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Paget Schroetter Syndrome – in the Context of Work-related Injury: Case Report and Review of Literature

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Abstract

Paget-Schroetter syndrome (PSS) is a primary deep venous thrombosis of subclavian-axillary vein complex after repetitive and strenuous use of shoulders and arms. Here, we report a 24-year-old man who presented with left subclavian and axillary vein thrombosis after loading/unloading labor, diagnosed with PSS via confirmation with Doppler ultrasound, and treated with anticoagulation. By comparing our case with 44 published case reports, we also aim to analyze the patient's characteristics, diagnostic methods, and treatment options for the disease.

Keywords: Upper extremity deep vein thrombosis; Complications; Diagnosis; Therapy; Physiopathology; Ultrasonography; Treatment outcome

Background

Paget-Schroetter syndrome (PSS) or effort thrombosis is a deep venous thrombosis (DVT) of subclavian-axillary vein complex seen in association with repeated strenuous activity of shoulders and arms such as exercises like swimming, wrestling, and gymnastics [1]. While arm exertion is commonly reported in PSS patients, it is important to note that harmless daily activities can also result in PSS [2]. As PSS is seen with activities in the upper extremities, it is often seen in physically active young individuals, especially male, and has a low incidence rate of 1-2 per 100,000 populations [2]. Symptoms include arm swelling, discomfort as well as heaviness and redness of arm, cyanosis of arm, dilated and visible veins on shoulder and upper arm known as Urschel's sign [2-4]. As it is a form of upper extremity deep vein thrombosis, PSS can result in complications including pulmonary embolism and recurrent thrombosis; therefore, it is imperative to consider possible pulmonary embolism in PSS patients [1,3,5]. Here, we report a case of a 24-year-old male with developed PSS after days of repetitive loading/unloading labor and will compare the case with other case reports on PSS.

Case Presentation

This is a 24-year-old right-handed Hispanic male who presented to our occupational medicine clinic complaining of diffuse left upper extremity swelling and pain for the last 10 days. His symptoms occurred one week after he started working as a loader/unloader in a warehouse. He had never done any warehouse job before, and this was the first time in his life to experience these symptoms. His new job entitled constant lifting and moving of boxes from the trailer to the warehouse and vice versa. Many of these boxes were above his shoulder level. He also mentioned he felt a "knot-like" sensation at his left axilla. Examination revealed a well-appearing male with average to slightly muscular body habitus. Vital signs were normal. Generalized swelling was noted over the left upper extremity involving the arm, forearm, and hand (Figures 1 and 2). The left upper extremity also appeared to be erythematous compared to the right one with diffuse tenderness. The left arm circumference was 36 cm (right arm was 33 cm), and the left forearm circumference was 31 cm (right was 29 cm). There were conspicuous dilated veins at the upper left side of the chest (pectoral area) (positive Urschel's sign, (Figures 3 and 4). Radial pulse was intact bilaterally, blood pressure was symmetrical in the two upper extremities and Adson's test was negative. Shoulder range of motion was intact but he had pain with overhead activity, mainly at the axillary area. No palpable mass was identified in the axilla.

The first impression of the case was thoracic outlet syndrome, which is considered a non-industrial injury. Chest X-ray was performed and was negative for cervical rib. The patient was sent for Duplex ultrasound and CT venography scan of the chest to evaluate for possible axillary venous obstruction. The duplex ultrasound showed extensive occlusive acute deep vein thrombosis throughout the left subclavian vein and extensive non-occlusive thrombosis at the left axillary vein. The CT venography of the chest has also confirmed the findings

Figure 1: Patient's arm, more dilated on the left arm

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Figure 2: Patient's arm, more dilated on the left arm.

Figure 3: Urschel's sign on the left shoulder.

Figure 4: Urschel's sign on the patient.

and was negative for any pulmonary emboli. The patient was placed immediately on oral Apixaban. Comprehensive hypercoagulability workup was performed to evaluate for possible etiological agents. The patient's D-dimer was slightly elevated to 0.66 mcg/ml (normal <0.5), screening for Factor V Leiden, antithrombin, Protein S and protein C activity came back within the normal limits. Homocysteine, tissue-plasminogen activator (tPA), cardiolipin, Plasminogen Activator Inhibitor (PAI-I) level and plasminogen activity were all normal. However, Covid-19 Ig G antibodies came back positive. The patient did not recall any recent or past Covid-19 infection or Covid-19-related symptoms. At this point the diagnosis of Paget-Schroeder syndrome (or Effort-induced thrombosis) was confirmed and the patient was sent for urgent vascular surgery consultation for further evaluation and possible intervention. He was placed on restricted duty with no use of the left upper extremity.

