American Journal of Medical Case Reports, 2016, Vol. 4, No. 9, 315-318
Available online at http://pubs.sciepub.com/ajmcr/4/9/6

DOI:10.12691/ajmcr-4-9-6

Paget-Schroetter: Primary Axillo-subclavian Vein Thrombosis in a Young Patient

Cesar Gentille*, Michael Arriaga, Christopher Peckins

Department of Internal Medicine, Houston Methodist Hospital, Houston, Texas, United States of America
*Corresponding author: cgentillesanchez@houstonmethodist.org

Abstract Upper extremity (UE) deep venous thrombosis (DVT) is much less common than lower extremity DVT. Primary UE DVT, that which occurs in the absence of known risk factors is quite rare. We present a case of Paget-Schroetter Syndrome (PSS) also known as "Effort Thrombosis," in a healthy young woman likely brought on by thoracic outlet syndrome (TOS). Our patient developed an UE DVT with minimal symptoms. Ultrasound, and then angiogram demonstrated a dense, long clot in the axillo-subclavian vein. Symptoms improved after catheter-directed thrombolysis although it was only partially successful. A narrowed region of the subclavian vein persists where it exits the thoracic outlet. Clinicians need to be aware of the unusual diagnosis as treatment can improve symptoms and facilitate recovery.

Keywords: *venous thrombosis, thrombolysis*

Cite This Article: Cesar Gentille, Michael Arriaga, and Christopher Peckins, "Paget-Schroetter: Primary Axillo-subclavian Vein Thrombosis in a Young Patient." *American Journal of Medical Case Reports*, vol. 4, no. 9 (2016): 315-318. doi: 10.12691/ajmcr-4-9-6.

1. Background

The axillo-subclavian vein is an uncommon site for a deep venous thrombosis. Common risk factors for thrombosis include prolonged immobility, previous thrombosis, hormonal treatment, chronic inflammatory disease, hypercoagulable states and others. In the absence of known risk factors, axillo-subclavian thrombosis is called Paget-Schroetter Syndrome [1,3,6,9].

Paget-Schroetter syndrome is a primary upper extremity DVT that occurs spontaneously, usually in young, healthy patients after strenuous activity. Secondary upper extremity DVTs occurring in the context of a known risk factor such as a hypercoaguable state or the presence of an intravascular device are more common [7]. PSS it thus distinct not only from lower extremity DVT but also from secondary upper extremity DVT and may require specific management strategies [3,6].

We report a case of a young patient with no significant medical history, who presented with increased swelling in her right arm and was subsequently found to have a clot originating in the axillary vein extending into the subclavian and cephalic veins. We will discuss the presentation, diagnosis and treatment of this uncommon entity, and address management.

2. Case Presentation

A 30-year-old female patient without significant past medical history presented to our Emergency department with right arm swelling for one week. She first noticed a change in her right arm two weeks prior when, after doing upper body exercises, she had developed an unusual sensation of soreness in both arms. Subsequently she noticed that the superficial veins of her right arm became more prominent; however, she did not experience any tenderness or additional episodes of pain. One week later a family member noted swelling in her right arm, which prompted a visit to her Primary Care physician's office where a Doppler ultrasound was performed and demonstrated thrombosis in the right subclavian, right axillary and right cephalic veins.

She denied fever, chest pain, shortness of breath, weakness, numbness, weight loss, or recent extended travel. The social and family histories were noncontributory. She also denied using oral contraceptives or any other medications. There was no history of intravenous catheter placement, recent surgeries, prolonged immobilization, known hypercoagulability nor recurrent fetal loss.

On examination, the patient was a pleasant, healthy appearing female in no acute distress and with a BMI of 20 kg/m^2. Temperature 36.5C. Blood pressure 118/67 mmHg, HR of 78 bpm and RR 14. Her right arm appeared edematous with engorged superficial collateral veins, however, there was no tenderness on palpation. Pulses (ulnar, radial and brachial) were palpable on her right arm however a loss of her radial pulse was noted after external rotation, extension and elevation of her right arm. Muscle strength was preserved (5/5) in all extremities. Sensation to pain and touch and deep tendon reflexes were also normal.

