3181a5b980. PubMed PMID: 19404188.

4. **van Alfen N**. The neuralgic amyotrophy consultation. *J Neurol*. 2007 Jun;254(6):695-704. Epub 2007 Apr 20. Review. PubMed PMID: 17446996; PubMed Central PMCID: PMC2780663.

5. **Smith CC, Bevelaqua AC**. Challenging pain syndromes: Parsonage-Turner syndrome. *Phys Med Rehabil Clin N Am*. 2014 May;25(2):265-77. doi: 10.1016/j.pmr.2014.01.001. Epub 2014 Mar 18. Review. PubMed PMID: 24787332.

6. **van Alfen N**. Clinical and pathophysiological concepts of neuralgic amyotrophy. *Nat Rev Neurol*. 2011 May 10;7(6):315-22. doi: 10.1038/nrneurol.2011.62. Review. PubMed PMID: 21556032.

7. **van Alfen N, van Engelen BG, Hughes RA**. Treatment for idiopathic and hereditary neuralgic amyotrophy (brachial neuritis). *Cochrane Database Syst Rev*. 2009 Jul 8;(3):CD006976. doi: 10.1002/14651858.CD006976.pub2. Review. PubMed PMID: 19588414.

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