By the time the patient made it to the vascular surgery clinic approximately 5 weeks from starting of the symptoms, it was deemed the thrombus is considered sub-acute and that the patient might not benefit from catheter-directed thrombolysis at that point. First rib resection was offered for the patient (based on the center's experience) as an elective procedure. However, the patient decided that he wants to continue conservative treatment with the oral anticoagulant and the job modification.

The patient continued to have improvements in his symptoms. On the 12-week follow up the patient has significantly improved in the swelling of the affected arm and forearm (circumference decreased from 36 cm to 35 cm in the upper arm and from 31 cm to 30 cm in the forearm). Duplex ultrasound was performed and showed improvement of the left subclavian thrombosis which became non-occlusive and showed complete resolution of the axillary venous thrombosis. The patient has changed his job from warehouse worker and enrolled in school for computer programming in the hope of becoming a computer technologist in the future.

Discussion

PSS is a relatively rare condition of thrombosis of axillary-subclavian vein [2] which correlates to strenuous activities of upper libs such as swimming, handball, baseball, and bodybuilding [6,7]. It is thought that PSS is caused by micro trauma of venous intima from repetitive use of UE resulting in local coagulation [8-13].

In this review, we looked at 45 previously reported cases of PSS. Majority of the cases were male (35 males and 10 females) which corresponds to previous reviews that showed that most of PSS patients are male. Also, we found that it is more likely that right arm is affected than left arm or both as we found 26 cases involving right arm [14-30] while we found 15 cases involving left arm and 4 cases involving both arms [31-54]. While we found that the right arm is more likely to be affected by PSS than the left arm, it may have to do with the fact that the majority of the population is right-handed and so it is more likely that the right arm is used more, thereby causing such a finding in our review.

On the literature review, there is an association between PSS and repetitive hyper-abduction and/or external rotation involving shoulders; in our review, there have been association of PSS with various activities using shoulders including exercises like baseball [14,28], swimming [17], judo [19] as well as professions such as carpentry [18], photography [25], furniture lifting [27], waiter job [36], and kebab chef job [55-57]. This is consistent with what our patient had with his profession as his work involved frequent luggage loading/unloading.

Just like our patient who had a complaint of left arm swelling and pain, in the literature review, all the published case reports of PSS demonstrate pain, swelling, and color change of affected arm as symptoms of PSS. Though coldness of the affected arm is a possible

presentation of PSS, there was only one case with the symptom on our review [31]. While the clinical manifestation of PSS is one of factors to consider when making the diagnosis, the clinical symptoms of PSS have poor specificity of less than 50% [2]. Therefore, it would have to involve other diagnostic modalities to come to the diagnosis of PSS.

Ultrasound, being noninvasive and affordable, is the first test for PSS with strong sensitivity of 78-100% and specificity of 82-100% [10], in our literature review, most of the cases were diagnosed with ultrasound as well. While catheter-directed contrast venography is the most accurate diagnostic method and thus the gold standard, it is invasive and reserved for cases where upper extremity DVT was not confirmed from other tests [11]. With the incidence of pulmonary embolism (PE) in upper extremity thrombosis of 5.6%, it would also be prudent to consider CT chest angiography to excluded PE in patients with PSS [12]; in our literature review, there was also a reported case of recurrent PE in the setting of PSS (56) as well as a case of bilateral PE [57] which would raise concern of possible PE with PSS further.

Treatment-wise, anticoagulation is the usual and the standard treatments for PSS; 3-6 months of anticoagulation is recommended for patients with PSS. Catheter-directed thrombolysis is indicated for patients with severe symptoms and effective within 14 days from initial presentation [2]. For long-term prevention, surgical management to decompress subclavian vein such as first rib resection can be considered [3] but the recommendations are weak [1]. In our literature review, the majority of patients just received anticoagulation and the patient in our case only received anticoagulation as his case was too late for thrombolysis and he declined any surgical option. While he had improvement in his symptoms and his left subclavian and axillary vein thrombi with follow-ups, it remains whether he would have recurrence of PSS or not.

Conclusion

While PSS is a relatively rare condition, it can result in serious complications including pulmonary embolism. Therefore, it should be considered as one of the differentials in individuals with arm swelling and pain after repetitive exertion of upper extremities. When suspecting PSS, it is imperative to exclude secondary causes of upper extremity DVT with coagulation workups and confirm presence of DVT with diagnostic modalities. Furthermore, it would be prudent to exclude pulmonary embolism with CT chest angiography given PE is one of detrimental sequelae of PSS. The management of PSS includes not only anticoagulation but also surgical intervention such as resection of first rib and thrombolysis which should be discussed with multiple disciplines including vascular surgery to have long term prophylaxis.

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