

Paget-Schroetter syndrome: the importance of early detection and effective surgical intervention

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How to cite this article: Pejкова S, Aleksovski D, Hadjitrifon S, et al. Paget-Schroetter Syndrome: the importance of early detection and effective surgical intervention. *Arch Clin Cases*. 2023;10(4):200-204. doi: 10.22551/2023.41.1004.10275

ABSTRACT

Paget-Schroetter syndrome (PSS) is relatively rare condition of thoracic outlet syndrome characterized by thrombosis or blood clot formation in the subclavian vein. Due to the non-specific symptoms and low incidence rate, PSS is frequently missed by medical professionals, and as such it often leads to wrong diagnosis and untreated patients. We present the case of a 30-year-old CrossFit trainer who developed a thrombosis of the subclavian vein. Initially, the patient consulted an internist after experiencing swelling in the right shoulder region and discoloration of the right upper extremity. Angiography revealed occlusion of the subclavian vein and anticoagulant therapy was prescribed. For more than a year, the patient's symptoms remained unchanged, and the subclavian vein occlusion persisted. Venography suspected effort thrombosis of the subclavian vein. The patient underwent surgery for decompression of the subclavian vein. After six months, results from post-operative computed tomography angiography showed that venous flow was fully restored and no pathology of the venous vessel wall could be demonstrated. This report aims to increase awareness of PSS among medical professionals, leading to earlier diagnosis and adequate clinical-surgical management.

KEYWORDS: Paget-Schroetter syndrome; thoracic outlet syndrome; effort thrombosis; subclavian vein thrombosis; subclavius muscle resection

INTRODUCTION

Paget Schroetter syndrome (PSS) is a rare condition characterized by thrombosis in the subclavian vein, also known as effort thrombosis of the axillary-subclavian vein [1]. The costoclavicular ligament, which inserts more laterally in most PSS patients, contributes to compression of the subclavian vein [2,3]. It is typically caused by repetitive trauma to the venous wall and subsequent inflammation, leading to blood clot formation. PSS is often seen in athletes, manual laborers, and those who perform repetitive arm motions. This condition has low incidence rate of 2 cases per 100,000 individuals and is more commonly diagnosed in young and healthy males, with a mean age of thirty, and a 2:1 ratio compared to females [4]. Up to 20% of all instances of deep vein thrombosis (DVT) in the upper extremities are caused by PSS [5]. Clinical presentation on the affected arm includes pain, edema, and muscle swelling, often accompanied by visible collateral circulation. A history of exercise or vigorous activity involving the upper limbs is present in 60% to 80% of patients with Paget-Schroetter

syndrome [6]. Early diagnosis and treatment are important to avoid complications such as chronic venous insufficiency and pulmonary embolism.

CASE REPORT

In this case report, we present the clinical case of a 30-year-old CrossFit trainer without comorbidities diagnosed with PSS. The patient has been practicing weightlifting for more than 10 years. Three years ago, he experienced a traumatic event while snowboarding. He fell and landed on his outstretched left hand, resulting with painful restricted motions in his left glenohumeral joint. Following this incident, he sought medical attention from an orthopedic specialist who conducted thorough examinations and radiography. The results indicated that there were no fractures or morphological lesions present. As a result, he was referred to Physical Medicine and Rehabilitation. Following rehabilitation, there was a complete improvement in the mobility of the joint. After a year he noticed a slight swelling and prominent superficial veins in his left upper limb in February 2021. The patient underwent Doppler ultrasonography, which could not confirm an acute thrombotic process in the deep vascular structures of the left arm. However, despite

Received: September 2023; **Accepted after review:** December 2023; **Published:** December 2023.



