



Differential Diagnosis in Patients with Possible NTOS

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M. Libby Weaver, Sheldon E. Jordan,
and Margaret W. Arnold

Abstract

Patients presenting with possible neurogenic thoracic outlet syndrome (NTOS) exhibit symptoms including pain, paresthesias, and weakness of the affected neck, arm and hand. Though these symptoms may be reproducible with certain maneuvers, the subjective nature of much of the evaluation of a patient with NTOS makes for a complex diagnostic process in which several other musculoskeletal and neurologic disorders must be considered and evaluated completely. This chapter focuses on the differential diagnoses that must be considered in anyone presenting with concern for NTOS.

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M. L. Weaver
Division of Vascular Surgery and Endovascular
Therapy, Department of Surgery, The Johns Hopkins
Hospital, Baltimore, MD, USA

S. E. Jordan
Department of Neurology, Neurological Associates
of West Los Angeles, Santa Monica, CA, USA

M. W. Arnold (✉)
Division of Vascular Surgery, Department of Surgery,
Johns Hopkins Bayview Medical Center,
Baltimore, MD, USA
e-mail: Margaret.arnold@medstar.net

Critical Take-Home Messages

1. Neurogenic thoracic outlet syndrome can be difficult to diagnose, and several neurologic and musculoskeletal alternative diagnoses should be considered.
2. Alternate diagnoses can exist independently, or in conjunction with, NTOS.
3. The presence of some additional diagnoses can not only complicate the overall clinical picture, but can confer a poor prognosis after first rib resection and scalenectomy.

10.1 Introduction

The diagnosis of NTOS is a challenging one. The differential diagnosis of shoulder and arm pain, weakness, and neuropathy is wide, with several musculoskeletal and neurologic processes to consider as primary causes of, or co-existing contributors to, a patient's symptomatology. While other chapters in this book focus on the process of diagnosing NTOS, the present chapter will focus on those entities that must be considered additionally in the differential diagnosis. Co-existing pathologies that render treatment of NTOS less effective must also be considered carefully in the diagnostic process, so that each patient's management strategy is appropriately selected. Those entities are also discussed here.

10.1.1 Cervical Spine Disorders

Cervical myeloradiculopathies, including disc disease, arthritis, and cervical spine strain, can mimic NTOS, with symptoms including upper extremity pain, weakness, and paresthesias. The presentation of pain tends to be similar, though supraclavicular fossa pain is more consistent with a diagnosis of NTOS. Patterns of paresthesia may differ in that numbness of the 4th and 5th digit are less likely to occur in patients with cervical root compression, as disc herniation and osteophytic spurs are not common at the level of C8. Additionally, limited mobility of the neck and sensitivity to axial compression and extension are more likely to occur in patients with cervical spine disorders. A positive Spurling's sign is an indicator of cervical root pathology. If cervical radiculopathy is suspected in the patient presenting with NTOS-like symptoms, MRI can be useful in the diagnosis, as sensitivity of this imaging is high in patients with cervical disk disease [1, 2].

10.1.2 Cervical Dystonia and Other Segmental Dystonias

Cervical dystonia, or spasmodic torticollis, is a condition in which involuntary contraction of the muscles of the neck lead to spontaneous twisting or turning, resulting in neck pain and, in some cases, positional paresthesias of the hand [3, 4]. The spectrum of presentation of symptoms may vary, and those patients with more subtle dystonia and prominent pain and numbness may be referred for NTOS evaluation. Like NTOS, dystonia may be provoked by occupational or traumatic accidents. These patients may respond to botulinum injections of the affected muscles. Combined use of EMG and ultrasound guidance may more accurately define the specific muscles that are contracting and contributing to the patient's pain, therefore identifying involvement of muscles other than the anterior scalene and providing useful diagnostic information.

10.1.3 Rotator Cuff Pathology

Rotator cuff injury and pathology is the most common reason that patients seek work up and treatment for shoulder pain [5]. Typically, rotator cuff pathology arises from a partial tear of the undersurface or articular portion of the supraspinatus tendon [6]. Typical mechanisms for injury include trauma to the shoulder as well as sports or occupations that involve repetitive overhead activity. This can complicate the NTOS work up, as often NTOS patients have a similar mechanism of injury or repeated overuse, such as in the case of competitive swimmers or baseball pitchers.

