Vascular Thoracic Outlet Syndrome

Mohamad Anas Hussain, MD, *† Badr Aljabri, MD, FRCSC, ‡§ and Mohammed Al-Omran, MD, MSc, FRCSC *†‡§

Two distinct terms are used to describe vascular thoracic outlet syndrome (TOS) depending on which structure is predominantly affected: venous TOS (due to subclavian vein compression) and arterial TOS (due to subclavian artery compression). Although the venous and arterial subtypes of TOS affect only 3% and <1% of all TOS patients respectively, the diagnostic and management approaches to venous and arterial TOS have undergone considerable evolution due to the recent emergence of minimally invasive endovascular techniques such as catheter-directed arterial and venous thrombolysis, and balloon angioplasty. In this review, we discuss the anatomical factors, etiology, pathogenesis and clinical presentation of vascular TOS patients. In addition, we use the most up to date observational evidence available to provide a contemporary approach to the diagnosis and management of venous TOS and arterial TOS patients.

Semin Thoracic Surg 28:151–157 © 2016 Elsevier Inc. All rights reserved.

Keywords: Thoracic outlet syndrome, subclavian artery, subclavian vein, arterial thoracic outlet syndrome, venous thoracic outlet syndrome

OVERVIEW

TOS refers to a constellation of signs and symptoms attributable to compression of the neurovascular bundle in the thoracic outlet region of the upper extremity. A total of 3 distinct terms are used to describe TOS depending on which structure is compressed in the thoracic outlet. A total of 3 distinct terms are used to describe TOS depending on which structure is predominantly affected: neurogenic TOS (nTOS) from brachial plexus compression, venous TOS (vTOS) from subclavian vein compression, and arterial TOS (aTOS) from subclavian artery compression. The most common form by far is nTOS, which accounts for more than 90% of all TOS cases. This review focuses on the venous and arterial subtypes of TOS, which are seen in 3% and <1% of TOS patients, respectively. During the past decade, the approach to vTOS and aTOS has undergone considerable evolution due to the emergence of minimally invasive endovascular therapies.

ANATOMICAL FACTORS

The thoracic outlet area comprises 3 anatomic spaces: scalene triangle, costoclavicular space, and pectoralis minor (PM) space (Fig. 1). The borders of the anterior scalene muscle, middle scalene muscle, and first rib define the scalene triangle; trunks of the brachial plexus and the subclavian artery pass through this space (Fig. 1A). Cervical ribs and anomalous first ribs may compress the scalene triangle, resulting in symptoms of nTOS or aTOS. The costoclavicular space, which is the area between the first rib and the clavicle housing all 3 major structures (subclavian artery, vein, and brachial plexus), represents the most common site of subclavian vein compression (Fig. 1B). The PM space, defined by the PM muscle anteriorly and the chest wall posteriorly, is an extension of the thoracic outlet and a common site of neurovascular compression.

ETIOLOGY

Most patients with TOS are adults between the age of 20 and 50 years, although adolescents aged 18 years or younger present with vascular TOS more frequently than adults.1

1043-0679/$-see front matter © 2016 Elsevier Inc. All rights reserved.
http://dx.doi.org/10.1053/j.semtcvs.2015.10.008

Semin Thoracic Surg 28:151–157 © 2016 Elsevier Inc. All rights reserved.
Factors that increase the risk of neurovascular compression and development of symptomatic TOS include congenital anomalies in the thoracic outlet, repetitive motion or stress activities, and traumatic injuries. Arterial TOS is associated with bone abnormalities or trauma in nearly all cases, whereas vTOS is most often the result of repetitive overhead arm and shoulder activities, such as swimming, throwing, or weight-lifting in combination with anomalies of the costoclavicular space.

PATHOGENESIS
Arterial TOS is most commonly associated with bone abnormalities of the thoracic outlet, with cervical ribs being present in up to 85% of patients with aTOS in contemporary series. Cervical ribs are present in <1% of the general population, and about 70% of individuals with cervical ribs are women. Cervical ribs that cause symptoms are often large, and can have bony fusion to the first rib that results in subclavian artery compression. This causes injury to the third segment of the subclavian artery, leading to intimal damage, thrombosis, distal embolism or poststenotic dilation, and aneurysm formation. Rarely, complete occlusion of the subclavian artery may occur. Other less common anatomic abnormalities causing aTOS include anomalous first rib, prominent C7 transverse process, callus formation from an old clavicular or first rib fracture, and fibrocartilaginous band.