this, the patient continued with his exercise regimen and noticed a progression of the edema associated with light hyperemia, heaviness, and paresthesia while exercising. On October 12th, 2021, due to these signs and symptoms, the patient sought medical care from a vascular surgeon. He was found to have vein varices in his left arm, with asymmetry between the limbs, light local hyperemia, and radial pulse present without alterations. An angiography of his left upper limb revealed occlusion of the left subclavian vein underneath the clavicle. The vascular surgeon prescribed low-molecular-weight heparin (LMWH) Enoxaparin 4000ie/0.4ml once a day for 2 weeks. He was then referred to a transfusion medicine specialist where laboratory tests were performed, including a complete coagulogram and blood count, which showed only a deviation from the normal range value of international normalized ratio (INR) 1.3 than the enoxaparin was ceased, oral anticoagulant Acenocoumarol 4mg once a day was prescribed and was checked regularly during follow-up appointments over one year by the internist and transfusion medicine specialists. During which time the thrombosis's cause was unknown and the symptoms persisted. For this reason, the internist consulted an interventional cardiologist, Therefore, venography was performed to confirm a suspected deep vein thrombosis (DVT). Venography confirmed presence of intramural thrombosis followed by extraluminal compression in the medial anatomical position between the left clavicle and the first rib (Figure 1A).

The patient was referred to our clinic due to suspicion of effort thrombosis of the subclavian vein. Upon arrival, a physical examination was conducted, and a comprehensive history was taken. He was conscious and denied having fever, chest pain, dyspnea, or any other thrombotic events and comorbidities. The examination revealed swelling in his left arm with light local heat and redness, and the superficial collateral veins were prominent (Figure 2A), but the radial pulse was normal. Blood examination was performed, which included a complete blood count, renal function, liver function, blood coagulation profile, and D-dimer level. Results showed that all values were within their normal ranges. Chest radiography and electrocardiography were also performed, but they showed nonspecific findings. After a thorough clinical evaluation at our clinic and analysis of the radiological investigations, the patient was diagnosed with Paget-Schroetter syndrome, which is also known as effort thrombosis of the

subclavian vein. Subclavian vein decompression surgery was indicated and the patient was scheduled for the operation. In the preoperative phase, the oral anticoagulant therapy previously prescribed was ceased, and 5 days before the surgery the patient was transitioned to prophylactic doses of LMWH-Enoxaparin 4000ie/0.4ml once a day.

The surgery was performed in supine position under general anesthesia. The operation was performed through a subclavicular approach (Figure 3A), where the pectoralis major fibers were opened longitudinally. Subsequently, the hypertrophied subclavius muscle was identified and resected (Figure 3B and 3C). The decompression was made only by resection of the subclavius muscle and tendon. It was noted that the subclavius vein was expanded and completely decompressed (Figure 3D).

After the procedure, we initiated anticoagulation therapy with enoxaparin 60-mg twice per day for 4 days, followed by a switch to 15-mg oral rivaroxaban twice per day until discharge. The patient received regular follow-up at the outpatient department, with an initial frequency of once per week and subsequently reduced to once per month. The patient continued to take rivaroxaban 15 mg twice per day as prescribed at least 6 months.

Postoperatively, the patient had no complications in terms of bleeding, hematoma, or infection. Moreover, throughout the follow-up, the patient did not encounter any additional episodes of venous thrombosis or limb swelling (Figure 2B). Six months after surgery, computed tomography angiography (CTA) was performed, which revealed the absence of intraluminal thrombosis and extraluminal vessel compression. After the results of the radiological examinations and the complete regression of the patient's symptoms, oral anticoagulant therapy was ceased. A new CTA (Figure 1B) was performed in April 2023, which revealed the absence of intraluminal thrombosis and extraluminal vessel compression. One month after the cessation of anticoagulant therapy, the patient exhibits no clinical manifestations suggestive of subclavian vein thrombosis. He persists in fulfilling his professional responsibilities as a CrossFit trainer without any limitations.

DISCUSSION

PSS is a rare type of thoracic outlet syndrome caused by compression or trauma to the subclavian vein. Hughes first