The patient's primary presenting complaint is pain, which is typically located at the lateral deltoid. Patients may also have weakness, especially with external rotation of the shoulder. Isolated rotator cuff injuries should not result in paresthesia down the arm or hand numbness. On physical exam, these patients generally have pain with overhead positioning (similar to an NTOS patient) and have weakness on external rotation of the shoulder. They may have pain on palpation of the shoulder joint and deltoid, but generally do not have significant tenderness to palpation of the scalene triangle.

If a rotator cuff injury is suspected, MRI of the shoulder can accurately detect full thickness and most partial thickness tears, but may not pick up subtle pathology of the rotator cuff.

Complicating matters, patients may be diagnosed with rotator cuff syndromes or labral tears only to develop prominent NTOS symptoms in spite of apparently technically adequate shoulder surgeries. Whether NTOS was the problem to start with or was created secondarily by such therapy is unknown.

Similarly, athletes or others engaged in repetitive overhead activity may develop impingement syndrome, which was described by Hawkins, et al. as shoulder pain "due to impingement in the vulnerable avascular region of the supraspinatus and biceps tendons." These injuries can degenerate into rotator cuff injuries. The most common

physical exam finding in these patients is the “impingement sign” which is pain when the arm is flexed forward [7].

10.1.4 Suprascapular Nerve Entrapment

The suprascapular nerve arises from the superior trunk of the brachial plexus with contributions from the anterior primary rami of C5 and C6 nerve roots. It is a mixed motor-sensory nerve that supplies the supraspinatus and infraspinatus muscles, both of which are part of the rotator cuff muscle group. The nerve can be injured with repetitive overhead activities, with trauma, or can be associated with a rotator cuff injury. Patients with suprascapular nerve entrapment present with shoulder pain that is typically located on the top and back of the shoulder as well as weakness with forward flexion and external rotation. This condition is relatively rare, but should be considered when the patient presents with poorly defined posterior shoulder pain. Work up includes thorough physical exam, specifically focused on shoulder weakness [8]. If suprascapular nerve entrapment is suspected, MRI may be helpful in identifying and distinguishing pathology.

10.1.5 Neurogenic Pectoralis Minor Syndrome

Neurogenic Pectoralis Minor Syndrome (NPMS) may occur separately from, or in conjunction with, NTOS. In NPMS, the brachial plexus is compressed at the infraclavicular level as the neurovascular bundle traverses beneath the pectoralis minor tendon at its insertion into the superomedial surface of the coracoid process. In one series, patients with NTOS were found to have co-existing NPMS in 75%, and NPMS alone in as much as 30% [9]. Symptoms of pain, weakness, and paresthesias of the upper extremity are similar, but it can be differentiated from NTOS in that there is generally tenderness to palpation

directly overlying the coracoid process as opposed to overlying the scalene muscles. A pectoralis minor block with local anesthetic or Botox may aid in confirming the diagnosis. The surgical treatment for NPMS is pectoralis minor tenotomy.

10.1.6 Brachial Neuritis (Parsonage-Turner Syndrome)

Brachial neuritis, also known as neuralgic amyotrophy or Parsonage-Turner Syndrome, is an inflammatory disorder of the brachial plexus. The etiology is not fully defined, but may be due to autoimmune, genetic, infectious or biomechanical causes. Nerves most often involved are the suprascapular and axillary, but it may also involve the musculocutaneous, long thoracic and radial nerves. The typical presentation of brachial neuritis is sudden onset of severe shoulder pain followed by resolution of pain with subsequent development of weakness and paresthesias in the upper extremity. The paresthesias are typically located over the deltoid, the lateral aspect of the arm and the radial aspect of the forearm. It can also be associated with a winged scapula. It may be bilateral in up to 30% of cases. The classic presentation of acute onset (days) of pain followed by resolution with subsequent development of (sometimes profound) weakness can help differentiate it from NTOS.

