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Case Report

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A Case of Intractable Shoulder Pain with Deltoid Muscle Atrophy- A Presentation of Parsonage Turner Syndrome

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ABSTRACT

Parsonage-Turner syndrome is a rare, idiopathic plexopathy that causes severe shoulder pain followed by weakness. It is mostly unilateral, involves the brachial plexus and can present similar to several different diseases. It is ultimately a diagnosis of exclusion. Here is a case report describing the workup of a patient presenting with symptoms suspicious for Parsonage-Turner syndrome.

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Case Report

A 35-year-old male with no significant past medical history presented to the clinic after the sudden onset of severe pain in his right shoulder that woke him from sleep the night before. This was the first time this had happened and his pain was constant and sharp. The pain did not radiate and was rated at an 8/10 in severity. He has no known drug allergies. There is no family history of chronic disease. Vital signs were concurrent with acute pain. Physical exam showed that skin was intact bilaterally over both shoulders with no numbness or tingling. There was no evidence tenderness to palpation of the right shoulder. The patient had 5/5 muscle strength bilaterally. Passive range of motion was normal, but active range of motion in the right shoulder was limited due to pain. Orthopedic tests including "full cup", "empty cup", and the drop arm test were negative. Upper and lower extremity deep tendon reflexes were 2/4 bilaterally. Radial pulses were 5/5 bilaterally with capillary refill being less than 2 seconds on both index fingers. The patient had no lymphadenopathy. Heart and lung exams were unremarkable. In the following paragraphs we talk through our differential diagnosis with a brief description of each entity.

Differential Diagnosis

The immediate concern was of cervical radiculopathy or a mass lesion. Cervical radiculopathy can present as arm, neck, or shoulder pain with possible arm weakness, sensory changes, or a decrease in DTR's [1]. Cervical radiculopathy can be caused by a disc herniation or cervical spondylosis from degenerative changes, which often leads to cervical canal stenosis [2]. A mass can present similar to cervical radiculopathy depending on its location. A Pancoast tumor can cause compression of the brachial plexus, which most commonly causes shoulder pain that radiates towards the scapula or axilla. It could also cause arm paresthesia, areflexia, and weakness. This patient had no pain radiation, no decrease in DTR's, and no weakness which made a Pancoast tumor less likely, but it was still included on the differential. MRI's of the cervical spine, shoulder, and brachial plexus were obtained. These were all normal and showed no cervical radiculopathy, mass lesions, or other anatomical causes for our patient's pain.

After ruling out a mass and cervical radiculopathy, our differential diagnosis was expanded to include infections such as Herpes Zoster, HIV, Lyme disease, and Syphilis. Herpes Zoster can occur from reactivation of Varicella Zoster that gained access to sensory ganglia during primary varicella (chickenpox) infection. Reactivation can occur in times of immunosuppression, and most commonly happens in patients over the age of 50. It can cause severe neuritis, followed in 2-3 days by a painful vesicular rash, but the interval between pain and rash can be longer [3]. This patient had pain in the C5, C6, and C7 dermatomes localized to the shoulder and lateral arm, but no rash was present at this moment. Polymerase chain reaction of the patient's blood was done to test for Herpes Zoster, but this was negative. HIV was also considered. This patient lacked the classic findings of acute retroviral syndrome, such as myalgias, arthralgias, diarrhea, rash, lymphadenopathy, headache, fever, and mucocutaneous ulcers, but it was kept on the differential due to its variability in presentation [4]. An HIV antigen/antibody immunoassay test was performed with an HIV viral load test, but these were negative. Lyme disease was lower on our differential. It occurs after an infected tick bites a human, leading to transmission of Borrelia burgdorferi. It initially presents with the characteristic skin lesion of erythema migrans, which occurs in approximately 80% of patients. A variety of other symptoms can also be present, such as anorexia, headache, myalgias, and arthralgias. Early disseminated disease typically occurs several weeks to months after the initial tick bite and results in cardiac and neurologic abnormalities, but these can also be the initial presenting symptoms. This can include cranial nerve palsies, peripheral neuropathy, mononeuropathy multiplex, and heart block. Late disease consists of arthritis and encephalopathy [5]. This patient lacked the characteristic "bulls-eye" rash, but the possibility of peripheral neuropathy being the presenting symptom

kept it on the differential. Serologic tests were done for IgM and IgG to Borrelia burgdorferi, which are usually positive by the time the patient has findings of early disseminated disease [6]. These were both negative in our patient. Syphilis is caused by the spirochete Treponema pallidium, and has a variety of clinical manifestations. Primary syphilis includes a painless chancre, which is a small ulcer with a raised margin. Secondary syphilis leads to constitutional symptoms like fever, headache, anorexia, adenopathy, and rash. The rash can take almost any form, and is present on the trunk and extremities with involvement of the hands and the soles. Although it is commonly present, as many as 20% of cases of secondary syphilis will not have a rash. Patients with latent syphilis are asymptomatic, with only serologic evidence confirming infection. Tertiary syphilis can occur anywhere from 1 to 30 years after the initial diagnosis, and about 25-40% of untreated patients will develop it. Symptoms include aortitis, gummas, and CNS involvement leading to general paresis and tabes dorsalis [7-10]. Syphilis was on our differential diagnosis due to its ability to cause peripheral neuropathy, however the rapid plasma reagin (RPR) test was negative [11].

