

## **Primary deep vein thrombosis of the upper extremity in a 21 year old male – A Case Report of Paget-Schroetter syndrome.** Dr. Lawson Z. Hunley, D.O.

**Abstract:** Paget-Schroetter syndrome is a primary deep vein thrombosis of the upper extremity occurring typically in the subclavian or axillary veins. This case is of a 21 year old male who presented with an acute onset of left axillary swelling and tenderness three days after lifting weights. Ultrasound Doppler at the time revealed a thrombosis in the axillary and brachial veins. The patient was subsequently started on anticoagulation therapy. Although rare, cases such as this one illuminate the need for primary care, urgent care, and emergency room physicians to be aware of Paget-Schroetter syndrome in young healthy people in order to avoid the potential morbidity and mortality associated with the condition.

### **Case Presentation:**

A 21 year old male presented with a chief complaint of acute left axillary pain and pectoral swelling. The patient reported a gradual onset of swelling that started approximately three days prior. Associated symptoms were a non-radiating pain in his left axilla described as a “deep ache”, a decrease in his range of motion in the left upper extremity secondary to pain, and chills. He denied any associated chest pain, shortness of breath, numbness, tingling, erythema, skin changes, or temperature changes. The patient reported the pain was worsened with activity. He was unable to identify any palliative factors. He denied any recent or past trauma to his left upper extremity. He was not taking any medications. His past medical history and family history were negative for any bleeding disorders or malignancies. He did report recently starting a rigorous weight lifting program. He denied any current tobacco or illicit drug use, and no recent alcohol use.

Physical examination at the time found a well-developed, athletic male who was alert, oriented, and in no acute distress. No cardiopulmonary abnormalities were identified. There was mild to moderate swelling noted in his left axilla and lateral pectoral region. The area was mildly tender to palpation. There was no associated erythema, warmth, lesion(s), or mass. The superficial

veins proximal to the swelling were mildly engorged, but non-tender. Radial and brachial pulses were easily palpable and capillary refill was within normal limits.

An outpatient Ultrasound Doppler of his left upper extremity showed a deep vein thrombosis in the axillary and brachial veins. The patient was subsequently sent to the Emergency Department for further evaluation. A CT Venogram of the left upper extremity was performed with intravenous administration of Optiray 350. Sagittal coronal and volume rendered images were utilized during interpretation of the exam. The study confirmed the occlusion of blood flow through the left axillary vein. The radiologist noted at the time that although the study was performed with the arm in the elevated position, it appeared that the thrombosis may be related to narrowing of the subclavian vessels as they cross between the first rib and the adjacent clavicle.

After consultation with a vascular surgeon the patient was started on subcutaneous enoxaparin and oral warfarin, an outpatient appointment was made with the vascular surgeon, and the patient was discharged home.

### **Paget-Schroetter syndrome**

Paget-Schroetter syndrome, also referred to as Effort Thrombosis, is an idiopathic thrombosis of the deep veins of the upper extremity, usually the subclavian and/or axillary veins. (Vijaysadan, Zimmerman, and Pajaro) The syndrome was named Paget-Schroetter due to early descriptions of the syndrome by Dr. James Paget in 1875 and Dr. von Schroetter in 1894. In the 1960s the term “effort” thrombosis was coined, acknowledging that the syndrome often occurred in physically active individuals after unusually strenuous use of the arm and shoulder.

Paget-Schroetter Syndrome is a form of Thoracic Outlet Syndrome. As Illig and Doyle point out in their article, “A comprehensive review of Paget-Schroetter syndrome”, “There is a surprising amount of confusion surrounding Thoracic Outlet Syndrome in general, due to the mistaken assumption that this is all one entity.” (Illig KA, Doyle AJ.) In reality, Thoracic Outlet Syndrome is

divided into 3 broad categories: Neurogenic, Venous, and Arterial. Paget-Schroetter Syndrome is a form of Venous Thoracic Outlet Syndrome. Venous Thoracic Outlet Syndrome is further divided into either primary or secondary based upon pathogenesis. Paget-Schroetter syndrome is a subset of primary upper extremity deep vein thrombosis. **Table 1** lists the causes of primary and secondary upper extremity deep vein thrombosis.