Hypercoagulable workup was performed which included cardiolipin antibodies, lupus anticoagulant, dilute Russell's Viper Venom time, ANA, functional protein C and S, factor V Leiden and prothrombin G20210A

(mutation factor 2). They were all negative. She was up to date with age and sex-appropriate cancer screening. Vascular surgery was consulted for a suspected thoracic outlet syndrome. An upper extremity venogram demonstrated acute on chronic thrombosis of the right axillosubclavian vein with fresh thrombus in the cephalic arch.

3. Treatment

The results of the venogram, in the context of her presentation, and her lack of known predisposing conditions suggested a diagnosis of primary upper extremity DVT (Paget-Schroetter syndrome), possibly due to thoracic outlet obstruction. Two different intravascular wires (Cook Medical Bentson wire, Terumo GLIDEWIRE) were utilized unsuccessfully in the attempt to traverse the clot. Venogram was then performed through the sheath (Figure 1). Afterwards a Cook Medical Triforce peripheral crossing set was placed over the wire and using the GLIDEWIRE and catheter system, the thrombosed area was successfully crossed. Following this, a balloon angioplasty of the innominate vein, subclavian vein and axillary vein was done (Figure 2). Finally, a drug – delivery catheter with an endovascular ultrasound system (EKOS corporation) was placed, and heparin, Tissue Plasminogen Activator (tPA) and coolant infusion were administered without any complications (Figure 3).

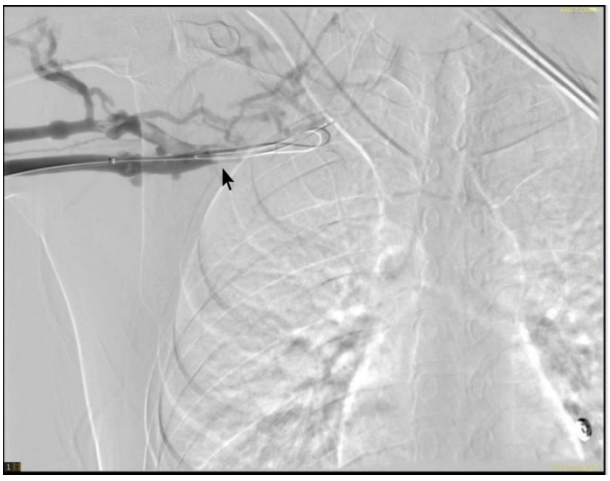

Figure 1. Venogram showing the acute thrombus in the right cephalic arc-h and the chronic thrombosis of the right subclavian vein (arrow)

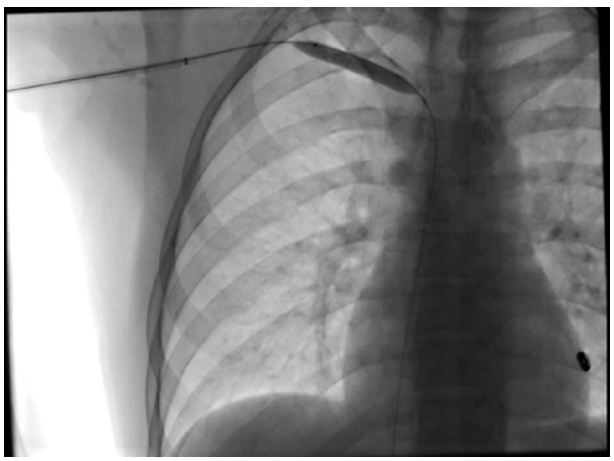

Figure 2. Balloon angioplasty of the innominate vein, subclavian vein and axillary vein

