Venous and Arterial Thoracic Outlet Syndrome Requiring Surgical Intervention: Two Case Reports

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\textbf{ABSTRACT}

\textbf{Background:} Thoracic outlet syndrome represents a spectrum of diseases depending on the structures involved. Most cases of thoracic outlet syndrome (TOS) are neurogenic while arterial and venous TOS constitute only 5\% of the cases. Paget-Schroetter syndrome represents an acute complication of venous thoracic outlet syndrome (vTOS) commonly occurring in physically active individuals participating in strenuous upper body exercises. Arterial thoracic outlet syndrome (aTOS) is usually associated with bony abnormalities. We discuss one patient of each type who presented with acute thrombosis to illustrate the different associated factors and treatment options.

\textbf{Case Report(s):} An 18-year-old Hispanic male presented with six days of left upper extremity (LUE) edema and pain that began following an upper body exercise. He was treated by pharmacomechanical thrombectomy followed by ultrasound assisted catheter-directed thrombolysis. Subsequently the patient underwent trans axillary surgical decompression of the thoracic outlet.

A 59-year-old South Asian woman presented with worsening LUE pain, weakness and paresthesia of 10 days duration. She underwent pharmacomechanical thrombectomy with angioplasty followed by supraclavicular thoracic outlet decompression.

\textbf{Conclusions:} Treatment of TOS presenting with thrombosis should consist of prompt thrombectomy followed by surgical decompression.

\textbf{Keywords} Venous thoracic outlet Syndrome, Paget–Schroetter syndrome, Arterial thoracic outlet syndrome, Mechanical thrombectomy, Thrombolysis, Thoracic outlet decompression, Supra clavicular approach, Transaxillary approach.

\textbf{Background} Thoracic outlet syndrome (TOS) is estimated to have an incidence 3–8/1000 \cite{1,2}. An overwhelming majority of cases are neurogenic TOS (95\%) with the remaining, 3-5\% venous and 1-2\% arterial \cite{3}. We present one case of venous and one of arterial...
TOS who presented to us within the space of a few months, the discussion of which will help to contrast the different demography, predisposing factors and surgical approach.

Venous thoracic outlet syndrome (vTOS) presents as occlusive venous symptoms following compression of the subclavian vein as it passes over the anterior scalene tendon and between the first rib and costoclavicular ligament. In an acute setting, vTOS may present as Paget–Schroetter syndrome (PSS), a complication involving acute venous thrombosis of the axillo-subclavian vein [4-6]. PSS presents an “effort-induced” thrombosis of the axillo-subclavian vein, often occurring in physically active individuals following repeated or strenuous use of the arm and shoulder [4,7-9]. These repeated cycles of compression-decompression likely damage the venous wall, causing luminal stenosis and eventual thrombosis presenting as acute upper extremity swelling, cyanosis, heaviness, and ultimately pain [2,6,10].

Due to the rarity of the condition, a paucity of literature exists about treatment and intervention guidelines, complications, and associated outcomes for management of PSS [6]. As a result, variation in healthcare provider treatment of PSS persists. As the threshold for decompression varies widely from mild to moderate symptoms, providers commonly reserve surgery for TOS to patients who failed conservative management [2]. In a recent study of 123 cases, management of PSS included first rib resection combined with catheter-directed thrombolysis (CDT) and anticoagulation in 60.1% of cases; 17% of cases underwent an unspecified surgical intervention; 5.7% of cases received balloon angioplasty or CDT and anticoagulation, without rib resection, 13% received only therapeutic anticoagulation, and 4.1% received an unspecified treatment [11]. Within the same study, 26.7% of subjects reported complications, with the most common being pulmonary embolism (8.2%) followed by re-thrombosis (5.7%), neuropathy (3.3%), persistent pain (1.6%), hematoma (1.6%), hemothorax (1.6%), and post-thrombotic syndrome (1.6%) [11]. Furthermore, the risk of post-thrombotic syndrome significantly increases morbidity and presents as pain, heaviness and swelling of the affected extremity and may present as a chronic condition [12-14].