**Table 1 Primary vs Secondary Upper Extremity Deep Vein Thrombosis**

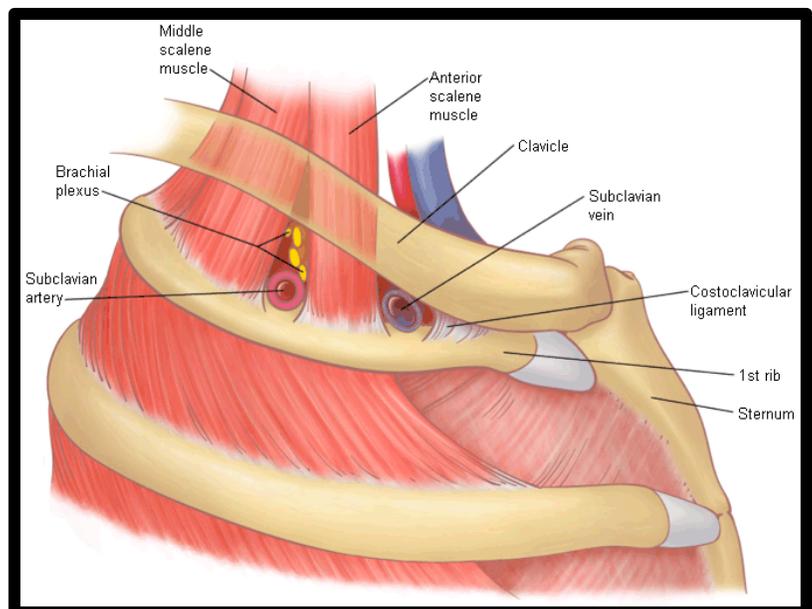
**I. Primary (20%)**

- A. Idiopathic
- B. Related to anatomical abnormalities (Thoracic Outlet Syndrome)
- C. Paget-Schroetter Syndrome (Effort Thrombosis)

**II. Secondary (80%)**

- A. Central Venous Catheters
- B. Pacemakers
- C. Malignancy
- D. Arm surgery or trauma
- E. Immobilization (plaster cast)
- F. Oral contraceptive use
- G. Pregnancy
- H. Ovarian hyperstimulation syndrome

The Thoracic Outlet consists of two distinct anatomic regions, the triangle formed by the anterior and middle scalene muscles which the brachial plexus and subclavian artery pass through, and the area more anterior near the junction of the clavicle and first rib where the subclavian vein re-enters the chest. The deep veins of the upper extremity are the ulnar, radial,



*Figure 1: Normal Anatomy of the Thoracic Outlet*

interosseous, brachial, axillary, cephalic, and subclavian veins. Regardless of cause, upper extremity deep vein thromboses occur mainly in the Subclavian vein (18-69%), axillary vein (5-42%), and brachial vein (4-13%).

Figure 1 shows the normal anatomy of the thoracic outlet and figure 2 shows the venous system of the upper extremity. (Alla VM, Natarajan N, Kaushik M, et al.)

Upper extremity deep vein thrombosis is a relatively rare presentation accounting for only 1-4% of all diagnosed cases of venous thrombosis annually. (Goshima, Kaoru) Primary upper extremity deep vein thrombosis represents only 20% of upper deep venous thrombosis. Paget-Schroetter syndrome is relatively rare subset of primary upper extremity deep vein thrombosis with an estimated incidence of 1-

2 per 100 000 hospital admissions. Extrapolated out that would be approximately 3000 - 6000 cases per year in the United States of America. (Illig)

Paget-Schroetter Syndrome occurs primarily in young health individuals who engage in athletic activities or have professions that require repetitive arm movements. The median age at presentation is in the early 30's with men 2:1 as compared to women. The right upper extremity

