Parsonage Turner Syndrome

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INTRODUCTION

Because many work and recreation activities involve repetitive movements, shoulder pain is not uncommon, even in young otherwise healthy adults. In many cases, the cause of such pain is readily identified, and appropriate treatment is prescribed. In other cases, the source of shoulder pain may be more difficult to diagnose, particularly in a primary care setting. Parsonage-Turner Syndrome (PTS) is a rare neurologic disorder, but with a classic presentation. In 95% of cases, the syndrome is characterized by a sudden bout of acute, severe shoulder pain, followed by progressive neurologic deficits, notably muscle wasting and weakness in the affected area, and sensory abnormalities. However, due to its presumed rarity, an initial diagnosis of PTS is not likely. Instead, patient discomfort is often attributed to more common neck, shoulder, and upper limb conditions. Treatment for PTS is generally conservative, and resolution is often spontaneous. Therefore, it is important to avoid misdiagnoses, which can result in costly, unnecessary, or counterproductive procedures, and prolonged patient suffering. Fortunately, understanding the classic signs of PTS can reliably lead to a diagnosis in the vast majority of cases.

CASE PRESENTATION

A healthy, 22 year-old white male presented with atrophy and weakness of the left supraspinatus and infraspinatus muscles. Three months prior, the patient underwent a fat grafting operation under general anesthetic. The procedure involved removal of adipose tissue in the abdominal area and subsequent transplant to the subcutaneous tissue of the left forehead. Two days after the procedure, the patient awoke to excruciating pain in his left shoulder. The pain was described as 10/10, and mainly localized to the posterior aspect of the shoulder, with no radiation. The pain resolved spontaneously after three days, and pain medication was no longer necessary. However, while lifting weights three months later, the patient noticed weakness in his left arm. Two months after the onset of weakness, he began to notice atrophy of his left supraspinatus and infraspinatus muscles. Physical exam was remarkable for 3/5 muscle strength in abduction and a positive empty can test in the left arm. Vitals were within normal limits and review of systems was unremarkable for fever, chills, night sweats, weight changes, neck pain, headache, nausea, and vomiting. The patient was prescribed prednisolone, and an MRI, nerve conduction study, and physical therapy were ordered.

The potential causes of shoulder pain are numerous, ranging from trauma, to disease, to hereditary issues. Here we report on a rare, but emerging potential culprit. Parsonage-Turner Syndrome (PTS) is a neurologic disorder affecting peripheral nerves, usually the upper trunk of the brachial plexus. It is generally characterized by the rapid onset of severe shoulder pain and eventual muscle wasting and weakness in the affected area. PTS is of unknown etiology, but postulated causative factors include disease, heredity, medicines, trauma, and surgery. A definitive diagnosis of PTS is difficult as symptoms overlap with other shoulder ailments such as rotator cuff pathologies, impingements, adhesive capsulitis, cervical spondylosis, and suprascapular neuropathy. Ultimately patient history, MRI, and EMG are necessary to confirm PTS. Conservative treatment, including pain management and physical therapy, is key. Most patients progressively recover within a period of 3 years. The present case involves a 22 year-old white male and former collegiate athlete. The patient underwent a surgical procedure in order to cosmetically correct an injury sustained previously in an automobile accident, and presented with severe left shoulder pain two days post-op. Atrophy, and weakness of infraspinatus and supraspinatus muscles were observed three months after surgery, and EMG was positive for suprascapular nerve dysfunction. The patient was treated with oral steroids and hydrocodone, and underwent physical therapy for a year. Most of the strength and bulk in both muscles was regained three years after initial presentation, but he failed to have a full recovery.
INVESTIGATIONS
No labs were performed, and the MRI came back negative showing no signs of inflammation, muscle tears, cysts, or abscesses. A nerve conduction study of the left upper extremity was performed eight months after onset of initial symptoms, and it showed abnormalities in the infraspinatus and supraspinatus muscle.

TREATMENT
The patient was treated with Prednisolone for four weeks. The medication was discontinued after two weeks due to side effects of the medication. He did not notice any changes after taking the medication. The patient was then referred to physical therapy for six months, after which the patient’s muscle mass increased by about 50% and strength improved. Another six months of physical therapy was performed and muscle mass and strength continued to improve to about 80-85% of baseline.

OUTCOME & FOLLOW-UP
The patient responded well to physical therapy. However, full muscle mass and strength were not restored. Infraspinatus was more atrophic than supraspinatus, and there was more atrophy on the medial scapular border. Muscle strength was not 100% but 5/5. Physical therapy was discontinued after 12 months due to lack of insurance coverage. Activities of daily living are not affected and he is able to perform weight lifting activities again with only slight limitation in certain exercises, specifically those involving abduction.

