CLINICAL VIGNETTE

Effort Thrombosis: Venous Thrombosis Triggered by Exercise

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Introduction

Effort thrombosis (also called Paget-Schroetter syndrome) is a rare condition defined by primary thrombosis of the axillary-subclavian vein associated with strenuous activity of the upper extremities. While the presentation of lower and upper extremity deep vein thrombosis (DVT) can be similar, treatment can be markedly different based on the underlying pathophysiology. Therefore, it is important for physicians to recognize when unprovoked DVTs need further workup, understand the mechanisms involved, and expedite treatment in order to minimize morbidity and mortality. The following case describes an acute presentation of unprovoked upper extremity DVT in a young man caused by effort thrombosis and the subsequent management of this disorder.

Case Presentation

A 37-year-old, right-handed man presented to the emergency department with sudden onset of painless right upper extremity swelling and discoloration following an intense weight lifting session. His past medical history was significant for papillary thyroid carcinoma and thyroidectomy two years earlier. He recently started an exercise program including upper extremity weight lifting. He reported no recent injuries or trauma. Review of systems was negative for fever, cough, chest pain, and shortness of breath. His only medication is levothyroxine. Family medical history is negative for venous thromboembolism. He does not use tobacco, alcohol, or illicit drugs.

Physical exam revealed a well-appearing, well-developed, muscular young man with asymmetric swelling and decreased capillary refill of his right upper extremity. Vital signs were within normal limits, and basic laboratory work-up was unremarkable. CT chest angiography revealed a small pulmonary embolism within a subsegmental pulmonary artery in the right lower lobe, and Doppler ultrasound of the right upper extremity revealed an acute DVT of the subclavian and axillary veins. Duplex ultrasound of his arteries was normal. Heparin drip was initiated, and he was admitted to the hospital for further evaluation and treatment.

Hematology/oncology was consulted, and they had no concerns of recurrent thyroid cancer. TSH was appropriately suppressed at 0.03 mcIU/L (normal 0.3-4.7 mcIU/L). Ultrasound of the parathyroid and thyroid demonstrated the surgically absent thyroid without evidence of a recurrent mass. A PET scan 6 months prior to presentation was also negative.

Due to the absence of known thrombotic triggers, a magnetic resonance venography was ordered to look for thoracic outlet obstruction. MRV of the right upper extremity revealed an occlusive thrombus of the right axillary and right subclavian veins with high-grade compression of the subclavian vein between the right clavicle and first rib (Figure 1). This finding is consistent with Paget-Schroetter Syndrome, also known as effort thrombosis.

Interventional radiology and vascular surgery were consulted, and the patient underwent right upper extremity thrombolysis with tPA infusion two days after presentation. Follow-up angiography revealed residual luminal narrowing and irregularity of the right axillary-subclavian veins, indicating chronic thrombosis and the need for first rib resection. Five days after presentation, the patient was discharged from the hospital with plans for outpatient surgery.

The patient underwent partial resection of his first rib, subclavius, and scalene muscles 5 weeks after hospital discharge. Intraoperatively, the subclavius was noted to be approximately three times normal size with an exostosis, pushing the subclavian vein against the anterior scalene. The anterior scalene was approximately twice normal size, and the venous channel between the anterior scalene and the subclavius was approximately 3 mm. Subtotal resections of the subclavius, anterior scalene, and middle scalene muscles were performed, and the first dorsal rib was resected.

On one-month post-operative follow-up, venography revealed residual stenosis that was then corrected with balloon angioplasty and continued anticoagulation. Another month later, the patient was asymptomatic with normal function of his right arm and anticoagulation was discontinued.

Discussion

The mechanism of effort thrombosis is two-fold, involving both anatomical anomalies that narrow the thoracic outlet and repetitive microtrauma of the endothelium causing activation of the coagulation cascade.[1] The costoclavicular space within the thoracic outlet is bordered by the clavicle and first rib. This space contains the subclavian artery and vein, as well as the brachial plexus. The subclavian vein lies in a groove created by the anterior scalene muscle, clavicle, and first rib, and is highly vulnerable to injury at this location. Anatomical anomalies of the thoracic outlet are fairly common in the general population,[2] including abnormal insertion of the

anterior scalene onto the first rib, a congenital cervical rib, muscle hypertrophy, and fibrous bands in the thoracic outlet. These anomalies can cause narrowing of the costoclavicular space, predisposing patients to thrombosis.