Next, we moved to possible metabolic causes such as diabetes mellitus and electrolyte disturbances. Uncontrolled diabetes can lead to many complications, such as retinopathy, nephropathy, and polyneuropathy. A small fraction of these patients can develop painful diabetic neuropathy [12]. Our patient had no signs or symptoms of diabetes, but this still needed to be ruled out. His hemoglobin A1c and fasting glucose levels were checked and found to be within normal limits. After ruling out diabetic neuropathy, we considered electrolyte disturbances such as hypocalcemia and hypomagnesemia that could lead to muscle cramps. A basic metabolic panel with magnesium levels were ordered, which were both normal.

After ruling out metabolic causes, we started to consider thoracic outlet syndrome and Parsonage-Turner syndrome. Thoracic outlet syndrome (TOS) involves compression of the neurovascular bundle in the area between the first rib, the cervical vertebrae, and the clavicle. The most common symptoms are caused by compression of the brachial plexus, leading to pain, dysesthesias, numbness, or weakness, which is called neurogenic TOS. Very rarely, prolonged compression can lead to atrophy of the muscles. However, pain with TOS is usually reproduced by sustained elevation of the arms, whereas our patient had pain at rest. In addition, our patient's sudden onset of severe pain is less consistent with TOS and the MRIs that were done showed no signs of it. Consultation with a neurologist resulted in nerve conduction studies and a needle electromyogram being performed. These studies showed axonal loss with denervation of the deltoid and infraspinatus muscles. There was mild fascicular sparing allowing him to keep his full range of motion. This finding supports Parsonage-Turner syndrome, a diagnosis of exclusion.

Parsonage-Turner syndrome, or neuralgic amyotrophy, is a rare disease with an unknown cause. Several theories exist, such as an infectious or autoimmune etiology, but none have been proven. Its incidence is thought to be 1.64-3.00 people for every 100,000. It is a nontraumatic plexopathy of the brachial plexus that often first presents at night as a severe pain that awakens the patient. The pain, usually unilateral, is often described as sharp, aching, burning, or stabbing in the shoulder and lateral arm regions. The pain can last for up to 4 weeks. The pain is then followed by weakness, anywhere from 24 hours to 2 weeks after. The nerves most commonly affected include the suprascapular, long thoracic, musculocutaneous, radial, anterior interosseous, and axillary nerves , leading to weakness in their distribution. Most patients develop a winged scapula, but our patient did not. Weakness can range from mild loss to complete paralysis, and may not resolve for several years, if it ever returns. Atrophy may also be seen in these muscles. It is a diagnosis of exclusion and the differential diagnosis can include cervical radiculopathy, masses, thoracic outlet syndrome, diabetic neuropathy, Lyme disease, and Herpes Zoster. Diagnosis of neuralgic amyotrophy is mostly clinical, but can be supported by electrodiagnostic testing, such as a needle electromyography that shows denervation. Treatment is mainly supportive, including physical therapy to maintain the remaining strength and range of motion. Glucocorticoids may be used, but their efficacy for this disorder has never been demonstrated. For persistent weakness, nerve transfers in the OR can be performed in an attempt to restore strength. There is a clinically similar hereditary version of this disorder called hereditary brachial plexopathy which is autosomal dominant and results from a mutation in the septin 9 gene (SEPT9).

Discussion

During this case report, we talked about the workup of Parsonage-Turner Syndrome. It can be suspected with the classic history of the sudden onset of severe shoulder pain at night that wakes up the patient. The differential diagnosis for this presentation is very broad and multiple possibilities were investigated and excluded prior to reaching a diagnosis. Initially Cervical radiculopathy or a mass lesion was considered as the onset of pain was acute, unilaterally, and could have originated from nerve compression. This was ruled out via an MRI of the cervical spine, brachial plexus, and shoulder that was negative for any findings that would suggest compression or mass lesion. Infectious viral etiologies that can present with neurological findings were also considered. Herpes Zoster was ruled out via PCR and HIV was ruled out with immunoassay and viral load. In regards to bacterial etiology Borrelia burgdorferi and Treponema palladium were both included in the differential diagnosis as they could both present with neurological symptoms. Our patient did not present with classic early symptoms of Lyme disease but, serologies were still obtained and were negative. Tertiary syphilis was also ruled out after rapid plasma reagin was found to be negative. Metabolic etiology was investigated with a Hemoglobin A1C, fasting glucose, a complete metabolic panel, and magnesium level. These laboratory values were all within normal limits confirming that the symptoms were not the result of diabetic polyneuropathy or another electrolyte aberration.

Findings that support the diagnosis of Parsonage-Turner syndrome, aside from the exclusion of other diagnoses, include the history and presentation. Our patient presented with Acute onset of unilateral pain within the region of the brachial plexus that lasted for a period of weeks and then resolved into weakness and muscle atrophy of the infraspinatus and deltoid muscles''. The patient had persistent weakness that took a 4 months to begin to recover strength without intervention. Another supportive finding of the diagnosis is a needle electromyogram was preformed and showed axonal loss with denervation of the deltoid and infraspinatus muscles.

Due to the sparse knowledge of Parsonage-Turner Syndrome it remains a diagnosis of exclusion. It requires multiple diagnostic test to exclude these other possible pathologies, making it a costly and time consuming diagnosis to make. Upon diagnosis treatment options are limited and in cases like our patient the syndrome resolves on its own without intervention and with varying levels

of residual neurologic effects. More research needs to be done on this disorder to determine an etiology and a treatment to better help the patients affected by it.

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