Thoracic outlet syndrome: a narrative review.

Síndrome do desfiladeiro torácico: uma revisão narrativa.

INTRODUCTION

In 1956, Peet et al. \(^1\) first used the term Thoracic Outlet Syndrome (TOS) to describe a constellation of symptoms caused by compression of neurovascular structures in the thoracic outlet region. This compression may take place between the clavicle and the first rib or by a number of anatomical variations. Neurological compression is the most common form of thoracic outlet syndrome. Vascular complications occur infrequently. Arterial complications usually result from compression of the subclavian artery by a complete cervical rib. Venous complications are often related to muscle compression of the subclavian vein. The neurogenic form, previously described, is the most common, constituting more than 95% of cases, while the venous represents 2% to 3%, and the arterial, about 1%. Risk factors include biotype and individual variations such as genetics, age and gender. In Brazil, there are no data on the epidemiology of TOS. Given the suspicion of TOS, a detailed clinical evaluation is necessary, followed by complementary exams to elucidate the cause. The treatment is directed according to the etiology and the presence or absence of complications. The purpose of this study was to perform a narrative review on TOS, focusing on its etiology, pathophysiology, epidemiology, clinical evaluation, complementary exams, differential diagnoses, and treatment.

LITERATURE REVIEW

We searched for scientific articles in PubMed databases using the MeSH term: "thoracic outlet syndrome". Inclusion criteria were publications between 2012 and 2017 and the accepted study types were review, systematic review, meta-analysis, clinical trials, and observational studies. After the search, we found 47 articles, of which we selected eight for this work.
RESULTS AND DISCUSSION

Etiology and pathophysiology

TOS occurs due to an anatomical or muscle variation that compresses the subclavian-axillary vessels and/or the brachial plexus in the region of thoracic outlet. Compression occurs mainly in three areas: in intercostal-scalene triangle, the costoclavicular space and the retrocoraco-pectoral space. The vascular and neural regions pass through these spaces and, due to their small size, any bone or muscle deformity may exert compression and cause neurogenic and/or vascular clinical symptoms.

The compression of the neurovascular bundle as it crosses the thoracic outlet can result from a combination of anomalies development, injuries and physical activities, which predispose to local compression. Both congenital and acquired thoracic outlet anatomy variants are common and mainly include variations in bone and muscle anatomy. Changes in the brachial plexus anatomy and muscle histology may also contribute. The anatomical causes of TOS can be organized into soft and bone tissue categories. Soft tissue causes are associated with up to 70% of TOS cases, while bone abnormalities comprise the other 30%.

The most common way to develop TOS is through some type of trauma. This can cause some deformation of the ribs or cervical plexus, triggering compression of the structures. The syndrome can also arise from repetitive movements over a long period, from erroneous exercises performed in weight centers, or even at work, from repetitive movements. It is believed to be due to minor trauma over a long period and to the inflammatory process that forms in the region.

In addition, the pre-existence of congenital bone and muscle abnormalities that previously did not show symptoms typical of the clinical picture of TOS may trigger symptoms after trauma.

The anatomical-morphological findings associated with TOS include the presence of ectopic cervical ribs, prominent C7 transverse processes, tumor in the region or previous trauma sequelae (Figure 1). Although the presence of cervical ribs may cause symptoms of TOS in the absence of trauma, 80% of patients with TOS and cervical ribs show symptom development only after injury. These patients usually have a large cervical rib fused to the first rib.

Epidemiology

TOS’s epidemiological data should be interpreted with caution, since cases are rarely reported due to the lack of agreement on the diagnostic criteria for the disease. On average, the incidence ranges from three to 80 cases per 1,000 people. The neurogenic form accounts for over 90% of TOS cases. About 3% to 5% manifest the venous form and less than 1% manifest the arterial form. The incidence of the venous form is 1:100,000 people per year.
The typical age range of development is between 20 and 30 years for vascular forms and between 20 and 40 years for the neurogenic form. The prevalence of the neurogenic form is higher in women, at a ratio of 3.5:1. The venous form, in turn, seems to be more common in men. The arterial form has no gender prevalence.

**Clinical evaluation**

The symptomatology of the patient with TOS depends primarily on the pathophysiological mechanism involved in the syndrome. Daniels et al. state that the patient with the arterial form of TOS may complain initially of persistent, non-radicular pain, numbness and discomfort in the affected extremity, becoming worse with physical exercise and improving with rest. However, there are no pathognomonic signs and symptoms of the arterial form. On physical examination, it is common to find coldness and paleness of the limbs. Paleness is more common in the upper limb, and skin changes in the distal limb. Ulcerations and signs of microembolic events are rare.

