Venous Thoracic Outlet Syndrome: A Short Review

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Abstract

Venous thoracic outlet obstruction can be either primary or secondary. Effort-induced thrombosis of the upper limb ranges from 1% to 4% of all venous thrombosis has been the focus of discussion in many an international and national journal recently because of an increase in the number of cases being seen, diagnosed, and treated. In this paper, we discuss an overview of the problem and our management approach based on available evidence and share the experience gained from treating patients with effort-induced axillary-subclavian thrombosis or Paget–von Schroetter syndrome, as it is otherwise called.

Keywords: Paget–Schroetter syndrome, primary effort thrombosis, upper extremity deep vein thrombosis, venous thoracic outlet syndrome

INTRODUCTION

Thoracic outlet syndrome (TOS) remains an enigma. Recently, the Journal of Vascular Surgery published an article on “Reporting Standards of Society for Vascular Surgery for TOS,”[1] which would require those seeing patients with this condition to perform a more detailed examination and record the findings in a more systematic manner before labeling a patient to be suffering from any of the forms of TOS.

In 1875, Sir James Paget drew attention to a “gouty phlebitis” of the upper limb. In 1884, von Schroetter described upper limb deep vein thrombosis (DVT) to be secondary to direct damage of the vein caused by its stretching from muscular strain. In 1948, Hughes, an English Surgeon, gave the term Paget–von Schroetter. Thereafter, venous TOS (VTOS) has had many an eponym.[2-5]

To better understand VTOS, one needs to revisit the anatomic spaces. The brachial plexus and subclavian artery pass through a triangle formed by the scalenus anticus and medius muscles along with the first rib. The subclavian vein, on the other hand, enters the chest wall more anteriorly, passing close to junction of the clavicle, the first rib, and the subclavius muscle and tendon [Figure 1].

VTOS has been described as being of three types: intermittent/positional venous obstruction also called as McCleery syndrome, secondary subclavian vein thrombosis (postcentral venous catheters or pacemaker lead insertion), and primary “effort thrombosis” (Paget–Schroetter syndrome).[1,5]

There are no randomized control trials (RCTs) that have been conducted and one has to rely on review articles, expert opinions, case series, and book chapters to plan treatment.

At the time of writing this article, there is no evidence in English literature that discusses VTOS – from the Indian subcontinent.

The number of cases being diagnosed has seen a rise owing to increased awareness of the problem among treating doctors, besides increase in the use of central venous catheters and availability of interventional radiology suites and qualified personnel.

EPIDEMIOLOGY

Effort-induced thrombosis of the upper limb ranges from 1%–4% of all venous thrombosis. In the United States, the incidence is about 3000–6000 cases yearly. Some cases are being missed because of lack of awareness among the nonspecialists. The age group seems to be predominantly

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patients in their 30s, with a male-to-female ratio of 2:1. The right arm is involved in 60%–80% of cases and is more in those who do vigorous exercise such as swimming or use of the involved upper limb for activity related to their profession, for example, automechanics and painters.\[^{[5-7]}\]

In a study published by Christian Medical College, Vellore, in 2006, the incidence of VTOS was 3% of all diagnosed TOS which is in keeping with the published literature.\[^{[8]}\]

Between January 2010 and March 2017, we have had 10 cases. The average age of presentation was 35.8 years with male preponderance (1.5:1) [Table 1].

**Etiology**

As shown in Figure 1, the anterior part of the thoracic outlet, where the subclavian vein passes between the intersection of the clavicle and first rib, is the anatomical space that explains VTOS. The area posterior to this region is open – the scalenus anterior lies posterior to the vein. A hypertrophied scalene can, however, compress the vein posteriorly. The subclavius muscle provides bulk at the costoclavicular junction. The first rib and clavicle do not move much; however, in extremes of leverage, the vein gets maximally compressed. Adams and DeWeese demonstrated that the vein could be compressed even in the normal population. A cervical rib is usually posterior and does not cause venous compression.\[^{[5,9]}\]