Hyper-abduction and retroversion of the upper limb, especially in an obstructed thoracic outlet, can result in venous intima damage and the initiation of deep vein thrombosis. Repeated insults to the venous endothelium, such as those incurred by strenuous activities including weight lifting, wrestling, and other sports, are particularly likely to induce significant thrombosis.

Patients with effort thrombosis typically present as young and otherwise healthy men with acute or sub-acute swelling and arm discomfort in their dominant arm. Patients may also present with dilated and visible veins across the upper arm, reddish-blue discoloration, and arm heaviness. A specific event such as rigorous exercise that immediately elicited the onset of symptoms may be reported.[3]

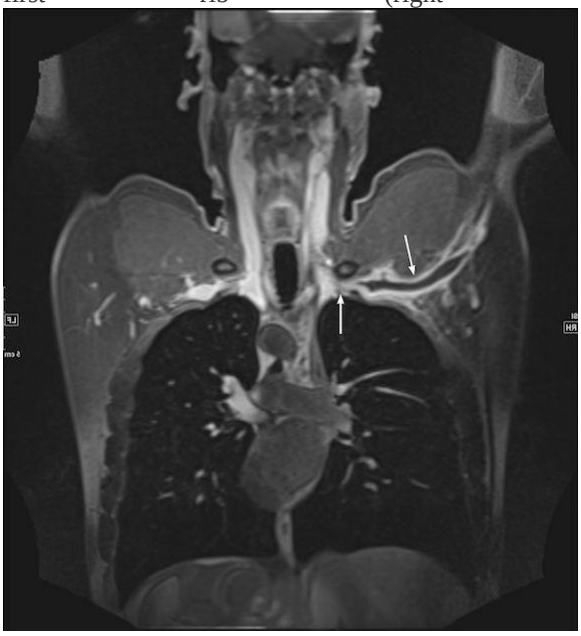
Effort thrombosis treated correctly in a timely manner will lead to minimal long-term consequences but if neglected can lead to severe negative sequelae such as recurrent thromboembolism, pulmonary embolism, and post-thrombotic syndrome. Effort thrombosis is often misdiagnosed as a simple muscle strain or incorrectly treated solely with anticoagulation as is commonly done for lower extremity DVT. Effort thrombosis managed with anticoagulation alone leaves the patient susceptible to acute pulmonary embolism in 6% to 15% of cases, residual upper extremity obstruction in up to 78% of cases, and permanent disability in the affected arm in 39% to 68% of cases.[3-5] Thus, it is important that physicians recognize the possibility of effort thrombosis in patients with otherwise unprovoked upper extremity DVT, order the appropriate imaging studies for diagnosis, and refer to knowledgeable specialists for timely and definitive treatment.

Diagnosis of effort thrombosis can be made with a duplex ultrasound scan noting occlusion of the axillary-subclavian vein at the costoclavicular junction.[3,6] Absence of flow, lack of compressibility, and an echolucent lesion indicate the presence of a recently developed thrombus, whereas a more chronic obstruction appears echogenic and fibrotic.[6] CT or MR venography offer high anatomical detail of nearby structures and can be used if the ultrasound scan is unequivocal.

Thrombolytic therapy followed immediately by first rib resection and scalenectomy is indicated for patients with less than 2 weeks of symptom duration. Lysis is less likely to be effective in patients with more chronic symptoms, and these patients should thus immediately undergo first rib resection and scalenectomy.[3] Following rib resection, all patients should receive anticoagulation for 3 to 6 months in addition to periodic ultrasound scans to assess flow in the subclavian. If necessary, venoplasty should be performed until the subclavian vein is patent.

No validated treatment algorithm currently exist for patients with effort thrombosis. In patients with short duration of symptoms prior to diagnosis, and an absence of identifiable anatomical anomalies, non-operative management has been shown to have similar outcomes as operative management. If non-operative management is preferred, patients should undergo catheter directed thrombolysis followed by 6 to 12 months of oral anticoagulation.[7] Patients should also frequently be assessed for vein patency with ultrasound. Ultimate goals of therapy should include patency of the axillary and subclavian veins, resolution of symptoms without further need for medications, and return to occupation or activities that induced the condition.

Figure 1. Contrast MRV revealed extensive clotting (left arrow) and subclavian vein compression at the clavicle and first rib (right arrow).



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Submitted June 14, 2016