In vTOS, the subclavian vein is often compressed between the first rib, costoclavicular ligament and subclavius tendon within the costoclavicular space (Fig. 1B). Repetitive arm movements traumatize the vein, causing posttraumatic inflammation, focal intimal fibrosis, stenosis, blood flow stasis, and eventual thrombosis leading to acute symptoms of upper extremity deep venous thrombosis. This clinical condition is referred to as effort thrombosis or Paget-Schroetter syndrome, and is often observed in young individuals and competitive athletes who engage in physical activities requiring repetitive arm and shoulder movements. Chronic compression and repetitive trauma also cause inflammation external to the subclavian vein. This point is underscored by the fibrotic, hypertrophied and relatively fixed surrounding anatomic structures often observed during vTOS surgery.

CLINICAL PRESENTATION
Early in the disease process, aTOS patients most commonly present with chronic arm, shoulder, or neck pain with increasing activity due to subclavian artery stenosis or thrombosis. Hand or arm ischemia due to arterial embolization is the most common acute presentation of aTOS. A subset of aTOS patients present with concomitant symptoms of nTOS, such as chronic arm or hand paresthesias, numbness, or weakness from coexisting brachial nerve compression. Physical examination for aTOS focuses on measuring bilateral blood pressures of the upper extremities to assess for a marked discrepancy between the symptomatic and asymptomatic side, palpating for cervical ribs or a pulsatile mass in the
supraclavicular area, examining hands for signs of digital ischemia, and auscultating for bruits in the supraclavicular fossa. Moreover, narrowing the artery through shoulder abduction may provoke bruits if not present at rest.

The classic clinical finding of vTOS is upper extremity edema with dilated subcutaneous collateral veins evident over the arm, shoulder, and chest wall. These collateral veins develop to accommodate the increased venous pressure from subclavian vein stenosis or thrombosis. In the largest series of vTOS to date (626 extremities), Urschel and Patel9 reported that vTOS patients most commonly present with visible collateral veins around the shoulder (99%), followed by arm swelling (96%), bluish discoloration (94%), aching pain with exercise (33%), and cervical ribs (10%). Only 4% of the patients presented with minimal symptoms. Severe complications of acute deep venous thrombosis, such as pulmonary embolism or venous gangrene, are rare.

DIAGNOSIS

Plain radiography of the neck and chest with cervical spine view or thoracic inlet view will often demonstrate bone abnormalities that may aid in the diagnosis of TOS. Cervical ribs, prominent, or elongated C7 transverse processes, abnormal or elongated first ribs, and large fracture calluses are easily detected on plain radiographs. In a study of 31 patients with TOS, only 2 had normal plain radiographs, whereas the other 29 had abnormalities, such as cervical ribs and enlarged transverse processes detectable on plain radiographs.10 However, the negative predictive value of plain radiography is debatable, and more definitive diagnostic imaging is required in nearly all cases to establish or exclude a diagnosis of TOS.

Duplex ultrasound (DUS) examination is an effective initial approach in confirming clinical suspicion of vascular TOS. Aneurysmal change, arterial stenosis, and thrombosis can be detected by DUS of the subclavian and axillary arteries. Moreover, DUS has the advantage of assessing dynamic blood flow during compression maneuvers (hyper-abduction), with a decrease in arterial diameter, changes in peak velocity or reproducible symptoms considered to be diagnostic of aTOS. Figure 2 shows an arterial DUS of the upper extremity with normal flow in the left brachial artery at rest, and complete cessation of flow with 45° of arm abduction above the shoulder—a classic finding in aTOS. DUS also has good accuracy in detecting subclavian-axillary vein thrombosis, although its use is limited in providing information about surrounding structures or preoperative planning.

Conventional catheter-based arteriography or venography has traditionally been considered the gold standard diagnostic test for vascular TOS as it provides information about the exact location and nature of the vascular compression. However, because of its invasive nature, and lack of visualization of surrounding structures means catheter-based angiography and venography are largely reserved for intraprocedural interventional guidance.