For Grunebach et al., the first typical symptom of the venous form is upper limb edema, which may eventually be preceded by severe pain a few days earlier. On physical examination, the patient may present cyanotic coloration of the upper extremity. Superficial veins become dilated in the upper arm, neck and chest.

For Franklin et al., classic symptoms of the neurogenic form include pain, paresthesia in the fingers, and weakness in the upper extremity. On physical examination, it is common to find pain on palpation above the brachial plexus, trapezius and scalene muscles, and anterior chest wall, but it is not pathognomonic of the syndrome. Symptoms are usually worse with activities that involve overloading, such as picking up an object on a high shelf, or involving pronounced limb extension.

In a systematic review, occipital headache was present in 76% of patients.

Some triggering maneuvers for TOS has been described in the literature. The Adson’s maneuver is performed by palpation of the radial pulse, elbow extension, lifting of the upper limb and ipsilateral head rotation while the patient performs a deep inspiration. The test may produce paresthesia by the distribution of the brachial plexus and, often, obliteration of the radial pulse of the affected side. The test alone does not confirm the diagnosis of TOS, since 40% of people without symptoms may have pulse disappearance just by moving the arm away from the body, and it should not impact the diagnosis of the venous form of TOS. The Elvey’s maneuver is performed with extension of the affected upper limb and contralateral rotation of the head. It checks for the presence of nerve compression in the thoracic outlet and may trigger paresthesia and pain. It also has low sensitivity and specificity. The Ross’s test is performed with the affected limb in 90° abduction and 90° elbow flexion while the patient slowly opens and closes the hands for three minutes. The test constricts the costoclavicular space, causing the patient to become unable to perform it at a minimum time. The Wright’s test, on its turn, is performed by hyperabduction and external rotation of the affected limb, while the patient rotates the head to the contralateral side, being positive with the total or partial obliteration of the radial pulse. However, this test obliterates the radial pulse in more than 7% of the normal population.
There is still no consensus on the use and diagnostic accuracy of clinical maneuvers as the gold standard in the diagnosis of TOS. This is justified by the low positive predictive value of the maneuvers. Nevertheless, an approach that combines the patient’s clinical history with careful assessment of maneuvers is still recommended.

**Complementary exams**

Patients with symptoms of TOS are often subjected to several additional tests such as magnetic resonance imaging (MRI), computed tomography (CT), electromyoneurography (EMNG), sensory and motor nerve conduction studies (NCS), and ultrasonography (USG). Imaging tests are useful in diagnosing vascular forms of TOS, but may have normal results in patients with the neurogenic form. For Chavhan et al., imaging exams play an important role in the diagnosis and management of TOS.

USG is the initial imaging exam chosen for the evaluation of vascular forms of TOS, since it is easily accessible and noninvasive. According to Moore et al., the duplex scan has high sensitivity and specificity in detecting venous stenoses or occlusions when performed by an experienced professional. For Chavhan et al., MRI is preferred for its ability to show bone, soft tissue, vascular and nerve abnormalities with its contrast superior and resolution. Limb angio-MRI allows excellent imaging of vessels and can be an excellent diagnostic tool. Furthermore, MRI neurography can detect brachial plexus compression, contributing to the diagnosis of the neurogenic form of TOS. Czihal et al. states that CT demonstrates good enhancement of vascular structures in the vicinity of bones and muscles. Angio-CT and venography produce high-quality, three-dimensional reconstruction images of the central vasculature and vessels extremities, which facilitates the identification of the vascular compression point and extent of the condition.

According to Kuhn et al., neurophysiology studies may show abnormalities of nerve activity when compared with other nerves in the cervical and thoracic spine. These manifestations may occur due to abnormalities in conduction velocity in the medial cutaneous nerve of the forearm and in the motor part of the median nerve of the short thumb abductor muscle. EMNG may show fibrillation in C8 and T1 distributions, although it does not show changes in conduction velocity.

**Differential diagnosis**

The clinical presentation of TOS is quite diverse, ranging from mild discomfort to severe symptoms. In addition, patients may present with unilateral or bilateral signs or symptoms related to compression of a combination of neurological and vascular components. Isolated vascular forms of TOS are more easily diagnosed but are also rarer. Thus, the examiner must distinguish which symptoms are related to brachial plexus compression, which are of vascular nature, and which are unrelated to the pathology of the thoracic outlet.

