

Case Report

Paget-Schroetter Syndrome: A Case Report of effort induced upper extremity venous thrombosis

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Effort induced thrombosis of the axillary-subclavian vein, also referred as Paget-Schroetter syndrome, is a relatively uncommon type of deep vein thrombosis of the upper extremity. Although rare, understanding its clinical presentation and etiology is essential for timely diagnosis and formulating correct treatment protocols. Most upper extremity deep vein thrombosis are secondary to either underlying pathology including cancer, chronic indwelling catheters, and hypercoagulable states making their clinical diagnosis relatively straight forward. On the contrary, effort induced deep vein thrombosis most commonly occur in young otherwise healthy people. Early detection and treatment of this type of thrombosis are essential in preventing late-term complications including post-thrombotic syndrome, and pulmonary embolus. There is no current consensus in treatment of effort induced upper extremity thrombosis with literature supporting conservative versus surgical treatment depending on patient presentation and physician preference.

INTRODUCTION

Spontaneous thrombosis of veins in the upper extremity is a relatively rare complication in otherwise healthy young individuals. Sir James Paget was the first to postulate the symptoms of acute pain and swelling of the upper extremity to venous thrombosis but it was Von Schroetter, in 1884, who clinically correlated these symptoms with occlusion of the axillosubclavian venous system (Haire 2011). Sub-

sequently termed Paget-Schroetter syndrome, spontaneous thrombosis of the upper extremity is rare, roughly 2 in 100,000 (Joffe and Goldhaber 2002); however the category of axillosubclavian venous thrombosis is relatively common. Joffe et al. reported that roughly 10% of all DVTs occur in the upper extremity with risk factors differing from lower extremity DVTs. Joffe and Goldhaber cited cancer, pacemakers, and the increasing use of central venous catheters for dialysis, chemotherapy, bone marrow trans-

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plant, and parental nutrition as likely causes of UEDVT. In comparison with patients with lower extremity DVT, upper extremity DVT patients were younger (59.2 ± 18.2 vs. 64.2 ± 16.9 years $p < 0.0001$), less often white (65 vs. 73% $p < 0.01$), leaner ($BMI 26.8 \pm 7.1$ vs. 28.5 ± 7.3 kg/m. The importance of early detection and diagnosis of these secondary causes of thrombosis is paramount as undiagnosed or untreated UEDVTs can lead to pulmonary embolus, superior vena cava syndrome, loss of venous access, and disabling pain.

The etiology of Paget-Schroetter syndrome or spontaneous axillosubclavian venous thrombosis (ASVT) is often unexpected in an otherwise healthy young adult who is likely involved in athletics or repetitive overhead activities most commonly long distance swimming, body building, wrestling, handball, and baseball throwing (Oktar and Ergul 2007; Haire 2011). Although multifactorial in etiology, there is a consensus of extrinsic compression of the subclavian vein leading to repetitive micro trauma of the vessel followed by stenosis and eventually thrombosis secondary to inflammation and stasis. It is postulated thoracic outlet syndrome referring to a compression of the neurovascular bundle at it exits the thoracic inlet may play a large role in the development of spontaneous ASVT. The venous system of the upper extremity is considerably prone to compression at the junction where the axillary, basilic, and cephalic vein converge to form the subclavian vein. It is at this junction where a tunnel is formed by the clavicle and subclavius anteriorly, the anterior and middle scalene laterally, and the first rib superiorly. Compression by any of these structures and even as far lateral as the pectoralis minor insertion onto the coracoid may contribute to the underlying pathology of spontaneous ASVT.

While the axillo-subclavian vein is a primary culprit in effort induced thrombosis, our current case report reveals an effort induced compression leading to a thrombus of the internal jugular vein.

