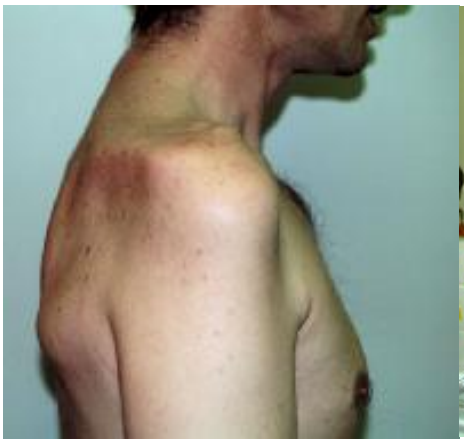
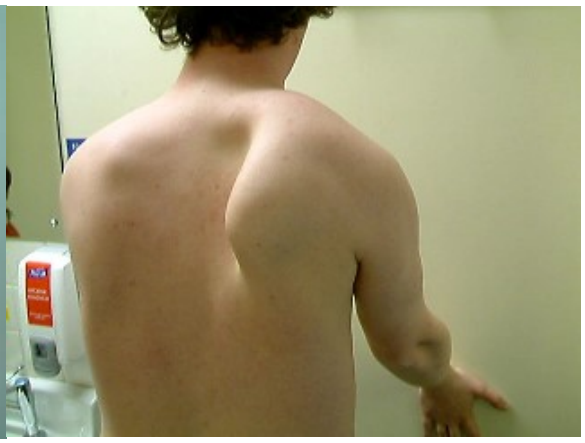
Case 3

45 –year-old presenting with a history of injury to the right shoulder whilst working in the freezing work. He was loading a sheep over an incline with his arm around the sheep. He felt pain in the shoulder but could manage to work. Subsequently there was weakness and wasting of the right arm.





Your Diagnosis?

Diagnosis: Neuralgic amyotrophy [NA] (Parsonage-Turner syndrome or brachial plexus neuritis)

1. Cause is unknown.
2. The suprascapular and axillary nerves are affected most frequently.
3. The disorder exhibits a broad range of clinical manifestations
4. Patients frequently present to physicians of different subspecialties.
5. Accurate diagnosis can be challenging and requires a thorough history and physical examination.
6. Nerve conduction velocity and imaging studies assist in the evaluation.
7. Treatment consists of symptomatic management.
8. Symptoms can persist for more than a year, but most patients note resolution of symptoms over time.

Introduction

First described by Dreschfeld in 1887 and clinically defined by Parsonage and Turner2 in a cohort of 136 patients in 1948.
It is typically characterized by attacks of neuropathic pain and subsequent patchy paresis in the upper extremity, occasionally associated with scapular winging;
Occurrence 1.6 and 3 cases per 100,000 annually, although actual incidence is likely to be at least 20 to 30 cases per 100,000 individuals secondary to under diagnosis and recognition.
The hereditary form (HNA) is much rarer.
Males are predominantly affected;
Autoimmune, genetic, infectious, environmental, and biomechanical processes have been implicated. In idiopathic neuralgic amyotrophy (INA), several antecedent events that may serve as triggers for an immune mediated etiology have been reported.
Two studies showed mononuclear inflammatory infiltrates in brachial plexus biopsies, whereas another study showed an increase in complement-fixing antibodies to peripheral nerve myelin in the acute phase of the condition.

The hereditary form of NA is autosomal-dominant and is caused by mutations in the gene septin 9 on chromosome 17q23.

Presentation

Patients typically present without constitutional symptoms

A sudden onset of severe, unrelenting shoulder pain that radiates to the arm or neck and lasts for hours to weeks.

The pain frequently can awaken patients from sleep and may be exacerbated by shoulder and elbow motion.

As the pain subsides, a flaccid paralysis with muscle weakness, muscle atrophy, and sensory loss of the shoulder girdle develops.

Average age, 28.4 years.

The most common nerves affected in NA are the suprascapular, axillary, radial, musculocutaneous and long thoracic nerve.

Clinical and electrophysiologic findings have suggested the involvement of axonal lesions of the peripheral nerves, occurring singly (mononeuritis) or in various combinations (mononeuropathy multiplex), which is reported in 75% of cases.

Pain is the first symptom noted in 90% of cases; in approximately one in five patients, pain is episodic in nature. Phases of pain can exist, starting from continuous, converting to neuropathic, and subsequently becoming musculoskeletal-type pain.

In general, weakness develops within 24 hours in approximately 33% of patients, within 2 weeks in 70% of patients, and within 1 month in 85% of patients.

The most common pattern is weakness affecting the distribution of the upper part of the brachial plexus, with or without involvement of the long thoracic nerve.

The muscles commonly affected include the infraspinatus, supraspinatus, serratus anterior, biceps, deltoid, and triceps.

