The case:

A 41-year-old man presented with moderate right shoulder pain on abduction. He had a history of sudden-onset right shoulder pain (10/10 in intensity) at rest and profound weakness on right shoulder abduction 1.5 years earlier. The pain and weakness remained at the same level for approximately 2 weeks and then gradually improved. At presentation, the patient had pain rated 7/10 in intensity and muscle strength rated 4 out of 5 (5=normal) on right arm abduction, but no pain at rest. Significant atrophy of the deltoid muscle and mild atrophy of the right pectoral muscle were noted, in addition to numbness along the lateral aspect of the right shoulder.

Figure: Axial (A) and coronal (B) T2-weighted fat-suppressed and large-field-of-view coronal short tau inversion recovery (C) magnetic resonance images of the right shoulder.

Your diagnosis?

For answer see page 130
Diagnosis:
Neuralgic Amyotrophy (Parsonage Turner Syndrome)

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A 41-year-old man who was a heavy machine operator presented with a history of sudden-onset right shoulder pain at rest and profound weakness on right shoulder abduction 1.5 years earlier. The pain was 10 in intensity (0, no pain; 10, worst pain) at onset, remaining at that level for approximately 2 weeks and then gradually improving. At presentation, the patient reported pain intensity of 0 at rest and 7 on arm abduction and external rotation. The weakness had also gradually improved from 70% dysfunction at onset to 40% dysfunction at presentation. The patient also reported the development of numbness along the lateral aspect of the right shoulder during the first 2 weeks; this remained relatively unchanged. The patient regularly engaged in boating, fishing, and hiking but reported no history of trauma immediately preceding the onset of symptoms. Physical examination revealed atrophy of the right deltoid and mild atrophy of the right pectoral muscles. Neurological examination was significant for 4/5 strength in right shoulder abduction and an 80% decrease in light touch and pinprick sensation over the right deltoid muscle area. Electromyography was suggestive of axonotmesis of the right axillary nerve, partial axonotmesis of the right pectoral nerve, and polyphasic motor units in pectoralis major consistent with nerve reorganization.

Magnetic resonance imaging (MRI) of the shoulder revealed significant atrophy and bright signal intensity involving the right deltoid (white arrows) and teres minor (black arrows) muscles, suggestive of denervation injury. Large-field-of-view coronal short tau inversion recovery magnetic resonance image showing bright signal intensity involving the clavicular head of the right pectoralis major muscle (asterisk), also suggestive of denervation injury (C).

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was present. A subtle non-specific, asymmetric brighter signal was noted in the right brachial plexus. No mass lesions of the brachial plexus, intrinsic nerve lesions, quadrilateral space or glenoid labrum lesions, or other abnormalities were evident on MRI. Overall, the MRI findings were suggestive of denervation injury involving the axillary nerve, which supplies the deltoid and teres minor muscles, and the lateral pectoral nerve, which supplies the clavicular head of the pectoralis major muscle.

Although concomitant damage to the 2 individual nerves was possible, it was considered unlikely because the patient had no history of trauma and MRI demonstrated no other potential cause. Because the lateral pectoral nerve carries fibers from the 5th, 6th, and 7th cervical nerves, the axillary nerve carries fibers from 5th and 6th cervical nerves, and these 2 nerves arise from different cords of the brachial plexus, a more central cause, such as a spinal cord lesion, was considered. However, cervical MRI showed no abnormality.

When the imaging findings of multiple nerve involvement with no obvious cause were reviewed in light of the clinical course of the disease, including sudden-onset pain intensity of 10, profound weakness of the shoulder with gradual improvement, profound atrophy of the deltoid muscle, and reorganization potentials in the pectoral muscles on electromyography, it was realized that this was a typical presentation of neuralgic amyotrophy.