10.1.7 Carpal Tunnel Syndrome

Carpal Tunnel syndrome (CTS) is a condition whereby entrapment of the median nerve occurs at the level of the wrist as the nerve traverses the carpal tunnel. Characteristically, patients present with numbness to the palmar aspect of the first three and one half digits of the hand. Clinical findings, such as positive Tinel’s sign and Phalen maneuver, are combined with electrodiagnostic studies to confirm the diagnosis [10]. The Cervical Brachial Symptom Questionnaire

(CBSQ) is a useful tool to diagram symptoms. This can help distinguish paresthesia patterns of carpal tunnel from those of NTOS, which will generally involve the entire hand, or the ulnar aspect of the hand and forearm. It is worthwhile to note that patients presenting with CTS will commonly have a positive Tinel's sign at the wrist (overlying the median nerve), whereas those with NTOS generally exhibit a positive Tinel's sign overlying the ulnar nerve or brachial plexus at the level of the cubital tunnel, interscalene triangle, or axilla. In both CTS and NTOS, patients may experience worsening of symptoms at night, with repetitive movements of the hand, or with elevation of the affected extremity. Most patients with CTS, however, exhibit distal upper extremity symptoms, whereas those with NTOS often also have a proximal component.

Nerve conduction studies across the wrist are often very helpful in distinguishing CTS from NTOS. Most patients with CTS will have slowed sensory or motor velocities for the median nerve at the wrist. If atrophy is present, patients with CTS will have weak and atrophic thenar muscles while those with severe NTOS will demonstrate more widespread atrophy involving the hypothenar muscles as well. The relative importance of NTOS or CTS in a patient with multifocal disease may be determined by analyzing the results of a scalene test block for TOS and the responses to wrist splinting or steroid injections for CTS.

10.1.8 Ulnar Entrapment Syndrome

Ulnar entrapment syndrome results from compression of the ulnar nerve, most commonly at the level of the elbow as the nerve passes through the cubital tunnel. This can be associated with traumatic or exercise-related injuries, as well as in the perioperative period as a result of patient positioning, most commonly in male patients undergoing intra-abdominal or pelvic operations [11]. Symptoms include paresthesias of the fourth and fifth digit with pain at the elbow and ulnar aspect of the forearm (Fig. 10.1) [12]. The patient may also exhibit weakness of the adductor pollicis muscle, which can be investigated by instruct-

ing the patient to hold a piece of paper between the thumb and index finger while the examiner attempts to remove the paper from the patient's grip. Electrodiagnostic criteria include slow motor conduction across the elbow. A Tinel's sign may be present at the elbow overlying the nerve, and patients will often describe concordant paresthesias with elbow flexion after 1 minute of positioning.

10.1.9 Peripheral Nerve Tumors

Peripheral nerve tumors affecting the brachial plexus are rare, comprising approximately 5% of all upper extremity nerve tumors. Benign tumors, including Schwannomas and Neurofibromas, are the most common. These tumors arise from the nerve sheath and can present with symptoms of nerve compression. They may be distinguished from NTOS by identification on CT or MRI [13].

10.2 Coexisting or Complicating Factors

Conditions may coexist with NTOS that will require treatment in parallel with those that target NTOS itself. The previously mentioned conditions may be NTOS mimickers, or may co-exist with NTOS. The following list of conditions often coexist with the NTOS presentation. The presence of these coexisting conditions does not supersede NTOS therapy, but rather directs the clinician to prioritize and coordinate treatment of each element so that the best outcomes are obtained.

10.2.1 Myofascial Pain

The term myofascial pain refers to the presence of localized and regional pain associated with tender and palpably tight muscles. Needle insertion or tapping the tender muscle trigger point will produce pain that may radiate down the back or along the limb some distance from the targeted