CASE REPORT

A 13 year old right hand dominant female presented with pain and heaviness of her right shoulder. She was at practice for fast pitch softball the day before the onset of her symptoms. Her initial complaint was pain laterally in her right shoulder and eventually she had problems lifting her shoulder above her head for any period of time complaining weakness/heaviness of the right upper extremity. She denied any recent trauma prior to the onset of symptoms. Palliative efforts included ice, which made it worse and heat, which made it better. She also took 200 mg Motrin daily prn. Upon further questioning, her mother explains that she practices softball pitching about 4 times a week with 2 practices with her team and 2 practices with her coach. She has taken the last 3 weeks off with some but not complete relief of the right lateral shoulder pain. Patient denies any numbness, tingling, cold intolerance, or swelling of the right upper extremity. Of note, her mother states she did have an engorged vein behind her right ear growing up which became painful for her when it was engorged. She

has no past medical or surgical history and has no allergies. She does not use tobacco or etoh.

On physical exam she stands at 5'2" tall and 117 lbs with a BMI of 21. Examination of her right shoulder revealed no acute abnormality or previous injury to the clavicle or AC joint. She did have palpatory tenderness to the lateral aspect of her shoulder and proximal humerus. Her right extremity did not feel warm or edematous nor was there any evidence of muscle atrophy. She had full active range of motion of her right upper extremity with an equivocal exam on the left. Patient demonstrated 5/5 muscle strength of her supraspinatus, infraspinatus, subscapularis, and abduction of her shoulder although she did have a great deal of pain against resistance in abduction of the shoulder. Special examinations revealed a positive Neer test, but negative for Hawkins's, Speed's, cross-arm., O'Brien's, Mimpri, apprehension, and relocation. She had good radial and ulnar pulses and was motor intact distal. AP, axillary, and scapular Y x-rays of the right shoulder were within normal limits (Figure 1). At that time the patient was sent for an MRI and told to continue to rest her pitching arm. Upon return, a review of her MRI showed a normal appearing supraspinatus, infraspinatus, subscapularis, and labrum. There were increased vascular channels in the teres minor muscle extending into the suprascapular and spinoglenoid notch, posterior aspect of the scapula, and lateral aspect of the proximal humerus extending into the greater tuberosity (Figures 2 and 3). The radiologist considered these findings consistent with a slow flow vascular malformation suggesting further work up. Upon these findings, the patient was instructed to follow up with a vascular surgeon, again with the instructions to rest the right shoulder. On follow up with the vascular surgeon, peripheral arterial Doppler testing was performed on the right upper extremity which showed no evidence of peripheral arterial disease at rest in the right arm. The right wrist/brachial index was 1.09 compared to 1.0 on the left. However, the duplex did show an occlusion of the right jugular vein with extensive collateral circulation feeding a very large right subclavian vein. There was minimal collateral circulation in the neck.

The patient in this case study received serial ultra sound studies to monitor the thrombus, took time away from her sport, and underwent anti-coagulation. She has had no surgical intervention at this time. Treatment was focused on dissolution of the thrombus with medical intervention alone.

DISCUSSION

This case represents a unique presentation of an already rare disease process. Ilhan, Ture, Yilmaz, and Arslan have previously reported PSS with extension from the subclavian vein into the internal jugular vein. They discuss the importance of including PSS in the differential of effort induced upper extremity pain and swelling. Similar to our case report, they were able to achieve a successful with conservative management alone.

Many questions remain as to the aggressive nature in treating exertional venous thrombosis of the upper extrem-