**History of the Condition**

Neuralgic amyotrophy is a neurological condition that usually involves the nerves arising from the brachial plexus. The condition was described as early as 1942 by Burnard and Fox, who called it “multiple neuritis of the shoulder girdle.” Spillane described 46 cases with similar presentation and called the condition “localized neuritis of the shoulder girdle.” In 1948, the term “neuralgic amyotrophy” was coined by Parsonage and Turner, who described 136 cases. The condition came to be called “Parsonage Turner syndrome” after them. It has also been called “acute brachial radiculitis” and “paralytic brachial neuritis.” The incidence of neuralgic amyotrophy reported by the available population-based studies varies between 1.64 and 3.00 cases per year per 100,000 individuals. This indicates that neuralgic amyotrophy is encountered as frequently as other common neurological conditions such as primary central nervous system tumors, Guillain-Barré syndrome, myasthenia gravis, disk-related acute cervical myelopathy, and human immunodeficiency (HIV) encephalopathy.

**Characteristics of the Condition**

Neuralgic amyotrophy is more common in males and on the right side. Patients are usually older than 30 years, although reported patient ages range from 3 months to 79 years. Neuralgic amyotrophy occurs in 2 forms: sporadic or idiopathic neuralgic amyotrophy and hereditary neuralgic amyotrophy. Hereditary neuralgic amyotrophy is rare, has an autosomal-dominant inheritance pattern, and presents at an earlier age compared with idiopathic neuralgic amyotrophy.

The largest study of neuralgic amyotrophy published to date was conducted by van Alfen and van Engelen and highlights many important aspects of the condition. Neuralgic amyotrophy typically presents with acute-onset severe pain in the shoulder, neck, or arm regions that reaches its peak in a few hours. The patient usually reports a pain score of 7 or greater (0=no pain; 10=worst pain), and that change in posture or nonsteroidal anti-inflammatory drugs do not provide relief. The pain rarely resolves within 24 hours; however, most patients report improvement within 2 months. In 10% of patients, the initial episode may persist for 60 days. Some patients may experience recurrent episodes and some may develop persistent neuropathic stabbing or shooting pain or persistent musculoskeletal-type pain for varying periods.

The pain is usually followed by development of weakness and muscle atrophy. In most cases, the weakness develops within the first 2 weeks and usually involves the shoulder region, although any muscle supplied by the brachial plexus may be affected. Earlier studies indicated that the serratus anterior muscle (innervated by the long thoracic nerve) was the muscle most commonly involved. However, more recent studies have indicated that the muscles innervated by the suprascapular nerve (ie, the supraspinatus and infraspinatus muscles) are involved just as often, if not more so. The 2 largest imaging-based studies of neuralgic amyotrophy describe the supraspinatus and infraspinatus as the most commonly involved muscles. However, this may have been due to these imaging studies being mainly based on shoulder MRI and the serratus anterior muscle not being included in the field of view on MRI. In most cases, the weakness improves within 2 years, although some cases continue to show weakness after 3 years. However, van Alfen et al suggest that a significant proportion of patients continue to have functional limitation as a result of pain and fatigue.

The muscular atrophy may be seen as early as 2 weeks and has been reported in up to 88% of male and 75% of female patients. The aforementioned findings are accompanied by sensory signs and symptoms that may not be obvious to patients or clinicians but are reportedly seen among the majority of patients on careful examination. The distribution of paresis and sensory changes is “patchy” and may not match the distribution of pain. In fact, van Alfen suggests that if the pain, paresis, and sensory changes have the same distribution, an alternative diagno-
sis, such as cervical radiculopathy, should be considered. The physical examination findings are usually negative on passive arm rotation or abduction.14

Neuralgic amyotrophy typically involves brachial plexus nerves.4,7,8 However, studies have reported involvement of nerves outside the brachial plexus, such as the cranial nerves, phrenic nerves, and lumbosacral plexus nerves.8,11,15,16 Van Alfen and van Engelen8 reported that up to 55.6% of patients with hereditary neuralgic amyotrophy and up to 17.3% of patients with idiopathic neuralgic amyotrophy had nerve involvement outside the brachial plexus. The most common extrabrachial involvement in their study was the lumbosacral plexus.