Sometimes <20%, weakness is limited to the muscles supplied by a single nerve.

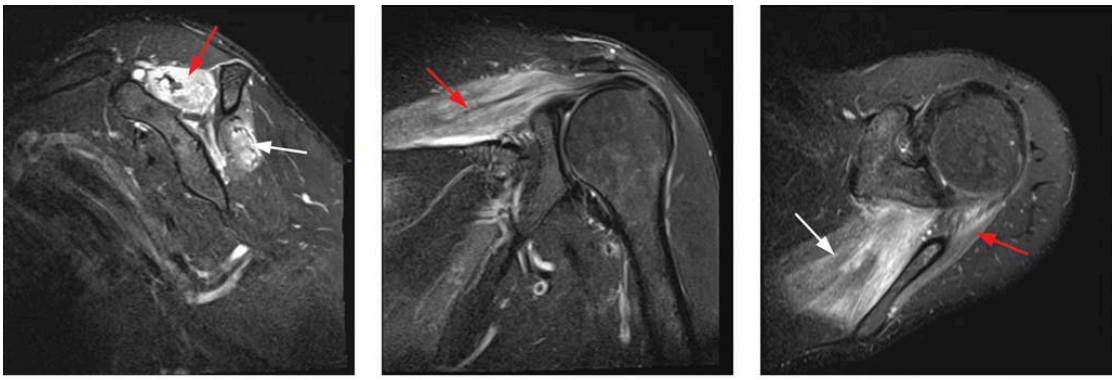
Sensory changes occur in 78% of patients. The most common sites of sensory loss are over the deltoid, the lateral aspect of the upper arm, and the radial aspect of the forearm.

Although NA typically affects motor nerves, sensory dysfunction can occur, albeit rarely, in isolation.

In pure sensory NA, the lateral antebrachial cutaneous, median, and medial

antebrachial cutaneous nerves are most often affected.

MRI



A B C

An increased intramuscular T1-weighted signal suggests atrophy with fatty infiltration and can involve one or more muscle groups of the shoulder girdle. The supraspinatus, infraspinatus, deltoid, and teres minor muscles most commonly exhibit MRI changes.

Differential Diagnosis

Neurologic Vs orthopedic conditions

In the acute stage, patients usually have full shoulder range of motion, thereby making NA more likely than primary glenohumeral joint pathology.

In addition, the abrupt onset of pain makes degenerative conditions unlikely.

Similarly, in NA, sensory changes are often peripheral rather than dermatomal in distribution.

In a patient with cervical radiculopathy, a meticulous neurologic examination can correlate nerve root pathology to the symptoms and signs. As the patient develops progressive weakness while pain resolves, the diagnosis of classic NA can be distinguished from a diagnosis of brachial plexus injury secondary to traumatic or iatrogenic reasons.

Diagnosis

1. Diagnosis is clinical
2. MRI
3. Electromyography
4. Shoulder radiographs should be obtained and evaluated for calcific tendinitis, another diagnosis that can present with severe, incapacitating pain.

Management

The natural history of NA is one of resolution over time, with abatement of neuropathic pain followed by gradual return of muscle strength and function.

No specific treatment protocol currently exists.

Early corticosteroid therapy may have a positive influence on pain in some patients and possibly hasten recovery in a few.

The primary role of physical therapy in the early phase of the syndrome is to provide the patient with strategies to help alleviate the traction on the involved nerves, which may have an increased mechanical sensitivity because of inflammation.
Adhere to physiotherapy so that biomechanical stability can be maximized.
The acute pain of NA responds best to a combination of a long-acting NSAID.
Weakness, atrophy, and decreased range of motion can be addressed with physical therapy and rehabilitation.
It is essential to realize that denervation that persists for >1 year is unlikely to recover well; any intervention to restore function should ideally be undertaken well before this time. In general, if by 6 to 9 months there is no clear evidence of regeneration or early recovery within a nerve distribution, then nerve transfer procedures should be considered.

Prognosis

The course of NA has been shown to be quite variable, with some patients showing resistance to recovery and others demonstrating complete recovery within a month after nonsurgical management.
Excellent recovery in 36% of patients within 1 year, 75% of patients by 2 years, and 89% of patients by 3 years. A more recent study found chronic pain and persistent functional deficits in almost one third of affected patients after an average follow-up of >6 years.
Two thirds of patients show beginning recovery of motor function within 1 month of the onset of weakness.
Patients with predominantly upper trunk involvement tend to have a better prognosis

than do those with lower trunk involvement.

In addition, in general, the duration of pain is correlated to the duration of muscle weakness.

Prolonged recurrent pain with no sign of motor recovery after 3 months is associated with a poor prognosis.

No relationship was found between recovery and age.

26.1% symptoms may recover

Reference

JAAOA July 2012, Vol 20, No 7; 443