DISCUSSION
PTS, which is now also known as brachial plexitis, neuralgic amyotrophy, or shoulder girdle neuritis, among others, was likely first identified in the late nineteenth century. However, it was not until the 1940s, with two articles appearing in the Lancet, that this syndrome was recognized as such. In the latter piece, appearing in 1948, Parsonage and Turner described their findings in 136 cases as “shoulder girdle syndrome”, later to be known as Parsonage-Turner syndrome. The etiology of PTS is unknown, but a wide range of causes have been postulated, including infection, trauma, surgery, anesthetics, antibiotic therapy, immunization, pregnancy, heavy exercise, and heredity. The syndrome has been considered rare, historically occurring in 2-3 out of 100,000 people. However, findings that are more recent have questioned the classification of PTS as a rare disease, since it may be 30-50 times more common than previously thought. Indeed, such figures suggest that 1 in 33 patients with new onset neck, shoulder, or arm complaints have PTS. Patients aged 3 months to 74 years have been afflicted, with a peak incidence in the third and seventh decades. PTS occurs more frequently in males, but recent findings indicate that neither sex, nor age of onset is helpful in distinguishing PTS from other shoulder ailments. PTS is primarily a disorder of the upper trunk of the brachial plexus and its branches, but it has been documented in other peripheral nerves, including those associated with breathing and phonation, and the lumbosacral plexus. Finally, in 97% of cases the disease is asymmetric.

Though PTS is a well-recognized clinical syndrome, with a very characteristic presentation, a clear diagnosis may be difficult for a few reasons. Foremost, it is a rare, possibly under-diagnosed syndrome, which may not immediately enter consideration in a primary care setting. Consequently, PTS is likely to be mistaken for other, more common conditions such as rotator cuff pathologies, suprascapular neuropathy, acute calcific tendinitis, adhesive capsulitis, cervical spondylitis, tumors, and cervical radiculopathy. Compounding the problem of PTS’s relative obscurity is the fact that no single test exists to confirm the syndrome. However, when equipped with the proper knowledge, the practitioner can reliably provide an initial diagnosis of PTS simply from patient history. Subsequent confirmation can generally be obtained from MRI, and EMG results.

According to van Alfen, a solid preliminary diagnosis of PTS can be made by posing these three questions (Figure 1).
Question 1:
Was there severe pain, unlike any previously experienced by the patient? If yes, PTS is likely. If no, PTS is still possible, but alternative diagnoses should be considered. Our patient described 10/10 pain; 9/10 and above are hallmarks of PTS. The intense pain often occurs upon awakening from sleep, is present in over 95% of cases, and manifests in one or more episodes over a few days to a week.1,12,14

Question 2:
Were there limitations to passive arm abduction and external rotation? If no, PTS is likely. If yes, structural pathologies, such as bursitis, calcifying tendonitis, adhesive capsulitis, rotator cuff tears, impingements, are likely. Our patient had full movement of his shoulder joint despite the pain, which most certainly ruled out the aforementioned conditions.

Question 3:
Did neuralgic deficits occur in the same root distribution? If no, PTS is likely. If yes, cervical radiculopathy is more likely. In our patient, two observations supported a diagnosis of PTS. One, the patient did not experience any radiating pain, which would imply same root distribution. Second, subsequent testing determined the patient’s only neurological deficit involved the suprascapular nerve.

Before the episode of intense pain, our patient’s history was generally unremarkable, in particular regarding activities or conditions typically associated with more common shoulder pathologies. The patient was a former college football player, but denied events that may have led to mechanical trauma of the shoulder in the period leading up to the development of pain. Moreover, the patient was right-handed, and the afflicted shoulder was the left one, a situation that further diminished the likelihood of mechanical injury to the shoulder area.

The patient’s MRI was unremarkable, showing no signs of inflammation, muscle tears, cysts, or abscesses. This finding was valuable in excluding more common causes of shoulder pain such as rotator cuff pathologies, impingement syndromes, labral tears, or adhesive capsulitis. Though an MRI can exclude many conditions, it also represents a key tool in diagnosing PTS, since it is sensitive to signal abnormalities related to denervated muscle.19 However, the MRI for this patient was taken several months after the onset of the bout of severe shoulder pain, which may explain, at least in part, the negative result.

EMG abnormalities in PTS are generally evident within 3 weeks of the onset of symptoms.20 Since PTS is believed to involve disruption of nerve conduction, EMG testing is considered the most important diagnostic test available.1,15,20 Moreover, EMG results permit the localization of the pathology to specific nerves and the muscles they supply. Our patient demonstrated an abnormal response to nerve stimulation in both infraspinatus and supraspinatus muscles, but normal function in all other nerves and muscles of the shoulder area. As well, our patient experienced atrophy of both supraspinatus, and infraspinatus muscles (Figure 2). In the majority of cases, deterioration of the affected muscle begins immediately, or within days of the remission of pain, but again in other cases, muscle wasting may not be noticed for months after the period of intense pain.1,12,22 While these results are very typical of PTS, they are also suggestive of suprascapular neuropathy (SSN).24 However, a diagnosis of SSN was not favored, for two major reasons. First, the patient was afflicted on his non-dominant side. SSN is typically, though not exclusively associated with traction of the nerve.24,25 Traction may result from repetitive activities associated with manual labor, or sports that feature prominent overhead activities, such as throwing.24 Traction of the suprascapular nerve may occur due to space-occupying lesions at the suprascapular, or spinoglenoid notches.24 However, the negative MRI would seemingly rule out these possibilities. Second, SSN is most commonly associated with insidious onset of dull, aching pain.25 In contrast, PTS is most commonly, though not always, associated with the sudden onset of constant, excruciating pain.1,2,12,21,22