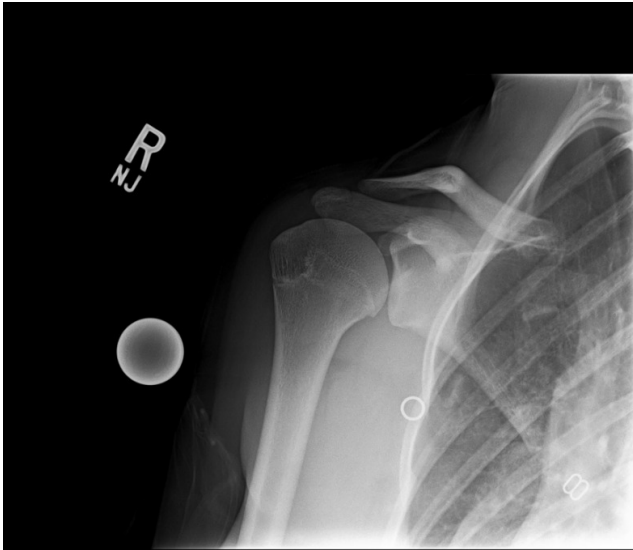


Figure 1

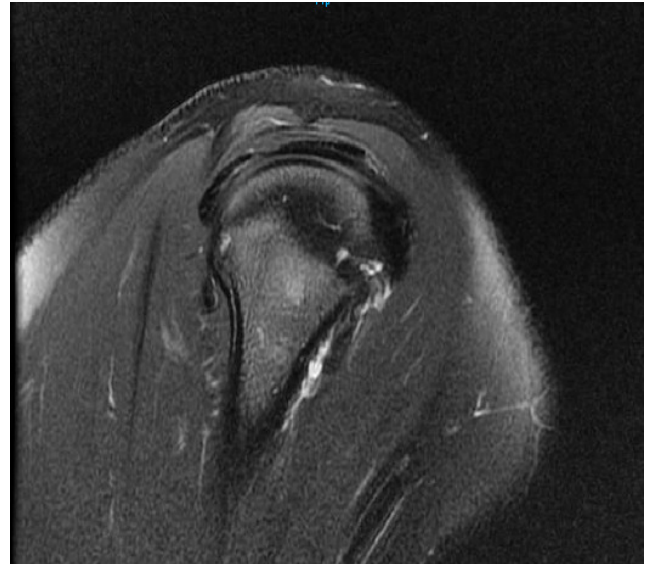


Figure 3

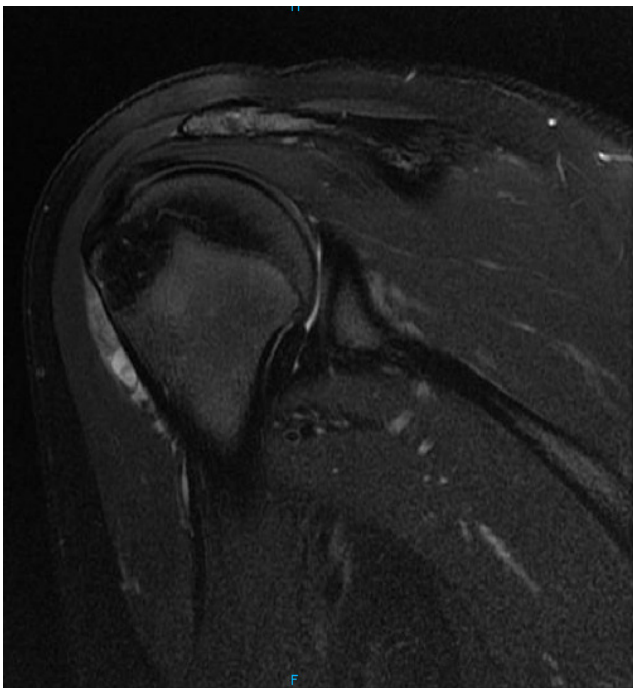


Figure 2

ity. Upper extremity DVTs were once considered benign and self limited although it is now known that they can lead to symptomatic pulmonary embolisms in 9% cases (Mustafa et al. 2003). Early treatment algorithms involving elevation of the upper extremity coupled with anticoagulation resulted in re-occurrence rates and persistent upper extremity symptoms in up to 50% of pts. Machleder was the first physician to recommend surgical intervention. His algorithm consisted of early thrombolytic therapy, a 3 month period of anticoagulation and delayed surgical decompression of the thoracic outlet with transaxillary resection and scalenectomy. His rationale for delayed surgical intervention was on the idea that a recently thrombosed vein would be extremely inflamed and thrombogenic. His theory was