Regardless of which of the structure is abnormal, the subclavian vein can be compressed by muscle or bone or a combination of both. Chronic compression and trauma are thought to lead to external inflammation, causing fibrosis and fixation of the subclavian vein to surrounding muscle and/or bone with perhaps intimal hyperplasia as well. During surgery, this is seen as a dense collagen scar. Whenever the costoclavicular space changes in diameter, the vein is stretched or torn. If it has been a chronic process, then, at imaging, “first pass collaterals” are seen connecting cephalic vein and profunda branches with the transverse cervical, scapular, and external and internal jugular veins.\[^{[5,9,10]}\]

In 2012, Magee et al. described the presence of “Langer’s axillary arch,” which is an accessory muscular band between the latissimus dorsi and pectoralis major. It has an incidence of 8% among young adults and could contribute to VTOS.\[^{[11]}\]

There is a subset of patients in whom an etiology cannot be found. This group on follow-up is known to be associated with developing lung cancer at 1-year follow-up with a Pancoast tumor bring the most common lesion.\[^{[4]}\]

**Symptoms**

Patient can present with symptoms of episodic arm discoloration and swelling brought on by exercise or arm elevation. Or they can present acutely with swollen arm. Usually, 60%–80% of

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**Table 1: Venous thoracic outlet obstruction Christian Medical College experience: January 2010-March 2017**

| Sex/age  | Occupation   | Side (CR) | Symptoms and duration (days) | Intervention | Decompression (D) | Return to work | Follow-up (weeks) |
|----------|--------------|-----------|-------------------------------|--------------|-------------------|----------------|------------------|
| Male/22  | Wood cutter  | Right     | Acute DVT, 14                | CDT + anticoagulation | No                | Yes            | 12               |
| Female/46| Homemaker    | Left      | Acute DVT, 2                 | CDT/stent + anticoagulation | No                | Yes            | 24 occlusion     |
| Male/38  | Manual laborer| Left      | Arm swelling, chronic DVT, 62| Anticoagulation | No                | Lost to follow-up |
| Male/43  | Manual laborer| Right     | Arm swelling, chronic DVT, 21| Anticoagulation | TA FRR           | Yes            | 24               |
| Male/38  | Mechanic     | Left      | Arm swelling, chronic DVT, 34| Anticoagulation | No                | Yes, part-time | 4                |
| Female/29| Manual laborer| Left CR  | Arm swelling, chronic DVT, 40| Anticoagulation | Supraclav CR      | Yes            | 24               |
| Female/38| Homemaker    | Right CR  | Arm swelling, chronic DVT, 30| Anticoagulation | Planned           | Lost to follow-up |
| Male/35  | Jewelry maker | Right     | Arm swelling, chronic DVT, 150| Anticoagulation | Planned           | Yes            | 6                |
| Male/33  | Teacher      | Left CR   | Arm swelling, 45             | Antiplatelet  | Supraclav CR      | Yes            | 52               |
| Female/36| Teacher      | Left CR   | Arm swelling, 60             | Antiplatelet  | TA FRR/CR         | Yes            | 24               |

CR: Cervical rib, TA FRR: Transaxillary first rib resection, TA FRR/CR: Transaxillary first rib resection and cervical rib excision, DVT: Deep vein thrombosis, Supraclav: Supraclavicular

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**Figure 1:** Costoclavicular space where the subclavian vein compression occurs
these patients will give a history of exertion before the onset of symptoms. A few patients present with dilated veins over the shoulder, neck, or anterior chest due to chronic occlusion of axillary-subclavian thrombosis (ASVT).[5,7,12]

In our series, six had presented with chronic DVT, two had acute DVT presentation, and two had venous edema [Figure 2]. Four of the patients had cervical rib at diagnosis [Table 1].

**Diagnosis**

The standard of care for diagnosis at presentation is a venous duplex. This imaging modality has high accuracy in experienced hands with sensitivity of 78%–100% and specificity of 82%–100%.[13] In our series, all the patients had venous duplex examination.