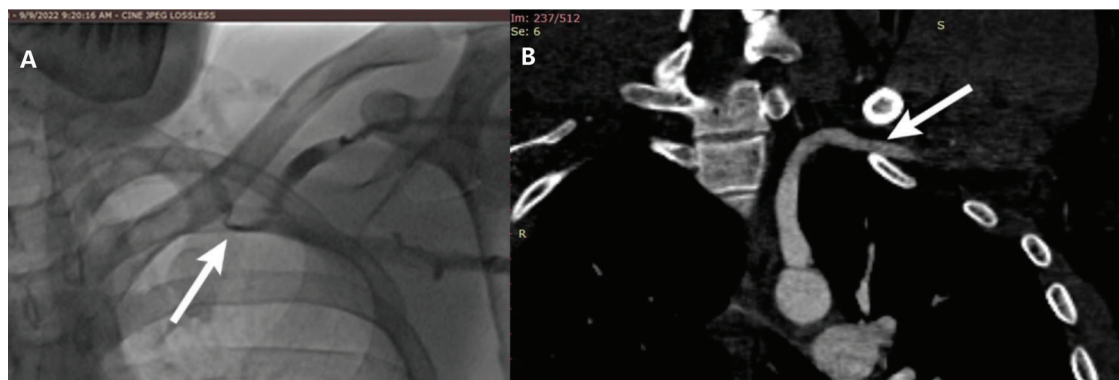


Fig. 1. Presence of intramural thrombosis - Imaging techniques: A) Preoperative venography reveals the presence of intramural thrombosis along with extraluminal compression situated medially between the left clavicle and the first rib; B) Postoperative CTA incorporating venogram demonstrates a complete expansion of the subclavian vein observed five months after the surgical intervention.



Fig. 2. Collateral circulation in the region of the left shoulder: A) Prior to the surgical intervention, there is a marked presence of collateral circulation; B) Following the decompression of the left subclavian vein - there is the absence of collateral circulation.

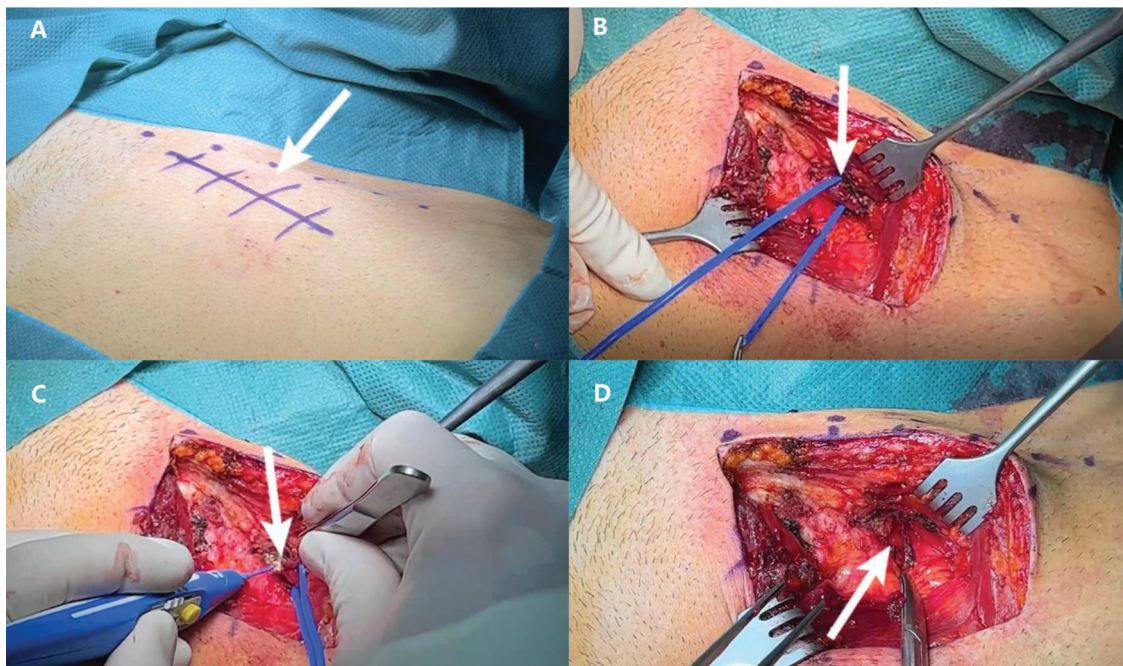


Fig. 3. Surgical steps of subclavian vein decompression: A) Subclavicular approach - the plan for the initial incision, focusing on the subclavian area. The subclavius muscle and the subclavian vein can usually be seen, along with other structures in this region. B) Evident hypertrophied subclavius muscle - a highlight of an enlarged or hypertrophied subclavius muscle. The muscle is likely to be noticeably larger than usual, possibly causing compression of adjacent structures, including the vein. C) Resection of the subclavius muscle - the hypertrophied subclavius muscle is being resected. This procedure is done to relieve pressure on the subclavian vein and other nearby structures. D) Fully expansion of the subclavian vein – this image shows the subclavian vein after the pressure has been relieved by the resection of the subclavius muscle. The vein is likely to be noticeably larger or more expanded as it is no longer being compressed by the muscle.