that a delay in surgical intervention after thrombolysis would allow for decreased inflammation and a subsequent recurrence. Urschel and Razzuk have the largest prospective, but not randomized, study examining different methods of treating Paget-Schroetter Syndrome (PSS). Their study, collected over 30 years, included 312 extremities in 294 patients divided into treatment groups ranging from anticoagulant only to thrombolysis and rib resection. Their data supported early diagnosis <4-6 weeks, expeditious thrombolytic therapy, and prompt rib resection as critical steps for best results. On the other hand, Lee et al. reported 27/35 (77%) patients treated successfully with anticoagulation and thrombolysis alone without the need for surgical intervention. These patients remained symptom free at a mean follow-up of 55 months. In the group of recurrence 8/35 (23%), younger age (22 vs. 36) and placement of a stent during initial thrombolysis were considered statistically significant factors prompting recurrence.

Unlike treatment protocols for DVT of the lower extremity, there does not seem to be a unified approach or algorithms in the treatment of UEDVT especially exertional ASVT. To date, there are no randomized controlled trials on the optimal treatment for patients with UEDVT and more importantly PSS. Many institutions have recommended their treatment algorithms (Caparrelli and Freishlag 2005), which almost uniformly follow the need for early thrombolysis and anticoagulation. The discrepancy usually entails when and if surgical intervention is necessary.

Hypercoagulable states and triggering factors including surgery, trauma, immobilization, pregnancy, and use of oral contraceptives have all been linked to LEDVT with a prevalence of clotting abnormalities in approximately 30% cases. There is limited data researching the prevalence of hypercoagulable states involving upper extremity DVTs. In the few journals published, there does not seem to be a clinical link between hypercoagulable states and exertional UEDVT (PSS) but there does seem to be a clinical link with idiopathic UEDVTs. Heron et al evaluated 51 consecutive

patients who had effort related or idiopathic UEDVT. Patients were considered effort induced based on a questionnaire which determined whether or not they engaged on strenuous effort intense activity the week before the thrombosis, regularly played sports consisting of overhead activity, or had a vigorous occupational activity. All patient's plasma was screened for antithrombin, protein C, and protein S deficiencies, and for antiphospholipid antibodies (lupus anticoagulant and anticardiolipin antibodies). Their DNA was also screened for factor V Leiden and for prothrombin gene G20210A mutations. Heron's results indicated that hypercoagulable states appear to play a significant role in idiopathic but not in effort-related UEDVTs with an odds ratio of 4.09 (95% confidence interval, 0.99-16.78; $p=0.06$) in the idiopathic UEDVT group compared with effort related UEDVT group. He concluded that the frequency of coagulation abnormalities in patients with PSS appears to be similar to the normal population while patients with idiopathic UEDVT behaved similar to that observed in primary lower extremity DVTs (Heron et al. 2000). This study appears to be limited to the author's definition of effort induced DVT and does not take into account any thoracic outlet structures or aberrant anatomy that could factor into a number of upper-limb thromboses not associated with strenuous effort. Another study by Martinelli found no additional risk factors in patients with idiopathic primary UEDVT compared to the general population. His study followed 36 patients with primary idiopathic UEDVT, 121 primary LEDVT, and 108 healthy controls. Patients in the control group and LEDVT group were frequency matched by age, sex geographic origin, and social status with patients who had UEDVT. All patients were screened for hypercoagulable states similar to the previous study. Martinelli showed the prevalence of abnormalities of the natural anti-coagulant system (9%) and hyperhomocysteinemia (6%) in patients with primary UEDVT was similar to the both factors in the control group (6% and 7% respectively). This was much lower than the patients with LEDVT (31% and 14% respectively) (Martinelli et al. 2001). Therefore, it is difficult to infer the relevance of a hypercoagulable work-up in patients with primary idiopathic UEDVT but there is some consensus that coagulopathies play a limited if not negligible role in effort induced UEDVT (PSS) and do not necessarily warrant a work-up if one has a detailed history and clinical exam pointing to effort induced thrombosis.