Positional venogram can be done in a patient who presents with episodic symptoms to look for vein compression. However, in effort thrombosis, the vein is usually thrombosed, and hence, examining the arm in neutral position is sufficient. There could be the presence of first pass collaterals. The American College of Radiology recommends a duplex as the best first approach while using a venogram when other noninvasive studies are inconclusive. Magnetic resonance and computed tomographic venography have proved to be disappointing in showing the region of the costoclavicular junction well. However, they do help visualize the central veins well.[5,14] [Figures 3 and 4].

The role of a thrombotic workup in this subset of patients is not clearly defined. A Swedish study published in 1988 found hypercoagulable states to be infrequent in these patients. A more recent study suggests otherwise. It states that a hypercoagulable state could be present in as many as 67% of patients.[6,15]

**Treatment Options**

As early as 1949, it was reported that if left untreated, it leads to significant morbidity.[4]

The aim of treatment is to prevent thrombus progression, reduce thrombus burden, and prevent early recurrence and Pulmonary Embolism. For want of large case series or RCTs, treatment options are based on evidence gained from treating lower limb DVT.

In patients who present with episodic symptoms and no gross anatomic abnormality, life style modification and physiotherapy help. The former includes weight loss, less vigorous movements, and postural changes. Structured physiotherapy helps break down any adhesions, increase range of movements, and reduce pain and edema.[5,16,17]

Those who present acutely would need limb elevation, rest, and anticoagulation till imaging done helps finalize a plan of management by anticoagulation or catheter-directed thrombolysis (CDT).

**Anticoagulation**

The 2016 ACCP guidelines recommend the use of anticoagulation over thrombolytic therapy in patients with upper extremity DVT [Grade 2C]. Once admitted, patients can be started on unfractioned heparin or low molecular weight heparin. This helps keep the collaterals open and prevents propagation of the thrombus. Warfarin therapy should then be initiated for 6–9 months, and extended, based on the assessment that includes recanalization and resolution of symptoms.[5,18]

In our case series, four patients were managed with anticoagulation alone [Table 1]. At follow-up after 6 months of anticoagulation on warfarin, all of them had reduction of edema and had returned to a normal life style. All patients are...
also taught complex decongestive therapy and use support stockings.

Experience with the use of new oral anticoagulant has not been published to date.

**Catheter-directed thrombolysis/thrombolytic therapy**
In patients with effort vein thrombosis, there are several proponents of CDT and suggestions that lytic therapy would help reduce the thrombus load. It should be initiated within the 1st week of onset of symptoms because resolution of fresh thrombus is higher. Results range from 62% to 84% and as high as 100% in cases where the thrombus was fresh.[4,5] Two of our patients underwent CDT and one was stented which occluded at follow-up.

Recent reports have shown that beyond 10 days after onset of symptoms, CDT is ineffective.[19,20] The University of Rochester, on the other hand, reported a success of 84% when treating patients who presented within 14 days.[21] In a report from Baylor for patients treated after >6 weeks, only 50% of the occluded veins could be partially opened; none completely.[16]

As per the ACCP 2016 guidelines, it is recommended that patients should be on the same intensity and duration of anticoagulation, postlysis, as those who received only anticoagulation (Grade 1B). [19]

**Stenting after anticoagulation and catheter-directed thrombolysis**
The underlying cause for thrombosis in these patients is a narrow costoclavicular junction, and therefore, stenting a narrowed segment of vein seen, without surgical decompression, although tempting, could lead to further damage of the endothelium, rethrombosis (reported to be as high as 40%), and stent fracture and complicate future surgical repair.[22-24]

**First rib resection**
Anticoagulation and CDT take care of acute thrombus; however, unless the chronic anatomic problem is addressed, up to one-third of patients will present with recurrence of thrombosis.[16,25,26] There are clearly two views about first rib resection (FRR) – one who proposes it and the other who feels it is not needed.