The ischemic events of arterial thoracic outlet syndrome (aTOS) likely occur as a result of repetitive cycles of compression-decompression causing to damage to the endothelium, ultimately resulting in partial or complete thrombosis, aneurysmal degeneration, stenosis with post stenotic dilation, and post stenotic aneurysms [15,16]. Arterial embolization represents the most common presentation for aTOS [16,17]. Patients with aTOS may initially present with signs and symptoms of claudication in the affected extremity [16,18,19]. Several clinicians postulate this phenomenon occurs due to chronic changes to the intima of the subclavian/axillary artery [16,17]. Abnormalities associated with aTOS include cervical ribs, abnormal first rib, elongated transverse process of C7, and congenital and acquired soft tissue problems [16]. Novel diagnostic tools such dynamic computed tomography angiography (CTA), Magnetic Resonance (MR) neurography, and Diffusion Tensor Imaging (DTI) may aid in the diagnosis of TOS [2,20].

Treatment varies depending on the anatomical structures involved. However, clinicians recommend immediate surgery for acute arterial insufficiency [16,21]. Moreover, several studies exist documenting conservative management of thoracic outlet syndrome yet none detail conservative management for aTOS specifically [16,22]. Surgical intervention for aTOS typically involves removal of abnormal ribs or fibrous bands [23,24]. Another study, however, recommends using initial catheter-directed thrombolysis (CDT) in cases of subclavian artery involvement and then following up with surgical decompression [16]. In cases of arterial degeneration, dilation, or aneurysm, authors recommend subclavian artery repair in order to prevent upper extremity ischemia [24,25].

To gain a deeper understanding of the treatment of TOS, we present two cases, an 18-year-old male with left-sided PSS and a 59 year-old female with left-sided aTOS. Both patients had positive outcomes following an endovascular approach with subsequent open thoracic outlet decompression and review of current literature.

Case Presentations

Case 1
An otherwise healthy 18-year-old Hispanic male presented with six days of left upper extremity (LUE) edema and pain that began following a session of pushups. Additionally, the patient complained of LUE paresthesia. A LUE Duplex ultrasound showed a deep vein thrombosis (DVT) in the left subclavian vein extending to the left brachial and left basilic veins (Figure 1). The laboratory values were unremarkable. Thrombophilia screening was negative. The patient was anticoagulated with heparin. On hospital day two, he underwent a LUE venography and pharmacomechanical thrombectomy using Angiojet followed by ultrasound-assisted catheter directed thrombolysis (EKOS), using tissue plasminogen activator (tPA) (Figures 2). An intravascular ultrasound showed a tight compression at the thoracic outlet. The patient underwent a transaxillary anterior scalenectomy and first rib resection two days later. He was kept on coumadin for 6 months, and received two subsequent ultrasound scans, which showed complete resolution of the clot (Figure 3). He regained full range of movements of his shoulder and the anticoagulation was stopped. The arm swelling resolved completely.

Case 2
A 59-year-old South Asian female with past medical history of hypertension, hyperlipidemia, hyperthyroidism and an unprovoked right lower extremity deep vein thrombosis diagnosed one year prior, presented to the emergency department with gradually worsening LUE pain, weakness and paresthesia of 10 days duration. She had undergone physical therapy for nonspecific arm pain, which included cupping. CTA of the LUE revealed a focal short segment stenosis/ occlusion of the mid left subclavian artery (Figures 4 and 5) and a complete occlusion of the proximal axillary artery through to mid brachial artery with weak distal reconstitution. In addition, there was a large area of callus from a previous malunited mid clavicular fracture on the same side.
Figure 1: Duplex ultrasound showing thrombosis of left axillary and brachial veins

Figure 2: Venogram and mechanical thrombectomy in progress of the thrombosed veins

Figure 3: Follow up Duplex scan 6 months later showing patency of the left subclavian vein indicating resolution of the clot
The patient had unremarkable laboratory studies. Thrombophilia screening was negative.

**Figure 4:** Sagittal and axial reconstruction of CT scan showing compression of the left subclavian artery, wedged between the anterior scalene muscle and the first rib.

**Figure 5:** 3D reformat of the CT scan showing narrowing of the left subclavian artery. Note the ipsilateral malunited clavicle fracture.