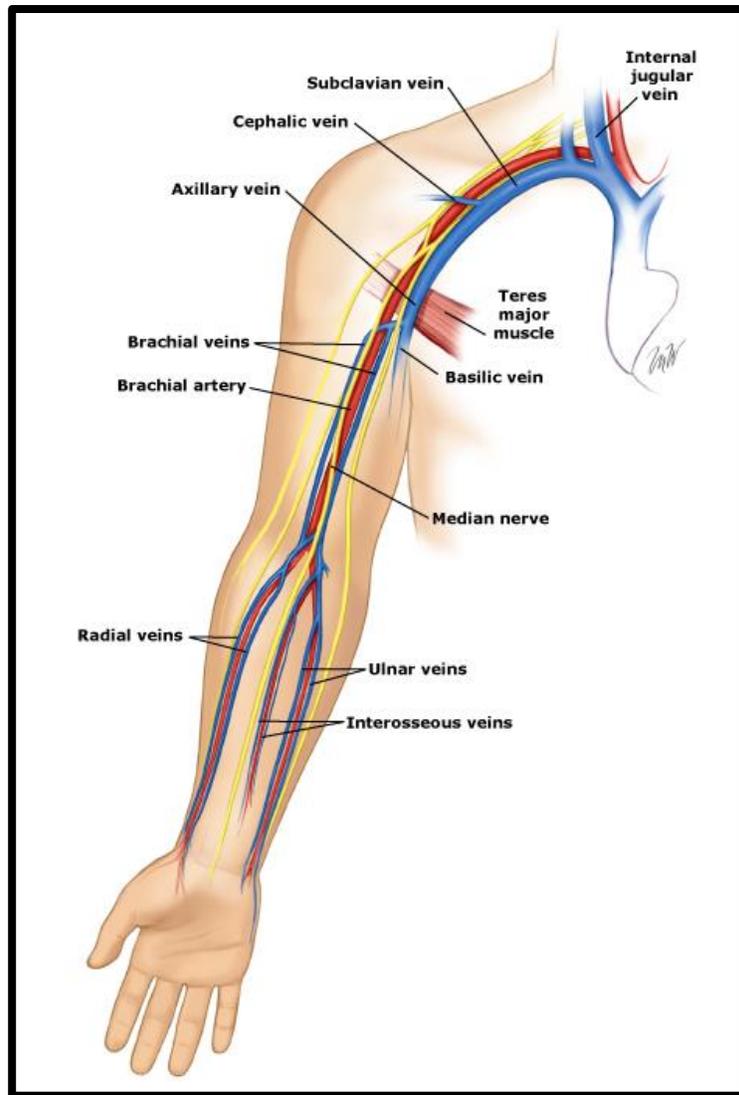


Figure 2: Vasculature of the Upper Extremity

is more common in the right than the left. Repetitive overhead arm movements that require hyper-abduction along with external rotation of the glenohumeral joint have been implicated. Common sporting activities include weight lifting, wrestling, baseball, gymnastics, and swimming. (Alla)

Although unknown, it is thought Paget-Schroetter syndrome is due to an underlying compressive anomaly in the thoracic outlet in patients who develop spontaneous deep vein thrombosis. This is frequently due to compression of the vein either between the first rib and a hypertrophied scalene muscle or subclavian tendon or between these tendons themselves. Compression between the clavicle and a cervical rib as well as partial occlusion of the vein by a congenital web has also been reported (Swinton and Fisher). It is also unclear if Paget-Schroetter is caused by a single injury or repetitive injuries to the affected vein.

Only 15% of the patients are symptomatic but the majority of them present with dull, aching pain in the shoulder and swelling of the arm and hand. These symptoms tend to worsen with vigorous use of the arm and improve with rest and elevation. Local trauma or, more frequently, strenuous use of the arm can be recalled by approximately one-half of patients, and symptoms generally are noted within 24 hours of the unusually strenuous activity. Diagnosis is made with a duplex ultrasound. A D-dimer can be used to exclude a thrombosis, but it will not exclude venous stenosis as a cause for symptoms. Therefore, a negative D-dimer does not exclude the need for an duplex ultrasound.

The optimal management of subclavian vein effort thrombosis remains a dilemma. According to Illig and Doyle "the central issues are whether this should be treated at all, the best method of treatment of the thrombosis itself, timing and method of 'permanent' correction of the underlying defect, and how to treat and follow the patients in the long term." (Illig) The dilemma exists because of the relative rarity of the disease, lack of awareness, and lack of large randomized trials. Current management of Paget-Schroetter Syndrome are based on isolated case reports and small retrospective series. Historically Paget-Schroetter Syndrome was

managed conservatively with elevation of the affected arm and anticoagulation. After long-term data began to show a high rate of continued symptoms, recurrent thrombosis, and disability, clinicians began to look at more aggressive treatment options. Anticoagulation is at the cornerstone of current treatment for Paget-Schroetter Syndrome, but treatment also now includes thrombolysis, thrombectomy, surgical and percutaneous venoplasty, and venous stents and bypass.