What most journal publications and literature reviews do agree on are the diagnostic parameters leading up to treatment. Clinical assessment of the involved extremity is extremely important in the early diagnosis of PSS and the more common UEDVT. It is important, prior to examination to look for an etiology that could be causal for the thrombosis. The most prognostic indicator of primary UEDVT is a history of strenuous exercise in otherwise young and healthy patients. Secondary UEDVTs are most commonly associated with indwelling catheters, cancer, and pacemakers. On clinical exam, PSS typically presents with, pain upper extremity swelling, venous dilatation, discoloration, and a feeling of heaviness and fatigue. Symptoms are usu-

ally positionally exacerbated whenever there is narrowing of the thoracic outlet i.e. abduction and external rotation (Kahn and Elman 2006; Urschel and Razzuk 2000).

There is no gold standard in diagnosing UEDVT or PSS. Routine chest, C-spine, and shoulder radiographs are routinely obtained first and evaluated for anomalous ribs, pathologic clavicular fractures, and space occupying lesions. Peripheral vascular studies including wrist/brachial index, pulses, and Doppler studies also aid in the diagnosis. According to Prandoni and associates, the lack of compressibility of a vein on the video monitor during ultrasound showed a 96% sensitivity and 94% specificity. The absence of a color signal within the lumen of the vein, or the direct visualization of an intraluminal color-filling defect showed sensitivity of 100% and specificity of 93% for the upper extremity (Mustafa et al. 2003). Venography is both diagnostic and therapeutic and lysis of the thrombus can be performed. Imaging with contrast arteriography or venography have frequently been used to diagnose thoracic outlet syndrome (TOS) with the arms routinely positioned at the side and then hyperabducted in order to evaluate the degree of compression. More recently, magnetic resonance angiography has been used because of its reproducibility, high spatial resolution and short acquisition times, and its ability to be used on obese patients (Hagspiel et al. 2000). A more recent diagnostic test involves paralysis of the scalene muscles using lidocaine. Provocative tests are completed before and after the injection and the patient is asked to estimate (percentage) how much relief they obtained from the injection (Abdollahi and Wood 2009). In summary, diagnostic testing is physician preference and is predicated on a complete clinical exam with clinical suspicion of TOS and or UEDVT.

Our current case falls out of range of many of the case reports, literature searches, and journal articles. Most case reports and literature reviews cite PSS around the axillo-subclavian juncture; our patient presented with thrombosis of the internal jugular vein with no involvement of the axillosubclavian region. At presentation, she clinically did not show any signs of upper extremity venous thrombosis with absence of edema, venous swelling, discoloration, or any provocative exams that could distinguish it from a rotator cuff tendinosis or impingement syndrome. Only her history of repetitive overhead activity and a feeling of weakness/heaviness with prolonged abduction of the arm fit the criteria of current data. It was only an incidental finding on a MRI of the shoulder in the evaluation of her rotator cuff that led to the diagnosis of a hemangioma by the radiologist and eventually the correct diagnosis of complete thrombosis of the internal jugular vein by a vascular surgeon.

A looming question is our patient's diagnosis is what actually caused the thrombosis to form within the jugular vein. We are aware of the causes of effort induced ASVT as mentioned above but can compression of the thoracic outlet also cause trauma or impingement more proximal venous anatomy. Aberrant anatomy including trauma to the clavicle, aberrant first rib, spastic or tight sternocleidomastoid or scalenes could all be a cause of impingement. As

of date, there are no literature reviews or case reports directly linking PSS to jugular venous thrombosis. Virchow's Triad relating to endothelial damage, alteration in blood flow, and coagulation abnormality, can best postulate etiologic factors for the patient's thrombus. The patient's medical history shows no indication that her thrombosis was caused by any medical condition i.e. cancer, coagulation defect, pregnancy, indwelling venous catheter. Given radiographs, magnetic resonance, and clinical exam refute any aberrant anatomy to the clavicle or musculoskeletal structures. Thus, the patient's thrombus was likely caused by impingement or repetitive micro trauma stemming from her strenuous pitching schedule.