The patient was heparinized and subsequently underwent a left brachial angiogram, which showed complete occlusion of the left subclavian, axillary, and brachial arteries. A pharmacomechanical thrombectomy with t-PA followed by angioplasty was performed (Figure 6) followed by anticoagulation with rivaroxaban. Postoperative duplex ultrasound revealed a tight stenosis of the proximal subclavian artery (Figure 7), and some residual thrombus in the brachial artery. The patient subsequently underwent thoracic outlet decompression via a supraclavicular approach, with resection of the first rib and the scalenus anterior. An interval transradial angiogram was performed with angioplasty of the distal brachial artery. The patient is totally asymptomatic now and is maintained on long term aspirin – rivaroxaban combination, in view of the previous DVT.

**Figure 6:** Transbrachial arteriogram and mechanical thrombectomy in progress. Note the intraluminal lucencies compatible with clot formation.

**Figure 7:** Post thrombectomy Duplex scan showing persistent compression of the proximal subclavian artery at the thoracic outlet.

**Discussion**

The majority of literature regarding PSS consists of isolated case reports (Table 1). Many authors practice protocol-driven, early CDT, followed by either immediate or staged surgical decompression and further adjunctive endovascular or open interventions if necessary. The multiple stages of therapy involves a complex decision process. There is no current consensus on guidelines describing the optimal management of this disease [6].

Table 1 displays various case presentations, mechanisms of injury, treatments, and outcomes demonstrating the inconsistencies in management of PSS. While some patients received conservative management, many, like our case, required aggressive endovascular and open surgical interventions. Some studies suggest that exercise is preventative; others state that high-intensity exercise may actually increase thrombogenic risk [36]. The authors postulate that the elevated levels of plasma catecholamines and lactic acid occurring during high intensity exercises significantly increase platelet aggregability [35,36]. Other suggested factors predisposing athletes to thrombosis include dehydration, endothelial injury from physical strain or trauma, immobility because of injury, long distance travel to competitions, and oral contraceptive use [35,37].
One study suggested that the bradycardia seen in athletes with a high level of cardiopulmonary fitness might even contribute to venous stasis [38].

Previous studies show that the treatment of PSS with anticoagulation alone leads to a higher incidence of residual symptoms, disability, and recurrent thrombosis [7,39]. Systemic fibrinolysis demonstrated greater efficacy than anticoagulation alone in attaining venous patency, but was accompanied by serious complications such as intracranial hemorrhage [7]. As a result, catheter-directed thrombolysis is more commonly used due to controlled, local fibrinolytic effects with minimal systemic bleeding risk [7]. Several studies suggest that initiation of catheter directed thrombolysis within 14 hours of presentation yields better clinical outcomes [7,39]. Researchers propose that fibrin clots formed longer than 14 hours exhibit greater strength and stability due to covalent linkage with Factor XIII [7,14,39-41]. One study reported success rates of 90-95% in patients who received rapid intervention with immediate decompression [14,39]. Other studies report early catheter-directed thrombolysis success rates ranging between 75–84% [14,42,43]. Another reported a 29% success rate of thrombolysis within 2–12 weeks following onset of symptoms [14,44]. However, some argue in favor of conservative, non-surgical interventions [9,27,30,32,35]. Both of our patients presented relatively late but still had successful outcomes.

Currently, no consensus exists for the duration of anticoagulation for PSS. The 2016 CHEST Guideline and Expert Panel Report recommends a 3-month course of therapy with new oral anticoagulants (NOACs) over vitamin K antagonists following any upper extremity deep vein thrombosis (DVT), regardless of thrombolytic interventions [14,45]. Others suggest using post-operative venography. Patients with adequate venous patency require no further intervention or anticoagulation. Despite initial relief of symptoms following intervention, re-thrombosis may occur in up to one-third of patients [14,46]. Therefore, some authors recommend a more definitive treatment with thoracic outlet decompression in patients who are good surgical candidates [14,47,48]. In the absence of specific anatomic abnormalities, surgeons achieve decompression with first rib resection via a transaxillary, supraclavicular or infraclavicular approach. If indicated, partial resection of the subclavius and anterior scalene muscle helps debulk the thoracic outlet and prevent recurrence of symptoms [14,47]. In cases of persistent stenosis or re-thrombosis,
patients should receive anticoagulation and repeat duplex ultrasound examination for 6 months [3,14,49,50]. Additional measures such as surgical thrombectomy, balloon venoplasty, and stenting showed poor success rates and high morbidity when used for persistent stenosis or re-thrombosis [14,51-53].