The first rib can be an approached transaxillary (TA), supraclavicular alone, or combined with paracervical. TA FRR which was popularized by Roos has been shown to yield good results in experienced hands.[27] In an inexperienced hand – resection of the second rib, injury to long thoracic nerve, and hemothorax have been reported. Urschel and Razzuk have reported “good-to-excellent” results in 85%–95% of their series.[28] The points to stress here are that the muscles around the costoclavicular junction should be nicely debulked and the fibrotic tissue around the vein (venolysis) be freed up to the level of the jugular vein.[29,30]

In 2007, Molina et al. published a 5-year follow-up with 100% patency and 0% symptom recurrence after CDT and TA resection.[31]

In our series, four patients underwent decompression (two underwent TA FRR and two underwent supraclavicular cervical rib resection).

In the absence of RCTs, it is safe to say that current evidence favors CDT and TA FRR, with excellent results in experienced hands.

**When should one offer resection of first rib?**
The controversy on timing for rib resection is higher than that about whether or not to resect.

Based on the theoretical risk of bleeding, endothelial function, and rethrombosis, the University of California, Los Angeles (UCLA) group performed resection/decompression 3 months after CDT. They reported 83% of the patients who underwent lysis and decompression to be free of symptoms. The group from Michigan reported that 90% of their patients returned to work.[24,32-34]

Based on evidence that rethrombosis can occur in 10% of patients during this interim period leading to morbid status and disability, there are several groups that offer lysis and decompression immediately with excellent results and low morbidity.[16,21,23,35-37]

Patients treated with anticoagulation therapy alone should undergo decompression soon after stopping anticoagulation therapy.

**Postlysis and rib resection – Venography, intervention**
After the above intervention, some would perform venography immediately, and some others, after a few weeks allowing the endothelium to heal. There is no consensus on the timing. Current evidence is in favor of angioplasty alone if any intrinsic lesion is found and against the stenting as this has been shown to have a higher incidence of thrombosis.[5,38]

**Vein reconstruction**
There could be a role for vein reconstruction in patients who do not improve after CDT, rib resection, and angioplasty. Surgeries described include creation of a temporary arteriovenous fistula and jugular vein turn down.[31,39,40]

None of our patients had venous reconstructive procedures.

**What if catheter-directed thrombolysis is unsuccessful?**
In such patients, it is recommended to treat patients with anticoagulation for 6 months. If at any point in time their symptoms worsen or do not resolve, decompression may be performed with continuing anticoagulation, with hope of recanalization and maturation of collaterals. Some patients are likely to need a jugular vein turn down.[5,41]

**Postoperative care**
Consensus is that anticoagulation is needed for a 6-month period, with periodic ultrasound assessment for recanalization. Contralateral symptoms while being low can occur, so patients need to be warned about the same and there may be a role for prophylactic FRR in the presence of cervical rib.[42]
Complications
PE can occur in 20%-36% cases as can stroke from a paradoxical embolism in the presence of a patent foramen ovale. Postthrombotic syndrome, right-sided heart failure, pulmonary hypertension, and phlegmasia cerulea dolens are some of the other complications mentioned.[17]

Treatment Algorithm
Our protocol has been drawn on the lines of that suggested by Illig and Doyle from Rochester Medical Center[5,21] [Figure 5].

Long-term outcomes
Due to a lack of RCTs and the availability of only several case series, it is difficult to express outcomes confidently. Illig and Doyle suggest that good outcomes were seen in 50%–66% of patients treated with anticoagulation (with or without decompression), 80%–90% in CDT and delayed rib resection group, and 90%–95% in CDT with immediate decompression group.[5]

Unanswered questions
- Natural history of the contralateral side
- Hypercoagulable states in effort vein thrombosis
- Management of ASVT in women
- Duration of anticoagulation postoperatively
- RCT to define time period of rib resection after CDT
- Cost and access to CDT for patients in India
- Adherence to protocol of management in India
- How to improve follow-up in India?

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Conflicts of interest
There are no conflicts of interest.

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Figure 5: Algorithm for treatment of patients with primary effort venous thrombosis of the upper limb
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