After approximately 24 hours of infusion, the patient was brought back to the operating room for a lysis check and treatment. The EKOS catheter was removed and a follow-up venogram was done which showed a residual area of stenosis in the subclavian vein in the thoracic outlet between the clavicle and the first rib. Due to the presence of residual disease, the vascular surgeon performed another balloon angioplasty; however, the area of stenosis persisted. This area of stenosis was presumed to be chronic and would likely lead to re-occlusion in the absence of definitive surgical correction, thus no further intervention was performed at that time. After a discussion with the patient, and in recognition of her minimally symptomatic state, additional surgical options were deferred. We initiated anticoagulation with apixiban and she was discharged home with follow up with Hematology and Vascular Surgery.

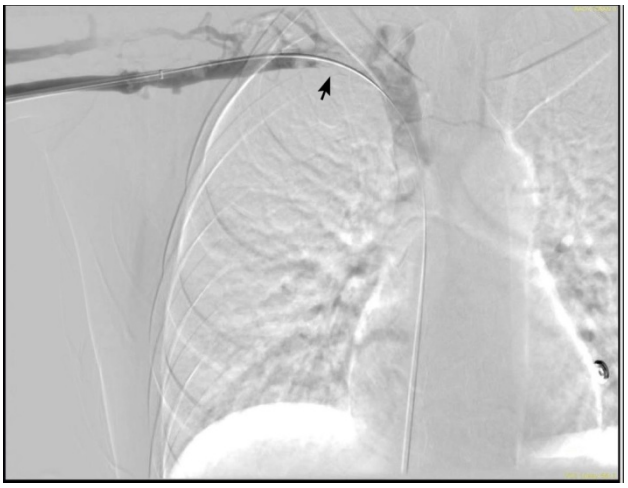

Figure 3. Residual thrombus in the subclavian vein in the thoracic outlet between the clavicle and the first rib (arrow)

4. Follow-up

The patient continued taking apixaban. Upon follow up with her hematologist she reported that the symptoms had improved significantly. Approximately three months later, a screening ultrasound demonstrated a thrombus in her contralateral (left) subclavian vein though she remained completely asymptomatic. She is continuing anticoagulation and seeking surgical advice regarding possible definitive treatment.

5. Discussion

In some instances, Paget-Schroetter syndrome has been described as an idiopathic entity and in others it has been associated with vigorous exercise involving the upper extremities. At times an anatomically narrowed thoracic outlet (TOS) was identified and considered to be causal. In the latter cases, it is thought that the subclavian, or the axillary vein is being partially compressed between the clavicle and the first rib [6,10]. In addition, the presence of other factors such as a hypertrophied scalenus muscle or subclavian tendon, a cervical rib, congenital bands, or abnormal insertion of the costoclavicular ligament in certain patients could increase the probability for

thrombosis in that area. Even in patients with normal anatomy, the subclavian vein can be easily compressed within the arm in extremes of abduction or external rotation [1,3,4,6,9].

A wide variety of sustained upper extremity movements may precipitate this condition. PSS has been reported in patients regularly doing weight training, gymnastics, or other sports as well as in occupations such as mechanic, painter and manual laborer [1,3,6]. Cases have also been reported in participants of baseball, softball, wrestling, swimming, hockey, martial arts, backpacking, and billiards [10]. Our patient exercised regularly and shortly before presentation had noticed an acute episode of pain in both arms after doing a vigorous routine that involved constant upper extremity abduction. Activities that involve hyperabduction, posterior rotation or extension of the arm have been implicated as contributory or causal to PSS [1,6]. Paget-Schroetter is also referred to as "Effort Thrombosis."

Clinical presentation often involves arm swelling and discomfort, but can also include discoloration of the arm and heaviness. The presence of dilated visible veins in the affected shoulder and arm can also be seen. In our case there were minimal symptoms with swelling and some dilation of superficial venous collaterals but without tenderness, discoloration or any other signs of thrombosis. A history of strenuous exercise involving the upper extremities antedating the development of symptoms is frequent among these patients [6,10,15].