There is multiple data on symptomatic primary upper extremity DVTs that present with painful swelling, heaviness, weakness of the affected extremity, and clinical correlation with specific tests. The consensus treatment for symptomatic PE's follows a specific algorithm ranging from anticoagulants, to anticoagulants with thrombectomy and more recently thrombolysis, to anticoagulants and thrombectomy/lysis paired with 1st rib resection. A recent review stressed a unified approach in treating axillosubclavian venous thrombosis consisting of diagnostic studies including duplex ultrasound followed by a diagnostic and therapeutic venogram with lysis of the thrombus. If after successful thrombolysis, compression of the vein is still identified then immediate surgical decompression via rib resection and scalenectomy is offered during the same hospital admission. Patients are discharged home on LMW heparin and scheduled with a repeat venography 2-3 weeks after surgery to reevaluate the patency of the ASV. If the vein has re-thrombosed, then repeat thrombolytic therapy is instituted. This is the main reason for the 2-3 week delay for follow-up venography as any earlier attempt initiating thrombolytic therapy could cause significant morbidity secondary to hemorrhage. If there is venous patency but residual stenosis, then venous angioplasty is undertaken without the placement of stents. Following follow-up venography, the patients are all converted to warfarin, INR 2.5-3.0, for 3 months (Caparrelli and Freishlag 2005). This algorithm offers a unified way to diagnose and treat PSS with multiple checks and balances to make sure the patient remains patent and symptom free. Unlike previous authors who recommend early surgical intervention (Urschel and Razzuk 2000; Brandao et al. 2006), this algorithm does not recommend rib resection and/or scalenectomy unless venography demonstrates impingement or stricturing of the venous anatomy. A suggestion might be to perform a dynamic venogram or a positional venogram in both neutral and elevated or hyperabducted/externally rotated posi-

tions as strictures might be missed if only the former imaging is undertaken.

Contrary to most case reports, our patient presented relatively asymptomatic in terms of UEDVT with no symptoms of edema or swelling. In Urschel and Razzuk's study, all patients presented symptomatic with Adson's sign, hyperabduction sign, or various other compressible signs (Urschel and Razzuk 2000). In our case, special clinical exams did not demonstrate any impingement of the distal vasculature with a documented negative Adson's sign. There were no symptoms of severe neck pain and swelling that is usually associated with symptomatic internal jugular vein thrombosis nor distension of the lower neck and upper chest veins (Urschel sign). Moreover, the patient did not demonstrate any neurogenic symptoms from the cervical structures down to the hand intrinsics. While most data supports early medical and possible surgical intervention for ASVT, should this patient necessarily need anticoagulation therapy, thrombolysis, and early surgical intervention? Sheikh et al cites that the optimal management if internal jugular venous thrombosis must be individualized and depends of the patient's spectrum of concomitant illnesses and recommended early anticoagulation treatment, if no contraindication, to prevent propagation of the clot in the form of a pulmonary embolus. Most authors would agree that IJVT should be treated similarly to a LEDVT with anticoagulation in the form of heparin or coumadin as untreated DVT can lead to PE.

Although not required in this case report, dynamic MRA has been postulated to demonstrate a compressible or constricted vessel with overhead abducted and external rotation movements. Based on these findings the patient can undergo a thrombolysis with or without a scalenectomy and first rib resection; the latter being performed if demonstrated compression on dynamic MRA.

In summary, there are multiple algorithms to diagnose and treat PSS. Every patient with PSS needs to be treated on a case-by-case basis. Although the authors above have tried to demonstrate the correct series of steps to treat exertional upper extremity DVT's, none of the studies have received level one evidence with any statistical significance. Our patient does not need to be aggressively treated as Urschel and Razzuk suggest. Initial attempts at medical management in the non-emergent or asymptomatic patient seems appropriate, or at least a viable option, before more drastic surgical measures, with increasing co-morbidities, are undertaken.

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