Kuhn et al. state that the problems of the cervical spine are more often characterized by constant pain in the neck and shoulder, and depending on the neck position, there may be a worsening of the pain. Irradiation of this pain sometimes occurs to the upper limbs. Shoulder position and direct palpation of joint structures aggravate symptoms.
Distal compression neuropathies, such as carpal tunnel syndrome, have isolated symptoms, predictable in relation to nerve distributions, and are aggravated by wrist and elbow position rather than shoulder or neck position.

The type of TOS and the structures involved directly influence the type of treatment. Initial clinical treatment seeks to relieve symptoms by using analgesics, anti-inflammatory drugs, and changes in lifestyle.

The neurogenic form of TOS is initially treated with physical therapy, postural changes and the use of anti-inflammatory drugs\textsuperscript{2}. Many patients experience pain relief without surgical approach. Botulinum toxin injection in the scalene muscles has been reported, with improvement in symptoms, although randomized trials of high quality do not indicate efficacy of this approach\textsuperscript{8}. Surgical decompression can be an option if the less invasive treatments are \textsuperscript{2}.

The treatment of the venous form of TOS has historically been symptomatic and based on anticoagulants\textsuperscript{7}. Studies, however, demonstrate several complications with this approach, such as persistent pain, recurrent thrombosis events and restrictions on arm movements, which hampers patients' daily activities. Thrombolysis with continuous infusion of plasminogen activator is the most common therapy and with better results in such case. Success rates for restoring the permeability of the subclavian vein are of nearly 100\% as long as thrombolysis be completed within two weeks after the onset of symptoms.

The treatment of the arterial form of TOS will depend on the structure affected and the most effective treatment is surgical, although it is susceptible to complications\textsuperscript{5}. For patients with mild degrees of acute arterial ischemia due to embolization, catheter-directed thrombolysis may be appropriate prior to surgical treatment. However, the presence of more severe ischemia usually requires embolectomy (with or without intraoperative thrombolysis) in conjunction with thoracic outlet decompression.

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**Table 1.** Main differential diagnoses for thoracic outlet syndrome, according to Daniels et al.\textsuperscript{5}.

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<thead>
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<th>Vascular</th>
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<td>Vasculitis</td>
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<td>Vasospastic disorders</td>
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<td>Raynaud's syndrome</td>
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<td>Acute coronary syndrome</td>
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<td>Musculoskeletal</td>
<td>Impact syndrome</td>
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<td></td>
<td>Rotator cuff syndrome</td>
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<td>Adhesive capsulitis</td>
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<td>Neurological</td>
<td>Cervical radiculopathy</td>
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<td>Ulnar neuropathy</td>
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<td>Carpal tunnel syndrome</td>
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<td>Brachial plexitis</td>
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<td>Multiple sclerosis</td>
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<td>Others</td>
<td>Pancoast tumor</td>
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<td>Trigger points</td>
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<td></td>
<td>Fibromyalgia</td>
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<td>Complex regional pain syndrome</td>
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**Treatment**

The treatment of TOS has two objectives: to relieve patients' symptoms and to prevent complications\textsuperscript{6}. Kuhn et al.\textsuperscript{3} advocate that treatment should be initiated clinically and become surgical only in case of treatment failure.
Surgical treatment in general occurs in a minority of cases and is related to symptomatic bone abnormalities, vascular complications, trauma, sensory loss with increased pain, which can develop into numbness of the upper limb, and failure of conventional treatment. Surgical intervention consists of vascular and/or nerve decompression performed by various access techniques, such as transaxillary, supraclavicular, infraclavicular, and anterior and posterior thoracoplasty. The choice of how to proceed depends on the anatomical region affected and the preference for the surgeon.

The transaxillary approach is advantageous because it allows excellent exposure of the anterior part of the first rib, where compression occurs. The entire rib can be resected and its most medial portion can be completely disarticulated and removed from the manubrium. This incision is the most cosmetically acceptable but does not allow venous reconstruction. Daniels et al. advocate the supraclavicular approach, which provides a fuller ribs exposition. In addition, it allows the compression site to be directly identified and arterial reconstruction performed as needed. The anterior and middle scalene muscles can be completely resected with this access, and it is also possible to perform brachial plexus neurolysis. The infraclavicular surgical method is rarely used, only when a larger reconstruction of the TOS venous form is required.

**CONCLUSION**

Our review demonstrates that, in view of a suspected TOS, a detailed clinical and radiological semiology is necessary, aiming at the correct etiological diagnosis to evaluate the best therapeutic option for each case.

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