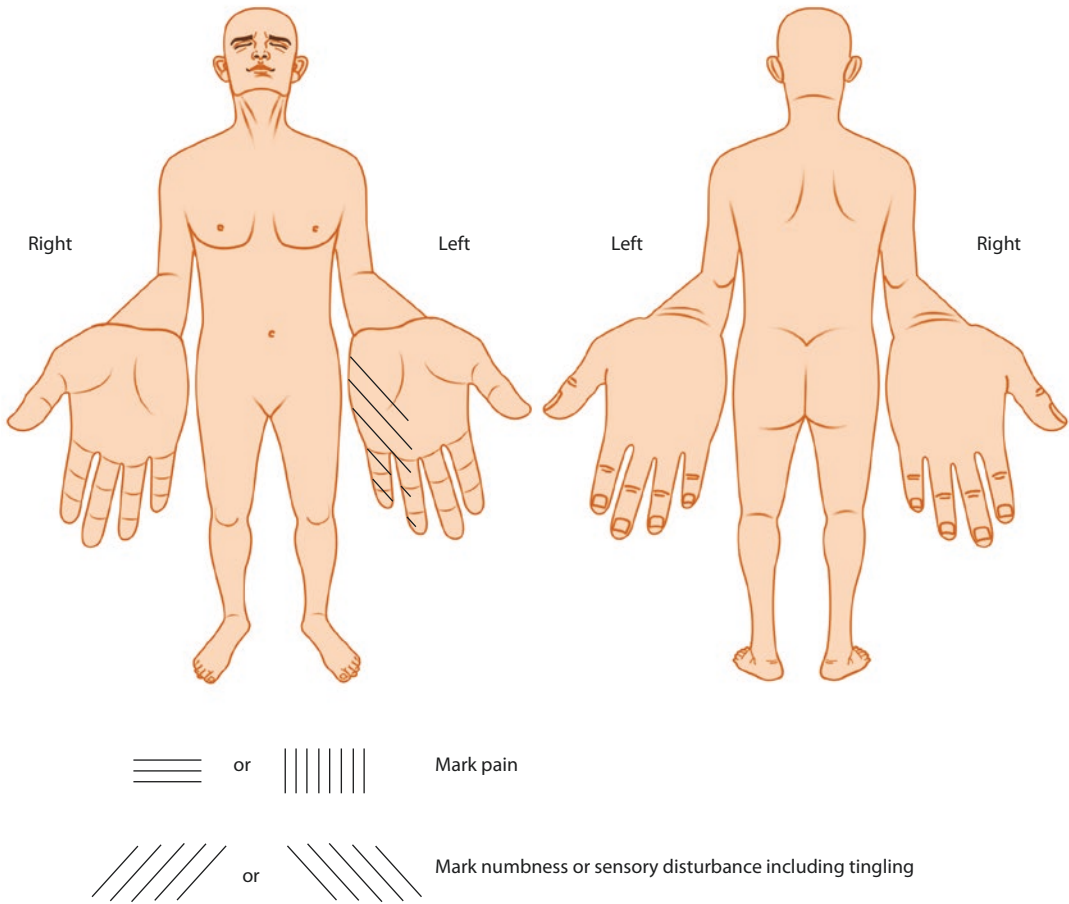


Fig. 10.1 CBSQ diagram for a patient with ulnar neuropathy

site. Many patients with muscle tenderness and tightness may not show active spontaneous motor unit activity by EMG which distinguishes this localized disturbance of muscle from patients with dystonia. The lack of active muscle contraction may be one reason why botulinum chemodenervation has not been uniformly effective in treating patients identified with myofascial pain [12, 14–17]. NTOS patients will frequently demonstrate myofascial trigger points in the trapezii, levator scapulae and rhomboid muscles.

10.2.2 Chronic Headache

Many patients will continue to have disabling headaches after technically successful thoracic

outlet decompression. The headaches may be caused by temporomandibular muscle dysfunction, frequent migraine, cervical facet arthritis, or occipital neuralgia. Referral to a neurologist or other physician specializing in the care of patients with these syndromes may be helpful.

10.2.3 Depression, Somatization and Iatrogenicity

Patients with chronic pain frequently express depressive symptoms; this condition adds to disability and will interfere with rehabilitation efforts. It is proposed that stress and emotional upset will cause or worsen pain, a process often referred to as “somatization;” for this reason

psychotropic medication, psychological counseling efforts, cognitive therapy, stress reduction techniques, and related interventions are often a part of multi-specialty pain management efforts.