In the absence of a definitive test for PTS, the steps described thus far can reliably diagnose the syndrome. Nonetheless, the diagnosis of PTS can be additionally confirmed by identifying an event in the patient’s history that may correlate with the onset of symptoms. As mentioned previously, dozens of causative factors for PTS have been postulated in the literature.1,2 In the present case, many of these factors were ruled out by patient history. For instance, antecedent disease, vaccination, or administration of medicine were all possibilities, but the patient denied any of these. As explained above, mechanical trauma was also excluded, based on patient history. Stressful exercise has been suggested as a cause for PTS, and the patient, a former college athlete, had a history of participation in strenuous exercise. However, such exertion was habitual for him and thus seems less likely as a precipitating factor. PTS may be caused by an autosomal dominant variant in the SEPT7 gene, but testing for this was not performed.14 In any case, reports in the literature suggest that the hereditary form of PTS is tenfold less likely than the idiopathic form, and it generally occurs in the second decade of life, with recurrent attacks in almost 75% of patients.2,14

In the present case, a singular event stands out. Approximately three days before the onset of pain, the patient underwent fat graft surgery as detailed above. The association of PTS with surgery has been well-documented in the literature, and of all known precipitating factors, appears to be most relevant to this case. It is unclear what specific aspect of surgery may lead to PTS, but iatro-
genic injury to the brachial plexus, stress-induced suppression of the immune system, inflammation, and anesthetics, have all been proposed.\textsuperscript{9,16,27,28,29,30,31,32,33,34,35,36,37,38} Treatment for PTS is based on both symptoms and etiology.\textsuperscript{20,37} Accordingly, no single or specific treatment modality for PTS has been validated.\textsuperscript{29} In general, the treatment for PTS is conservative, and may be divided into two phases, which reflect the typical course of symptoms.\textsuperscript{4,5,20}

**Phase One**

During this phase, the period of intense pain, NSAIDs, anti-inflammatories, opiates, and steroids may be prescribed with varying degrees of success. In the present case, a combination of NSAIDs and opiates were very effective in lowering the patient’s perceived pain from 10/10 to about 2/10. Such success in relieving patients’ pain has been reported elsewhere, and it has further been suggested that administration of these drugs may accelerate recovery.\textsuperscript{1,4,10,41} However, resistance of PTS-associated pain to such medicines has also been noted.\textsuperscript{12} Moreover, dos Santos\textsuperscript{29} has suggested that absences a clear etiology for PTS (viral vs. autoimmune, or other), administration of specific classes of drugs should be guarded.

**Phase Two**

During this phase, which commences when pain has subsided, the goal is to reestablish range of motion, and restore muscle strength.\textsuperscript{20} At first, muscle strength could be sufficiently compromised to warrant use of a sling.\textsuperscript{42} However, with consistent physical therapy over a course of 8 to 12 weeks, and encouragement that the condition will improve, positive results are generally observed.\textsuperscript{42} During rehabilitation, TENS (transcutaneous electrical nerve stimulation), and other similar modalities can help alleviate pain.\textsuperscript{1,20} Acupuncture may also be helpful, but reports of its success are mainly anecdotal.\textsuperscript{1,10} Surgery is indicated to remove any pathology (e.g., cyst, lesion), or if sufficient recovery has not occurred after two years.\textsuperscript{4,6}

Overall prognosis is good with long-term pain or weakness being a possible complication. Most patients recover by three years after onset of symptoms, however, about 10% of patients experience continued deficits.\textsuperscript{26} Patients with the hereditary form of PTS tend to have more severe symptoms and a higher recurrence rate. In the present case, pain subsided within a few days after onset of symptoms but after three years, minor weakness in abduction remained.

**CONCLUSION**

PTS is a rare condition of unknown etiology and pathophysiology, most commonly affecting the upper trunk, and branches, of the brachial plexus. It has a characteristic presentation of sudden, acute, and intense pain followed by predictable neuromuscular deficits. Several factors or conditions have been postulated to cause PTS, among them, heredity, disease, and multiple forms of medical treatment, including surgery. In the present case, a young, healthy male developed PTS following a surgical procedure unrelated to the brachial plexus, or shoulder region. A positive diagnosis is generally made from patient history, with confirmation by EMG, and MRI. Treatment is conservative, generally beginning with medicine to alleviate the initial pain, and continuing with physical therapy and related protocols, which restore muscular strength and range of motion. Increased incidences of PTS suggest this has been an historically under-diagnosed condition. As such, it is valuable to educate primary care providers on this syndrome to avoid costly, or destructive misdiagnoses.

**AUTHOR DISCLOSURE**

No relevant financial affiliations.

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