The preferred imaging for diagnosis is Doppler ultrasound. It is a rapidly available and a minimally invasive test with a sensitivity 78% to 100% and specificity from 82% to 100% [2]. Venogram is generally performed if the results from the US are inconclusive and/or as an initial therapeutic step. This invasive procedure entails more risk as well as cost. Second line imaging tests include MR and CT venography [1,6,9]. In our case, the venogram was performed as an initial step.

Treatment guidelines suggest management of UE DVT with anticoagulation as initial treatment, with further management in selected cases [5]. Several studies have described unacceptable high morbidity and recurrence rates with this single modality therapy [1,4,5,12]. Improved outcomes have been reported with a sequential multimodality approach which usually includes thrombolysis (catheter-directed, systemic) and/or thoracic outlet decompression [4,5,6,9,12,13,17]. A retrospective study reviewing therapy of PSS patients over the last 50 years from one medical center showed that from the group of patients that received only anticoagulation had persistent symptoms at follow-up while most of the patients receiving thrombolysis and first rib resection had complete resolution of their symptoms [15]. The apparent need for further management is likely related to the chronic compression associated with TOS and possibly from intimal hyperplasia of the involved veins, and inflammation and fibrosis of the surrounding tissues [6,11,15].

Despite the lack of consensus, some authors suggest a combined approach of anticoagulation and an intervention; most treatment algorithms include catheter directed thrombolysis (CDT), thoracic outlet decompression and post-operative anticoagulation [10,12,13,17]. Catheter directed thrombolysis for acute treatment of thrombus is associated with better outcomes if performed within two weeks of presentation [1,6,9]. The procedure typically involves venous anterograde access through the arm, penetration and traversal and catheter directed thrombolysis of the lesion (optionally with adjunctive ultrasonic thrombolysis), followed up by tPA infusion (usually from 24h-48h), along with heparin. A follow-up venogram is performed 24 hours later to look for signs of residual disease [16]. It is not uncommon to find intrinsic venous defects afterwards likely related to the underlying chronic injury associated with PSS [6]. Surgical decompression through first rib resection has been recommended in all patients with residual compression with a reported success rate of around 90-95% [6,10,15]. Even though some physicians prefer surgery with or without preceding thrombolysis, groups of patients have also been reported to do well without surgical treatment [1,4,6,8]. A retrospective study of 27 PSS patients receiving non-operative treatment (anticoagulation alone or with CDT) noted only 15% of patients had recurrence at two and a half years follow up, while 85% were asymptomatic and without need of further therapy with decompression [4].

The patient's specific characteristics, the nature of the precipitating event and the time to diagnosis all can affect the decision for therapy [14]. A conservative approach might be preferred in older patients with more than 2 weeks of onset, in the context of an isolated event (unusual strenuous activity), with no systemic factors that could predispose to DVT and with rapid improvement on IV heparin. Some physicians prefer conservative treatment with CDT and anticoagulation first in all patients, advancing therapy to decompression if there is persistence of symptoms [1,6,16].

Optimal treatment of PSS is not yet defined. A conservative, sequential approach individualized to each patient may be a way to optimize risks and benefits for each case. Our patient was treated with CDT and angioplasty that diminished her clot burden and effected recanalization with residual narrowing. Given her lack of significant symptoms, it was not felt that the balance of risk and benefits favored immediate decompression surgery. She continues on long-term anticoagulation and ongoing close clinical monitoring.

6. Learning Points

1. Paget-Schroetter Syndrome is an idiopathic DVT of the upper extremity and has been described in the context of repetitive upper extremity exercises and with thoracic outlet narrowing.
2. Secondary UE DVT is much more common, and usually follows placement of an intravascular device.
3. Management algorithms have not been delineated, but CDT (with or without endovascular ultrasound dispersion), angioplasty, long-term anticoagulation, and definitive surgical decompression have all been utilized.
4. Since Primary DVT is generally a benign process, a thoughtful, sequential and conservative approach may be warranted.