**ETIOLOGY**

Various causes, including autoimmune process, vaccinations, and viral and bacterial infections, have been proposed for neuralgic amyotrophy.16-20 However, the precise cause remains unknown.

**DIFFERENTIAL DIAGNOSIS**

Some important differential diagnoses that should be considered include direct trauma to the muscles around the shoulder or neck; denervation injury from damage to brachial plexus or nerves secondary to trauma, neoplasms, neuropathies, infections, vasculitis, or an autoimmune process; cervical radiculopathy; cervical spondylosis with referred brachial plexus pain; damage to nerves and muscles after radiation; and nerve tumors.

**LABORATORY INVESTIGATIONS**

No laboratory test exists that can diagnose neuralgic amyotrophy.8,21 Electromyography may help with diagnosis by providing details of the distribution of nerve and muscle involvement.

**IMAGING**

Magnetic resonance imaging can play a significant role in the diagnosis of neuralgic amyotrophy and is the imaging technique of choice. The imaging findings have been well described.12,13,22 In neuralgic amyotrophy, MRI findings are essentially those of denervation injury (ie, high signal intensity on T2-weighted images secondary to increased extracellular water23) in the acute to subacute phase, and muscle atrophy with or without fatty replacement in the subacute to chronic phase best seen on T1-weighted images. Magnetic resonance imaging may also help to exclude other potential causes of nerve injury such as a mass or a cyst compressing the nerve, neural tumor, and neuritis. Magnetic resonance imaging can also be used to evaluate other causes of shoulder pain, including rotator cuff tear, labral tear, impingement, cervical radiculitis, and acute infection or inflammation. By eliminating other potential causes, MRI can help clinicians avoid unnecessary testing and surgery. Because the nonimaging literature indicates that patchy involvement of nerves is common in neuralgic amyotrophy, the imaging finding that would most strongly suggest the diagnosis of neuralgic amyotrophy is muscle involvement in a pattern that does not match the distribution of any single nerve.8

**TREATMENT**

There is no specific treatment for neuralgic amyotrophy. It is a self-limiting condition that resolves on its own. However, supportive measures can be instituted to improve patients’ functionality and preserve joint function, depending on the stage at presentation (ie, during the initial acute episode or after the acute episode with residual musculoskeletal or neuropathic pain). Tsairis et al7 reported that early administration of steroids did not alter the course of the disease and only a few patients reported pain relief. Because over-the-counter nonsteroidal anti-inflammatory drugs do not seem to provide satisfactory pain control, adding opiates has been recommended.8

**PROGNOSIS**

Earlier studies suggested that neuralgic amyotrophy had a good prognosis with complete recovery in most patients.3,9 However, more recent studies suggest that a significant portion of these patients continue to have pain and functional limitation.8 Van Alfen et al10 reported that persistent pain was experienced by one-third of patients and functional limitations by two-thirds after a mean follow-up of 2.5 years. This is not surprising given that significant atrophy of the involved muscles, which would alter the shoulder biomechanics, is noted in these patients.

**CONCLUSION**

Neuralgic amyotrophy is an uncommonly encountered neurological condition that may cause significant morbidity. In a large series, the mean interval between presentation and diagnosis was more than 10 months,8 indicating that this condition may be overlooked or difficult to diagnose. Although there is no known treatment for neuralgic amyotrophy, correct and timely diagnosis is important to avoid unnecessary testing and surgery and provide supportive therapy. Correct diagnosis will also enable physicians to inform patients of the nature of neuralgic amyotrophy and potentially prevent what has been referred to as “doctor shopping.”

An orthopedic surgeon may be the first physician to evaluate these patients because of shoulder pain. The clinical history of extremely severe pain, but lack of limitation on passive arm movement, may be confusing and may lead to a long list of differential diagnoses. Similarly, a radiologist may suggest the diagnosis based on MRI evidence of denervation involving muscles supplied by different brachial plexus nerves without an identified cause. To provide a correct and timely diagnosis, radiologists and orthopedic surgeons must familiarize themselves with neuralgic amyotrophy, its typical clinical presentation, and its imaging findings.
REFERENCES