introduced PSS in 1949, although Paget and Schroetter had both earlier described the interplay between strenuous physical activity causing damage to the axillary vein, leading to deep vein thrombosis of the upper extremity [7,8]. This theory is now supported by further evidence of anatomical abnormalities including a cervical rib, congenital bands, scalenus tendons hypertrophy, subclavius muscle hypertrophy and abnormal costoclavicular ligament insertion. These anomalies can compress the costoclavicular space, restrict venous flow and may lead to chronic vessel wall changes that are conducive to developing a venous thrombus especially after vigorous arm activity. The repetitive trauma to the endothelium leads to inflammation, intimal hyperplasia, and fibrosis, which results in widespread collateral development, venous webs, and perivenular fibrosis [9]. As a result of morphological abnormalities and repetitive endothelial trauma caused by muscle strain, costoclavicular crowding leads to thrombosis initiation and progression. Most of the patients describe a specific trigger, usually arm-intensive sports, which triggers symptoms, but a few may present with chronic symptoms. Symptom onset is acute to subacute usually. Symptoms typically occur quickly or subacutely due to an abrupt event such as sport-related exercise, including swellings, heaviness, redness, and arm discomfort [10,11]. It is often observed to notice enlarged and prominent veins running along the shoulders and upper arms, which is known as (Urschels sign) [2]. Complications that may arise from this condition include pulmonary embolism, recurrent thrombosis, and post-thrombotic syndrome. The diagnosis is usually made by imaging studies, such as ultrasound or venography, which can confirm the presence of a thrombus in the subclavian vein. Compression ultrasonography with color Doppler is preferred and recommended for diagnostic purposes, however, contrast venography should also be considered as a gold standard for diagnosis [12,13]. These conditions should be considered during the differential diagnosis of PSS: neurological thoracic outlet syndrome, arterial thoracic outlet syndrome, tumors in the head, neck, or arm, Pancoast tumor, complex regional pain syndrome, brachial neuritis, compression of nerve roots, quadrilateral space syndrome and compression of nerves [14]. Over the past half-century, a surprising volume of literature has been produced on treating effort thrombosis. Catheter directed thrombolysis has now become the approach, for all patients, with acute effort thrombosis as it has shown better long-term outcomes compared to anticoagulation alone unless there are contraindications. The success rate of catheter-directed thrombolysis is 62% to 84%, but its effectiveness increases dramatically if it is administered soon after the symptoms appear [15]. It is important to understand that thrombosis is a sudden result of an ongoing, chronic condition that involves both external pressure and internal damage to the vein near where the clavicle and first rib meet. While thrombolysis can treat the immediate clot, it does not correct the underlying issues, leaving about a third of patients at risk for thrombosis coming back or continuing symptoms. Therefore, it is worth considering adding surgical decompression of the area near the clavicle and first rib to the treatment plan after thrombolysis. The usual approach, to treat PSS generally includes a combination of anticoagulation treatment and surgical procedures. This may involve decompressing the thoracic outlet, first rib resection, the scalene muscles resection and the resection of the costoclavicular ligament [15,6].

CONCLUSION

Paget-Schroetter Syndrome as a rare type of thoracic outlet syndrome requiring a high suspicion index for diagnosis. Early recognition and treatment can prevent long-term complications such as post-thrombotic syndrome, and surgical intervention can result in the complete resolution of symptoms. Effort thrombosis in the subclavian vein is different from forms of other deep vein thrombosis because it has its own unique pathophysiological attributes, clinical manifestations, and functional implications. Consequently, the effective management of subclavian vein obstruction necessitates treatment considerations specifically tailored to this condition. In conclusion, it is imperative to highlight an emergent trend that has been observed since the advent of the Coronavirus disease (COVID-19) pandemic, characterized by a surge in the incidence of thrombotic disorders, including Paget-Schroetter Syndrome, also known as effort thrombosis of the subclavian vein. This phenomenon warrants meticulous examination and analysis to ascertain the potential correlations and underlying etiological factors that may be contributing to the escalation in the presentation of such thrombotic disorders in conjunction with the COVID-19 pandemic.

Conflict of Interest

The authors declare that they have no competing interests.

Informed Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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