Diagnosis of thoracic outlet syndrome

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Thoracic outlet syndrome (TOS) is a nonspecific label. When employing it, one should define the type of TOS as arterial TOS, venous TOS, or neurogenic TOS. Each type has different symptoms and physical findings by which the three types can easily be identified. Neurogenic TOS (NTOS) is by far the most common, comprising well over 90% of all TOS patients. Arterial TOS is the least common accounting for no more than 1%. Many patients are erroneously diagnosed as “vascular” TOS, a nonspecific misnomer, whereas they really have NTOS. The Adson Test of noting a radial pulse deficit in provocative positions has been shown to be of no clinical value and should not be relied upon to make the diagnosis of any of the three types. The test is normal in most patients with NTOS and at the same time can be positive in many control volunteers.

Arterial TOS is caused by emboli arising from subclavian artery stenosis or aneurysms. Symptoms are those of arterial ischemia and x-rays almost always disclose a cervical rib or anomalous first rib. Venous TOS presents with arm swelling, cyanosis, and pain due to subclavian vein obstruction, with or without thrombosis. Neurogenic TOS is due to brachial plexus compression usually from scarred scalene muscles secondary to neck trauma, whiplash injuries being the most common. Symptoms include extremity paresthesia, pain, and weakness as well as neck pain and occipital headache. Physical exam is most important and includes several provocative maneuvers including neck rotation and head tilting, which elicit symptoms in the contralateral extremity; the upper limb tension test, which is comparable to straight leg raising; and abducting the arms to 90° in external rotation, which usually brings on symptoms within 60 seconds. (J Vasc Surg 2007;46:601-4.)

Indications for subclavian arteriography in thoracic outlet syndrome

Historically, when Adson1 first described his maneuver in 1927, what today is called thoracic outlet syndrome (TOS) was then called scalenus anticus syndrome. It was thought to be an arterial problem caused by compression of the subclavian artery by the anterior scalene muscle (ASM). Many things have changed over the past 80 years. Today, it is recognized that in addition to the subclavian artery, the subclavian vein or brachial plexus can also be compressed, and structures other than the ASM can do the compressing. This is what led in 1956 to renaming this condition by the all-inclusive label, thoracic outlet syndrome (TOS).2

TOS is a broad term whose definition is “upper extremity symptoms due to compression of the neurovascular bundle by various structures in the area just above the first rib and behind the clavicle”. The term TOS does not specify whether the compressing agent is muscle or bone, and it also fails to identify the structure being compressed.

However, since it is usually fairly easy to differentiate arterial from venous or neurogenic involvement, the term TOS should be preceded by an appropriate modifier: Arterial TOS (ATOS), venous TOS (VTOS), or neurogenic TOS (NTOS).

Incidence

NTOS comprises over 90% of all TOS cases seen today while ATOS is less than 1%. Although the incidence in any one medical center is influenced by referral patterns, ATOS is the least common of the three types in all series. Our 40 years of experience in treating TOS includes more than 2500 operations, but only 11 were for ATOS, an incidence of less than 1%. The incidence of operations for VTOS was 3%, while over 95% of the TOS procedures were for NTOS. Thus, the large majority of patients presenting with arm pain and paresthesia will have neurogenic, not arterial, TOS.

Etiology

The history prior to the onset of symptoms can be helpful. Most patients with NTOS have a history of neck trauma preceding their symptoms, auto accidents being the most common and repetitive stress at work being next most common. VTOS, sometimes called effort thrombosis or Paget-Schrotter disease, may be preceded by excessive activity with the arms. Symptoms of ATOS usually develop spontaneously, unrelated to trauma or work. Arterial TOS is almost always associated with a cervical rib or an anom-
lous first rib and, thus, a normal neck x-ray is a good screening test to rule out ATOS.

Cervical ribs

Cervical ribs occur in less than 1% of the population and about 70% are in women. Most cervical ribs are asymptomatic. However, the cervical rib is a predisposition to develop NTOS following neck trauma, most often whiplash injuries.

Symptoms

The term “vascular” TOS is nonspecific. It implies arterial or venous compression but it also expresses uncertainty as to which structure is involved. Thus, the term “vascular TOS” should be replaced by a more specific label, which can be differentiated by history, symptoms, and physical findings as outlined below.

A. Arterial TOS. The symptoms of ATOS include digital ischemia, claudication, pallor, coldness, paresthesia, and pain in the hand but seldom in the shoulder or neck. These symptoms are the result of arterial emboli arising either from mural thrombus in a subclavian artery aneurysm or from thrombus forming just distal to subclavian artery stenosis. In this situation, the pallor and coldness are due to arterial ischemia and not Raynaud’s phenomenon. The arm swelling seen in VTOS is not a feature of either ATOS or NTOS. Paresthesia in the fingers and hands is common in venous TOS and may be secondary to swelling in the hand rather than to nerve compression in the thoracic outlet area.