Contrast-enhanced computed tomography (CT) and magnetic resonance (MR) are now widely available, reliable, and reproducible modalities used to establish the diagnosis of vascular TOS. CT and MR are typically performed as 2-step procedures (neutral position and arm abduction) to reproduce the vascular compression seen on provocative maneuvers. CT angiography or venography provides superior analysis of the vasculature in relation to the bony structures, whereas MR angiography or venography is more efficient in the depiction of accessory muscles, muscle hypertrophy and fibrous bands.11 Both of these imaging modalities are useful in delineating the exact location and nature of the

**Figure 2.** Duplex arterial ultrasound with waveforms of a 21-year-old patient with aTOS showing normal flow in the left subclavian artery at rest (A), and complete cessation of flow with arm abduction 45 degrees above the shoulder (B).
vascular compression, and aid in surgical planning. The American College of Radiology Appropriateness Criteria recommendations suggest that CT and MR angiography or venography are both appropriate in establishing the diagnosis of TOS; therefore, which imaging modality is selected likely depends on clinician and institutional experience and expertise. Figure 3 presents a CT angiogram showing moderate impingement of the left subclavian artery by a cervical rib with poststenotic dilatation, and distal embolization to the left brachial artery in a patient with an acute ischemic arm secondary to aTOS.

MANAGEMENT

Arterial TOS

The management strategy for aTOS is guided by the nature and severity of arterial complications. Asymptomatic patients with subclavian artery compression without evidence of arterial degeneration may be managed nonsurgically due to a low risk of complications. It is reasonable to follow these patients with serial imaging of the arterial system, as the natural history of these patients is not clearly defined. We prefer to perform arterial DUS of the upper extremity every 6 months in these patients, although it is unclear if this practice leads to earlier diagnosis of arterial complications. Surgical treatment is required for patients with symptoms and evidence of arterial complications, such as intimal damage, mural thrombus, embolization, poststenotic dilatation, or aneurysm formation. The appropriate surgical strategy is guided by 3 main principles: decompression, arterial resection, and distal revascularization.

1. Decompression: At minimum, relieving the arterial compression involves resection of cervical or first ribs, fibrous bands, scalenectomy, and removing any other associated anomalies. Some authors suggest that the first rib should be routinely removed to prevent recurrence of symptoms because it acts as a key insertion point for fibromuscular bands that cause vascular compression. Others argue that anterior and middle scalenectomy alone is as effective as scalenectomy with first rib resection, and that a rib-sparing approach leads to less morbidity, including a lower risk of pleural or plexus injury and shorter hospital stay. However, the evidence for this is limited, and largely focused on patients with a past history of neurogenic TOS. Consequently, for now, one approach cannot be recommended over another.

2. Arterial resection: Resection of any potential source of arterial embolus such as a subclavian artery aneurysm or luminal stenosis with intimal damage is necessary to prevent ischemic complications of the upper extremity.

3. Distal revascularization: Vascular reconstruction in the form of primary anastomosis, interposition graft, or axillary-brachial bypass may be required depending on the extent of the subclavian artery resection. If evidence of distal embolus is present, intraarterial thrombolysis or thromboembolec-tomy may be used in conjunction with arterial reconstruction to improve outflow of the limb.

There are 2 main surgical approaches to thoracic outlet decompression: transaxillary and supraclavicular. The primary advantage of the transaxillary approach is that it provides complete visualization of the first rib for removal, and poses minimal risk to critical neurovascular structures that are distant from the first rib. However, this approach is largely used for nTOS, as it is not suitable for vascular reconstruction. The supraclavicular approach is preferable for aTOS as it allows resection of the cervical and first ribs, fibromuscular structures, and vascular reconstruction. However, care must be taken to identify and protect the brachial plexus and other...
neurovascular structures, such as the phrenic and long thoracic nerves, and the subclavian vein with this approach. The supraclavicular exposure of the thoracic outlet is illustrated in Figure 4.

Contemporary series report negligible operative mortality and a complete resolution of aTOS symptoms in >90% of patients. One of the largest and most recent series, which examined 49 cases of aTOS, showed that operative morbidity in this population is low (13%), and mainly related to transient nerve injury to the brachial plexus that resolves completely with conservative management. Other potential complications include arterial rethrombosis, hemothorax, pneumothorax, or chylous leak.

Although open surgical revascularization is considered the gold standard approach to aTOS, some case studies have described endovascular stenting of the subclavian artery in combination with surgical decompression. The advantage of this hybrid technique is a less invasive approach to subclavian artery revascularization; however, this must be weighed against a higher risk of stent fracture at this location due to compression from external structures such as the clavicle and first rib. More long-term data is needed before subclavian artery stenting can be considered a valid alternative to surgical revascularization. Intraarterial thrombolysis for a distal embolus or thrombus is another treatment option, particularly if the upper limb is acutely threatened from severe ischemia. Thrombolysis must be followed by thoracic outlet decompression and arterial repair to remove the source of embolus such as subclavian artery stenosis or aneurysm. Regardless of which arterial revascularization technique is selected, surgical decompression is fundamental to thoracic outlet treatment.