aTOS may occur asymptptomatically in the form of aneurysm, occlusion, or silent embolization or symptomatically as ischemia following embolization [54,55]. Less commonly, aTOS presents as an asymptomatic neck mass or as an incidental finding on imaging [55,56]. As a result of its rarity, aTOS is often misdiagnosed during initial presentation and may take greater than 6 months from symptom onset to accurately diagnose [55,57]. This delay frequently results in chronic and repeated embolization with symptoms of acute limb ischemia such as pain, weakness, pallor, and paresthesia [17,55]. In advanced disease, patients may present with rest pain and ischemic ulceration [55,56].

The results of a retrospective study with 41 patients showed subclavian artery stenosis with or without post stenotic dilatation in 26 (63%) of patients, subclavian artery aneurysms in 12 (29%), axillary artery compression in 2 (5%), and chronic subclavian artery occlusion in 1 (2.4%). However, in absence of obvious ischemia, clinicians may not suspect significant arterial insult resulting in underestimation of the incidence of arterial pathology secondary to compression of the thoracic outlet. Furthermore, as two-thirds of patients possess associated bone abnormalities, researchers suggest routine arterial imaging for patients evaluated for TOS. Proper understanding of the incidence, natural history, and clinical relevance of arterial compression and post stenotic dilatation occurring in TOS warrants further studies [17].

In patients with suspected aTOS, catheter-based angiography is the “gold standard” as the procedure is both diagnostic and therapeutic, allowing for direct therapy and planning for potential reconstruction or bypass [55,58]. Research suggests use of arterial ultrasound in patients referred for asymptomatic incidental cervical rib(s) for proper evaluation of subclavian artery stenosis or aneurysm and recommends duplex ultrasound with extremity manipulation to assess for compression during dynamic range of motion [55]. Young active patients may warrant annual or biannual follow-up duplex ultrasound to reassess for aneurysm [55]. Patients presenting with symptoms of neurogenic or arterial compression such as discoloration, claudication, or other signs of acute limb ischemia require further evaluation with CTA [55]. CTA, MR angiography, or traditional angiography should extend to the digits to properly evaluate these patients [55].

In patients with subclavian artery compression and aneurysm without significant distal ischemia, TOS decompression via a supraclavicular approach is recommended [55]. Excision of the cervical rib, the fibrous attachment, and the anterior scalene muscle serves as adequate treatment for most patients undergoing aTOS decompression. Mildly dilated subclavian arteries may be followed with serial imaging as most affected arteries remain unchanged or return to their normal size [55]. However, subclavian arteries with significant aneurysmal dilation, intramural thrombus, or history of thromboembolic disease need to be replaced. In these cases, first rib resection is also recommended to eliminate dynamic compression of the bypass graft [55].

Patients presenting with acute limb ischemia secondary to thromboembolism in absence of usual risk factors such as arrhythmia, atherosclerosis, or trauma, warrant evaluation for aTOS [55]. Severe limb ischemia often indicates emergent open thrombectomy via brachial artery cut down. In patients with mild acute ischemia and distal thrombus, CDT may be considered [55]. This was the case in our aTOS patient, for which we staged our thoracic outlet decompression. In cases of aneurysm, one study recommends therapeutic anticoagulation following thromboembolectomy and subsequent resection and bypass in the same hospital admission [55].

**Conclusion**

The current cases highlight the need for surgical intervention for acute presentation of TOS. Treatment of TOS presenting with thrombosis should consist of removing the thrombus, decompressing the thoracic outlet, and preventing recurrence. This may be achieved through early diagnosis, rapid initiation of systemic anticoagulation followed by thrombectomy/thrombolysis and surgical decompression.

**Authors’ Contributions:**

S. Mahmoudzadeh, A. Parrill, D. Shaikh, K. McGhee, S. Lev, and A. Ramanathan: study design, data acquisition, data analysis, and drafted manuscript. All authors approved the final version of the manuscript.

**References**