Competing Interests

The authors declare that they have no competing interests.

References

[1] Alla V, Natarajan N, Kaushik M, Warrier R, Nair C. Paget-schroetter syndrome: Review of pathogenesis and treatment of effort thrombosis. *West J Emerg Med 11(4): 358-362*. 2010; 11(4): 358.

[2] Chin EE, Zimmerman PT, Grant EG. Sonographic evaluation of upper extremity deep venous thrombosis. *J Ultrasound Med*. 2005; 24(6): 829-38; quiz 839-40.

[3] Feugier P, Chevalier J. The paget-schroetter syndrome. *Acta chir belg*. 2005:256.

[4] Goss SG, Alcantara SD, Todd GJ, Lantis JC,2nd. Non-operative management of paget-schroetter syndrome: A single-center experience. *J Invasive Cardiol*. 2015; 27(9): 423-428.

[5] Guyatt GH, Akl EA, Crowther M, Gutterman DD, Schuunemann HJ, American College of Chest Physicians Antithrombotic Therapy and Prevention of Thrombosis Panel. Executive summary: Antithrombotic therapy and prevention of thrombosis, 9th ed: American college of chest physicians evidence-based clinical practice guidelines. *Chest*. 2012; 141(2 Suppl): 7S-47S.

[6] Illig KA, Doyle AJ. A comprehensive review of paget-schroetter syndrome. *J Vasc Surg*. 2010; 51(6): 1538-1547.

[7] Joffe HV, Goldhaber SZ. Upper-extremity deep vein thrombosis. *Circulation*. 2002; 106(14): 1874-1880.

[8] Kidd M, Broderick V. An unusual presentation of a swollen arm: A case report. *J Med Case Rep*. 2014; 8:22-1947-8-22.

[9] Koury J, Burke C. Endovascular management of acute upper extremity deep venous thrombosis and the use of superior vena cava filters. *Semin Intervent Radiol*. 2011(28): 3.

[10] Mall N, Van Thiel G, Heard W, Paletta G, Bush-Joseph C, Bach BJ. Paget- schroetter syndrome: A review of effort thrombosis of the upper extremity from a sports medicine perspective. . *Sports Health*. 2013; 5(4): 353.

[11] Molina JE, Hunter DW, Dietz CA. Paget-schroetter syndrome treated with thrombolytics and immediate surgery. *J Vasc Surg*. 2007; 45(2): 328-334.

[12] Rosa Salazar V, Otalora Valderrama Sdel P, Hernandez Contreras ME, Garcia Perez B, Arroyo Tristan Adel A, Garcia Mendez Mdel M. Multidisciplinary management of paget-schroetter syndrome. A case series of eight patients. *Arch Bronconeumol*. 2015; 51(8): e41-3.

[13] Thiruchelvam N, Mbuvah F, Kistangari G, Anumandla AK. Upper-limb deep vein thrombosis in paget-schroetter syndrome. *Cleve Clin J Med*. 2015; 82(10): 658-659.

[14] Thompson JF, Winterborn RJ, Bays S, White H, Kinsella DC, Watkinson AF. Venous thoracic outlet compression and the paget-schroetter syndrome: A review and recommendations for management. *Cardiovasc Intervent Radiol*. 2011; 34(5): 903-910.

[15] Urschel HC, Jr, Patel AN. Surgery remains the most effective treatment for paget-schroetter syndrome: 50 years' experience. *Ann Thorac Surg*. 2008; 86(1): 254-60; discussion 260.

[16] Vedantham S. Endovascular procedures in the management of DVT. *Hematology Am Soc Hematol Educ Program*. 2011; 2011: 156-161.

[17] Young K, Tunstall O, Mumford A. Subclavian vein thrombosis in an otherwise healthy 9-year-old boy. *BMJ Case Rep*. 2014.