**Venous TOS**

The natural history of acute primary subclavian-axillary vein thrombosis is associated with high long-term morbidity and disability. Furthermore, historical series have reported persistent or recurrent symptoms in >70% of vTOS patients treated with anticoagulation alone. Therefore, treatment for vTOS involves consideration of 3 therapies in addition to anticoagulation: thrombolysis, decompression, and venoplasty. Which therapy is selected depends on the clinical presentation of patients with vTOS. Several contemporary series have suggested various treatment algorithms for vTOS based on their local experience. Taken as a whole, the approach to vTOS management can be summarized based on 3 patient presentations (Fig. 5).

1. **Acute:** For patients presenting acutely (<6 weeks since symptom onset), prompt catheter-directed thrombolysis with early surgical decompression of the thoracic outlet has been reported to produce >90% clinical success. A short course of anticoagulation may be required until venous patency is confirmed by postoperative imaging. Venoplasty of a residual subclavian vein stenosis after decompression may be considered to reduce the risk of rethrombosis.

2. **Chronic:** Patients presenting with chronic (≥6 weeks) stenosis or occlusion of the subclavian vein with evidence of vTOS benefit from surgical decompression. Preoperative thrombolysis may be considered in patients with total occlusion in an attempt to restore luminal patency before decompression. A course of postoperative anticoagulation may be required until venous patency is confirmed by postoperative imaging. Venoplasty of a residual subclavian vein stenosis may be considered to reduce the risk of rethrombosis.

3. **Intermittent obstruction:** Patients with chronic symptoms of intermittent venous obstruction but without evidence of thrombus or significant stenosis require only surgical decompression. Thrombolysis, anticoagulation, or venoplasty are generally not required.

Infraclavicular, paraclavicular, and transaxillary approaches have all been well described for subclavian vein decompression. Which approach is selected often depends on operator experience and preference, as there is insufficient evidence to recommend one.
Operative principles are similar to aTOS—first rib resection with scalenectomy to decompress the thoracic outlet, and venolysis. Given the most common site of venous compression is in the costoclavicular space, first rib resection is generally preformed routinely in vTOS patients in combination with scalenectomy. Furthermore, routine first rib resection has been associated with improved quality of life in patients with vTOS. A few studies have examined the role of venous angioplasty and stenting before decompressing the thoracic outlet, with suboptimal results. Venous stenting in a nondecompressed costoclavicular junction is associated with high rates of stent fracture and rethrombosis, likely due to external compression of the stent by the unyielding structures of the thoracic outlet. Therefore, primary stenting of the subclavian vein without thoracic outlet decompression is not indicated.

Residual stenosis of the subclavian-axillary vein is not uncommon following decompression due to fibrous strictures or thrombus; options for management include intraoperative or postoperative venography and angioplasty with or without stent insertion, vein patching, mechanical thrombectomy, or axillary-subclavian vein reconstruction. Although no study has directly compared outcomes between these different approaches, most authors prefer to perform venoplasty at the time of decompression or within 1-2 weeks postoperatively of any residual stenosis because of its minimally invasive nature and excellent reported 1-year primary and secondary patency rates of 92% and 96%. Venous DUS is an efficient noninvasive imaging modality for routine postoperative surveillance, although a venogram may be preformed at the first postoperative visit if residual venous stenosis is suspected and venoplasty is considered. With respect to anticoagulation, a short course of 3-6 months may be required if there is residual thrombus on postoperative imaging. Venous TOS series have generally only reported warfarin as the anticoagulation agent of choice, although alternative agents such as oral factor Xa inhibitors may be considered given their efficacy in the treatment of venous thrombosis.

CONCLUSION

Although uncommon, vascular TOS presents diagnostic and management challenges to the modern cardiovascular surgeon. Clinical evidence for the approach to vascular TOS is limited to case series, with a paucity of randomized controlled trials and prospective data. However, over the past decade, the continual emergence of
technological leaps in the form of endovascular techniques such as catheter-directed arterial and venous thrombolysis, and balloon angioplasty has considerably diversified the contemporary approach to vascular TOS. Further research is needed to define the role of stenting in vascular TOS.

3. Criado E, Berguer R, Green