Some observers have suggested that cumulative trauma disorders (including NTOS resulting from occupational factors) may result from an inability to cope with life stressors and may be partly iatrogenic in nature because of the doctor's role as an enabler [18, 19]. Such enabling may reinforce the sickness role when it leads to avoidance of unhappy work experiences and when it allows access to the rewards of entitlement programs (i.e., secondary gain). Physician and patient relationships after injury and physician participation in the process of litigation may contribute to this. The recent Australian experience with an "epidemic" of cervicobrachial occupational injuries, which was mitigated by changes in physician willingness to diagnose these cases and the scaling back of entitlement programs, is often cited as an example supporting the concept of iatrogenicity [20]. What is not often discussed in this context is the massive increase in alternative and complementary health care that occurred as clinical support from conventional medical circles waned, leading to an alternative explanation that a significant proportion of these patients simply migrated to alternative care when the conventional medical pathway became less available [20].

10.2.4 Complex Regional Pain Syndrome

Complex regional pain syndrome (CRPS) may occur independently or in association with thoracic outlet syndrome. CRPS comes in two subtypes, the most common being Type I in which the patient has no discernable peripheral nerve injury. There is no 'gold standard' test for diagnosis, rather symptoms and physical examination are suggestive of the diagnosis [21]. Diagnostic criteria were recently updated by an international consortium [22–25]. CRPS is characterized by

generalized regional pain and dramatically increased sensitivity to various somatosensory stimuli, along with associated features such as dystonia, diaphoresis, temperature fluctuations, swelling, discoloration, and late musculoskeletal changes including contracture and osteoporosis. Patients with NTOS who present with a component of CRPS may experience failure of NTOS therapy and interventions.

10.2.5 Fibromyalgia

Fibromyalgia, a chronic pain syndrome characterized by widespread achiness, pain, and stiffness, soft tissue tenderness, general fatigue, and sleep disturbances, is also associated with poor outcomes for a variety of procedures and chronic excessive utilization of medical resources [25–35]. Diagnostic criteria are based, in part, on scaled scores on the Widespread Pain Inventory (WPI) and Symptoms Severity Score (SSS) [36].

It should be noted that scales frequently used for evaluation of patients with cervicobrachial syndromes such as the DASH do not address widespread pain. By contrast the CBSQ concentrates on upper body complaints but allows the patients to map possible widespread pain on the figure of the body (Fig. 10.2). Fibromyalgia should be considered clinically when a patient has chronic pain in the upper and lower body, fatigue, non-refreshing sleep and a variety of functional complaints, often including functional bowel disease or pelvic organ dysfunction. These patients often describe multiple system complaints on a review of systems intake form. They often have concomitant depression and may complain of problems with focus or memory. Multiple allergies, food sensitivities and failed medical and surgical therapies may be present in the general history. Many of these patients may conceal a prior diagnosis of fibromyalgia out of fear of rejection. By contrast, more "acceptable" labels may be openly discussed such as Lyme Disease, Chronic Epstein Barr infection, Lupus, vitamin deficiency, heavy metal toxicity, hypothyroidism, or intestinal overgrowth.