B. Venous TOS. Swelling of the arm, plus cyanosis, is strong evidence of subclavian vein obstruction, either thrombotic or nonthrombotic. Pain or aching is often present, but may also be absent. The arm swelling seen in VTOS is a feature of either ATOS or NTOS. Paresthesia in the fingers and hands is common in venous TOS and may be secondary to swelling in the hand rather than to nerve compression in the thoracic outlet area.

C. Neurogenic TOS. Pain, paresthesia, and weakness in the hand, arm, and shoulder, plus neck pain and occipital headaches, are the classical symptoms of NTOS. Raynaud’s phenomenon, hand coldness and color changes, is also frequently seen in NTOS. It is the latter symptoms that can lead to an erroneous diagnosis of ATOS. The incidence of specific symptoms in our last 50 patients is shown in Table I.

In NTOS, the coldness and color changes are not caused by ischemia resulting from obstruction of the subclavian artery. Rather, they are due to an overactive sympathetic nervous system whose fibers run on the circumference of the nerve roots of C8, T1, and the lower trunk of the brachial plexus. When the nerves are irritated or compressed, the sympathetic fibers are activated, producing Raynaud’s phenomenon. This explains how the coldness and color changes are frequently seen with NTOS. Because the same coldness and color changes also occur in ATOS, differentiating between the two must be done by evaluating other signs and symptoms, which can easily establish the diagnosis.

Physical examination

VTOS is easily identified on physical exam by arm swelling, cyanosis, and distended superficial veins over the shoulder and chest wall.

NTOS and ATOS can be differentiated by physical findings. NTOS usually demonstrates tenderness over the scalene muscles and duplication of symptoms by the following provocative maneuvers:

1. Neck rotation and head tilting (ear to shoulder), which elicits symptoms of pain and paresthesia down the contralateral side.
2. Abducting the arms to 90° in external rotation (90° AER), which brings on symptoms within 60 seconds and often, in less than 30 seconds.
3. Modified upper limb tension test of Elvey (ULTT). The later test is comparable to straight leg raising in the lower extremity. We were introduced to the ULTT several years ago by Dr Ron Stoney and have found it very helpful in the diagnosis of NTOS. The numbers of positive findings in our last 50 patients are shown in Table II.

Upper Limb Tension Test of Elvey (ULTT)

We have modified the original test of Elvey by starting with Elvey’s second position, performing the test with the patient sitting up, and executing the maneuvers actively rather than having the examiner perform them passively (Fig). Carrying out the test in this way permits both limbs to be tested simultaneously and permits the asymptomatic side to serve as a control for the symptomatic one. The modified ULTT is performed with three maneuvers in succession as follows (Fig):

Position 1. Abduct both arms to 90° with the elbows straight.

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>50 patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neck pain</td>
<td>88</td>
</tr>
<tr>
<td>Trapezius pain</td>
<td>92</td>
</tr>
<tr>
<td>Supraclavicular pain</td>
<td>76</td>
</tr>
<tr>
<td>Chest pain</td>
<td>72</td>
</tr>
<tr>
<td>Shoulder pain</td>
<td>88</td>
</tr>
<tr>
<td>Arm pain</td>
<td>88</td>
</tr>
<tr>
<td>Occipital headache</td>
<td>76</td>
</tr>
<tr>
<td>Paresthesia:</td>
<td>98</td>
</tr>
<tr>
<td>All 5 fingers</td>
<td>58</td>
</tr>
<tr>
<td>4th and 5th fingers</td>
<td>26</td>
</tr>
<tr>
<td>1st-3rd fingers</td>
<td>14</td>
</tr>
<tr>
<td>No Paresthesia</td>
<td>2</td>
</tr>
</tbody>
</table>
Position 2. Dorsiflex both wrists.

Position 3. Tilt the head to one side, ear to shoulder. Each maneuver progressively increases stretch on the brachial plexus.

In ATOS, an absent radial pulse at rest is common, as emboli often lodge near the antecubital space. Arm abduction to demonstrate pulse obliteration is seldom necessary. There usually is no scalene muscle tenderness; neck rotation and head tilt elicit no symptoms; and the ULTT may be normal. In ATOS, most of the symptoms are in the hand and forearm, fewer in the shoulder girdle and neck. The diagnosis of ATOS can usually be established with clinical information alone.

**Diagnostic procedures**

**X-ray.** A simple x-ray of the chest or neck will easily detect a cervical or anomalous first rib. If there is none, this essentially eliminates ATOS. (Theoretically a tight scalene muscle or ligament could compress the artery, but we have never seen such a case). Even if there is a cervical rib, most symptomatic cervical ribs cause symptoms of neurogenic, not arterial, TOS.

**Adson test.** In 1927, and again in 1947, Alfred Adson described his now famous test. “The patient takes a long deep breath, elevates the chin, and turns it to the affected side. This is done as the patient is seated upright, with his arms resting on his knees. An alteration or obliteration of the radial pulse or change in blood pressure is a pathognomonic sign of a scalenus anticus syndrome.”