Paget-Schroetter Syndrome has a potential for considerable morbidity due to the risk of pulmonary embolism which is present in up to a third of patients. Other complications, such as persistent pain and swelling, superior vena cava syndrome, and problems with vascular access, can be disabling. Since the Paget-Schroetter Syndrome generally affects young, otherwise healthy individuals with an active lifestyle and long life expectancy, there is general consensus that the major goal of therapy is to minimize the likelihood of significant symptoms of venous insufficiency.

In summary, Paget-Schroetter Syndrome is a relatively rare primary idiopathic thrombosis of the upper extremity in young healthy individuals. It occurs more often in the right arm of males after repetitive strenuous activity. It is diagnosed with duplex ultrasound. Current management is geared towards decreasing repeat thrombosis and persistent symptoms. Due to the potential complication of life-threatening pulmonary embolism and the potential for long-term disability it is essential that primary care providers be aware of this condition in order to properly diagnose patients they may see in their primary care office, urgent care center, or emergency department.

## REFERENCES

- Illig KA, Doyle AJ. **A comprehensive review of Paget-Schroetter syndrome.** J Vasc Surg 2010; 51:1538.
- Alla VM, Natarajan N, Kaushik M, et al. **Paget-schroetter syndrome: review of pathogenesis and treatment of effort thrombosis.** West J Emerg Med 2010; 11:358.
- Nemmers DW, Thorpe PE, Knible MA, Beard DW. **Surgical management of subclavian-vein effort thrombosis as a result of thoracic outlet compression. Case report and literature review.** Orthopaedic Review 1990 Feb; 19 (2): 164-72.
- Molina JE. **Need for emergency treatment in subclavian vein effort thrombosis.** Journal of the American College of Surgeons 1995 Nov; 181 (5) : 414-20.
- Doyle A, Wolford H, Davies M G, Adams J T, Singh M J, Saad W E, Waldman J A, Illig K A. **Management of effort thrombosis of the subclavian vein: today's treatment.** Annals of Vascular Surgery 2007 Nov; 21 (6) : 723-9.
- Spiezie Luca, Simioni Paolo. **Upper extremity deep vein thrombosis.** Internal and Emergency Medicine 2010 Apr; 5 (2) : 103-9.
- Azaki A, McElhinney DB, Thompson RW, Raven, RB, Messine LM, SToney RJ. **Surgical management of subclavian-vein effort thrombosis as a result of thoracic outlet compression.** Journal of Vascular Surgery 1998 Nov; 28 (5) : 777-86.
- Roche-Nagle Graham, Ryan Ronan, Barry Mary, Brophy David. **Effort Thrombosis of the upper extremity in a young sportsman: Paget-Schroetter syndrome.** Br J Sports Med 2007; 41:540-541.
- Leebeek F W, Stadhouders S N A , et al. **Hypercoagulability states in upper-extremity deep venous thrombosis.** Am J Hematol, 2001, 67(1) : 15-9
- Aburahma A. F., Sadler D. et al. **Conventional versus thrombolytic therapy in spontaneous effort axillary-subclavian vein thrombosis.** Am J Surg, 1991, 161 (4) : 459-65
- Swinton N. W., Jr., Edgett J. W., Jr., Hallr. J. **Primary subclavian-axillary vein thrombosis.** Circulation, 1968, 38(4) : 737-45
- FISHER J. B., GRANSON M. A. **Congenital venous web causing subclavian vein obstruction: a case report.** J Vasc Surg, 1989, 10(4):460-2.
- MACHLEDER H. I. **Effort thrombosis of the axillosubclavian vein: a disabling vascular disorder.** Compr Ther, 1991, 17(5) : 18-24.
- STEVENSON I. M., PARRY E. W. **Radiological study of the aetiological factors in venous obstruction of the upper limb.** J Cardiovasc Surg (Torino), 1975, 16(6) : 580-5.
- Goshima, Kaoru. **Primary (spontaneous) upper extremity deep vein thrombosis.** UpToDate. Accessed on 25 September 2013.