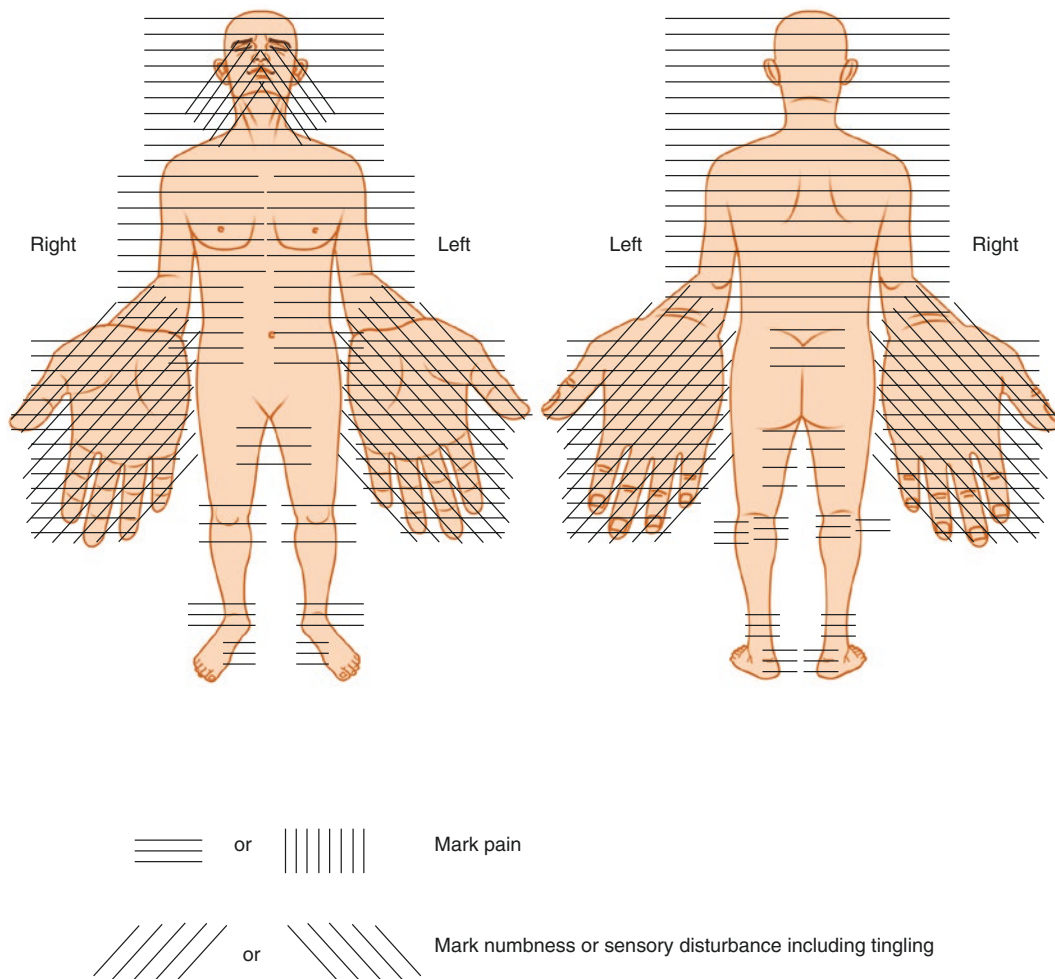


Fig. 10.2 CBSQ diagram for a patient with fibromyalgia

The available literature generally describes poor outcomes for a variety of procedures and chronic excessive utilization of medical resources in patients with widespread pain and fibromyalgia [24, 26–34, 36]. Increased awareness of, and better screening tools for, this condition will ideally delay surgical plans and allow for suitable referral. If surgery is contemplated in these patients, both doctor and patient should be aware of the potential for worsening of generalized symptoms and for the expectation of only modest functional gains in the long term with persisting poor quality of life assessments. An increased use of clinical resources will be expected in the post-operative period.

10.2.6 Opioid Hyperalgesia

In patients with chronic use of opioids, tolerance and dependency may develop. In addition, some patients will have the appearance of worsened regional pain and overall pain sensitization associated with chronic exposure to high daily doses of opioids, so-called opioid hyperalgesia. Since these patients are highly tolerant, opioids no longer seem to produce effective analgesia. It seems obvious that such patients will have more difficulty than other patients in experiencing adequate pain control in the post-operative period after thoracic outlet decompression. In fact, outcomes after any NTOS-targeted therapy

(including botulinum chemodenervation) are poor in patients with this condition.

10.3 Summary

Patients with possible NTOS can present both with NTOS mimicking conditions as well as a multifactorial cervicobrachial syndrome. In order to achieve successful outcomes with NTOS-targeted therapy, a three step process should be considered. First of all, patients should demonstrate the presence of positive features typical of NTOS including numbness, fatigue, and pain exacerbated with overhead reaching in conjunction with evidence of pathology at the supraclavicular fossa and/or pectoralis minor. Absence of these classic features should direct the clinician to broaden the differential diagnosis to look for other etiologies of the patient's symptoms. Secondly, other conditions (carpal tunnel syndrome or cervical disk disease) or co-existing conditions (depression, distal entrapment neuropathies) should be scrutinized and treated in parallel with NTOS. Third, a careful evaluation should be done to ensure that a patient does not have a coinciding condition that predicts failure of NTOS-targeted intervention (fibromyalgia, opioid hyperalgesia or CRPS). Using this algorithm, a robust and repeatable diagnosis can be assured, and patients who will have the highest likelihood of benefit from proper therapy can be identified.

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