Since its original description, Adson’s “infallible” test has been disputed by several investigators. In 1945, Wright noted that pulses could be obliterated by turning the head to either the ipsilateral or contralateral side. In 1965, Woods noted that in TOS patients, the Adson test was more often positive when turning the head to the contralateral side (63%) than to the ipsilateral side (22%). In reviewing several reports of patients with NTOS in whom the Adson test was performed, the incidence of positive responses ranged from 22% to 100% with a median of 31%.

Between 1980 and 1998, four studies of healthy volunteers investigated the effect of postural maneuvers on the radial pulse, including the Adson test. Pulse obliteration or reduction of amplitude occurred in 53%, 9%, 15%, and 11%, respectively.

**Ninety degree abduction in external rotation stress test (90° AER).** In 1963, Gilroy and Meyer introduced a modification of the Adson test, the 90° AER maneuver, which is performed by having the patient elevate the arms in a “stick-em-up” position. This is a provocative test that was popularized by Roos in 1966 and is sometimes referred to as the elevated arm stress test (EAST test of Roos). This test is very helpful in the diagnosis of neurogenic TOS. A positive test is reproduction of the patients’ symptoms of pain and paresthesia within 60 seconds but not necessarily reduction of the radial pulse. Our own statistics found that with this maneuver, 94% of NTOS patients had reproduction of symptoms while only 24% demonstrated a diminished pulse.

**Arteriography.** The role of arteriography in TOS is a very limited one. The primary purpose of arteriography is to help the surgeon plan arterial reconstruction. It should not be needed to differentiate between arterial and neurogenic...
compression. This should be done by history and physical exam plus pulse volume recordings and noninvasive duplex scanning when ATOS is under consideration. Arteriography should be employed only when a patient has signs and symptoms suggestive of arterial insufficiency or ischemia and when surgery is under consideration. Duplex scanning often can screen for a subclavian artery aneurysm or stenosis and when such is present, arteriography can then be used to confirm the diagnosis and plan surgical repair. In the absence of these criteria, arteriography is not indicated. When the criteria are met, subclavian abnormalities are usually evident with the arm at rest so that dynamic positioning during arteriography (abducting the arm to 90° and 180°) is seldom needed.

Magnetic resonance arteriography (MRA) does not carry the risks of transfemoral arteriography. Its only risk is the dye itself. However, the same indications should apply to all types of arteriography. MRA is not indicated in NTOS and is simply an unnecessary expense.

Subclavian arteriograms in patients with NTOS will be normal with the arm at the side but some patients will demonstrate subclavian artery compression on dynamic positioning. This finding does not establish a diagnosis of ATOS or NTOS.

Noninvasive vascular lab studies. Arterial pulse reduction with arm elevation can be documented with a pulse volume recorder. However, in patients with symptoms of NTOS, this test is not indicated for the same reason that arteriography is not indicated. Utilizing a vascular sign to diagnose a neurogenic condition is unreliable. Although there is no risk involved with vascular lab studies, it represents an unnecessary study and an unwarranted expense. These are indicated only in patients with clinical findings of ATOS.

Electrodiagnostic testing. Electromyography (EMG) and nerve conduction velocity (NCV) tests are normal in the large majority of patients with clinical signs of NTOS. When positive, the changes are usually nonspecific. In a small number of patients, usually those with cervical ribs and hand atrophy, electrodiagnostic studies reveal typical changes of ulnar neuropathy. This area may be about to change. Recently a study from France was published on the NCV of the sensory medial antebrachial cutaneous nerve. Measurements of this nerve revealed abnormalities in clinical NTOS patients in whom all other electrodiagnostic studies were normal. This could prove to be a very helpful objective test to confirm diagnoses that, to date, have been made only by subjective criteria.

CONCLUSIONS

1. The term “TOS” is not a specific diagnosis. It should be replaced by specifying the appropriate type of TOS before the term, such as neurogenic TOS, arterial TOS, or venous TOS, each of which can be differentiated by appropriate history and physical exam.
2. The term “vascular TOS” should be abandoned in favor of a more specific diagnosis.
3. Adson test is not reliable to either rule in or rule out a diagnosis of neurogenic TOS. It is employing a vascular sign to diagnose a neurologic condition, which frequently is misleading.
4. The upper limb tension test (ULTT) is comparable to straight leg raising in the lower extremity and is very helpful in the diagnosis of neurogenic TOS.
5. The purpose of arteriography is to assist the surgeon in planning and when surgery is under consideration. Duplex scanning when ATOS is under consideration. Arteriography should be employed only when a patient has signs and symptoms suggestive of arterial insufficiency or ischemia and when surgery is under consideration. Duplex scanning often can screen for a subclavian artery aneurysm or stenosis and when such is present, arteriography can then be used to confirm the diagnosis and plan surgical repair. In the absence of these criteria, arteriography is not indicated. When the criteria are met, subclavian abnormalities are usually evident with the arm at rest so that dynamic positioning during arteriography (abducting the arm to 90° and 180°) is seldom needed.

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REFERENCES


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