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Case Report

A Paget-Schroetter Syndrome: Case Report and Literature Review

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Abstract

Paget-Schroetter Syndrome (PSS), also known as 'effort thrombosis,' is a form of primary thrombosis affecting the subclavian vein at the costoclavicular junction. It is typically observed in young patients following repetitive and intense physical activities involving the shoulders and arms. Management often involves not only anticoagulation but also catheter-directed thrombolysis, along with first rib resection to prevent recurrence and complications [1]. Physiotherapy and rehabilitation should also be part of the management. This case report of Paget-Schroetter syndrome highlights the importance of anamnesis and physical examination for the appropriate management of the patient.

Keywords: Paget-Schroetter Syndrome (PSS); Urschel's Sign, Thoracic Outlet Syndrome; Subclavian Vein Thrombosis; Pulmonary Embolism; Post-Thrombotic Syndrome (PTS); Thoracic Outlet Decompression (TOD); Axillary-Subclavian Vein Thrombosis (ASVT).

Introduction

Paget-Schroetter Syndrome (PSS), also known as 'effort thrombosis,' is a form of primary thrombosis affecting the subclavian vein at the costoclavicular junction. It is typically observed in young patients following repetitive and intense physical activities involving the shoulders and arms [2].

We are going to talk about a young man of 22 years old, that presented himself to the emergency room, with a problem of pain, swelling and cyanosis of his right upper limb.

The diagnosis of Paget-Schroetter Syndrome was made.

The outcome for the patient was favorable, but this case report highlights a clinical situation that can mistakenly be downplayed and misdiagnosed as a muscle tear. Given the serious complications of an undiagnosed subclavian vein thrombosis, such as pulmonary embolism [3], it is crucial to emphasize the importance of a thorough clinical examination in the emergency room and to avoid trivializing pain that is too quickly classified as muscular.

Case Report

A 22-years-old man visited the emergency department, on the first of December 2024, with complain of swelling and pain of the right upper arm. The pain started five days ago, when he was at the gym, and was carrying heavy loads. He first felt the pain, then his half right upper arm became blue. He did not lose strength or sensitivity of his right upper arm. The symptoms involved after 30 minutes, and came back the morning of his admission at the emergency department, five days later.

His vital signs upon arrival were as follows: Blood pressure 116/65 mm Hg, Heart rate 57 beats per minute, Temperature 36.5°C, Respiratory rate: 18/min, Oxygen saturation: 98%. The patient did not present with dyspnea, cough or hemoptysis.

The clinical examination reveals diffuse edema of the right upper limb and the presence of dilated collateral venous circulation of his shoulder and arm (known as Urschel's sign) (Figure 1 and 2). Cardiopulmonary auscultation was normal. The patient has no notable medical past, and was taking no medication at that time. The patient regularly attended the gym, 5 times a week.

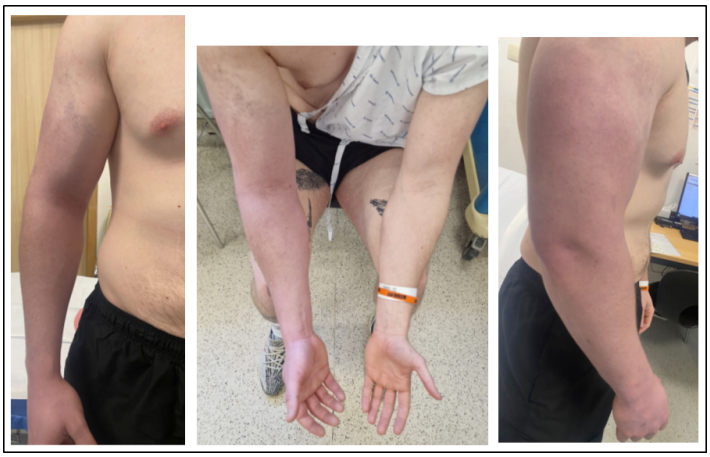

Figure 1: Urschel's sign: Dilated collateral venous circulation of the shoulder and upper limb.

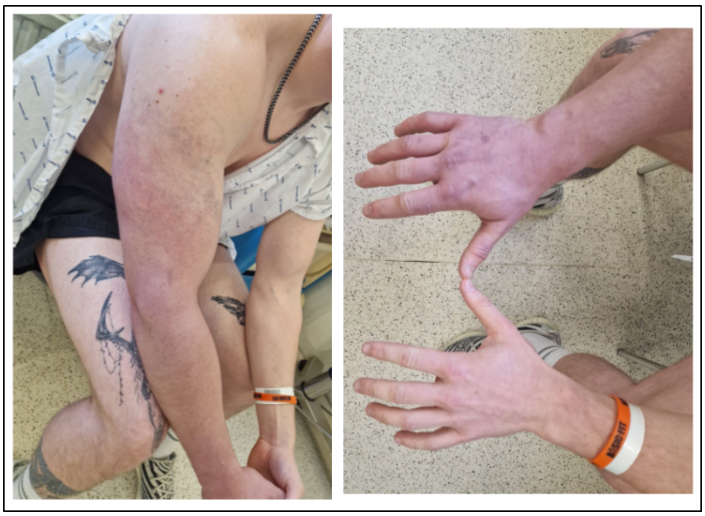

Figure 2: Urschel's sign: Dilated collateral venous circulation of the shoulder and upper limb.

The complementary tests performed included a blood sample with a complete blood count, with particular attention to the platelet count, inflammatory marker levels, and thrombophilia assessment. The results were as follows: There was no significant inflammatory syndrome: CRP: 3,1 mg/L (< 5,0), WC:10,65.10^3/mL (3,64-8,46), platelets were normal: 217.10^3/mL (143-325), but D-dimers were high: 1,896 ng/mL (19-440).

A Doppler ultrasound of the right upper limb was performed (Figure 3 and 4), as the diagnosis of Paget Schroetter Syndrome was suspected: There was no arterial abnormality, no muscle tear or hematoma. There was a venous thrombosis of the subclavian vein, over a length of 10 cm. There was no abnormality of the humeral vein, nor of the various veins in the forearm and wrist.

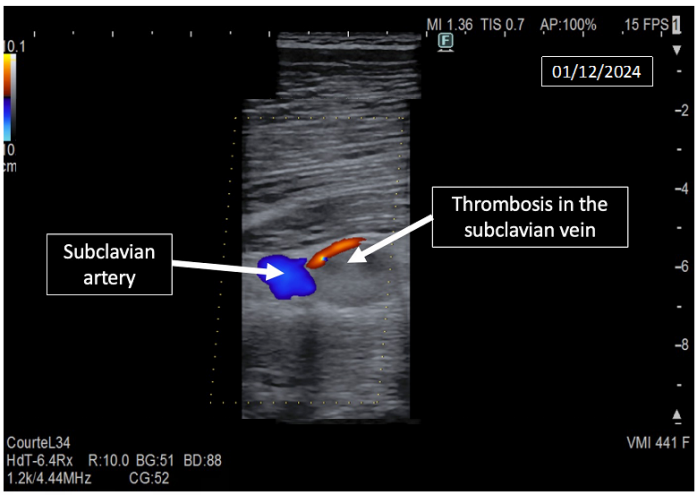

Figure 3: Doppler of the right upper limb showing thrombosis in the right subclavian vein.

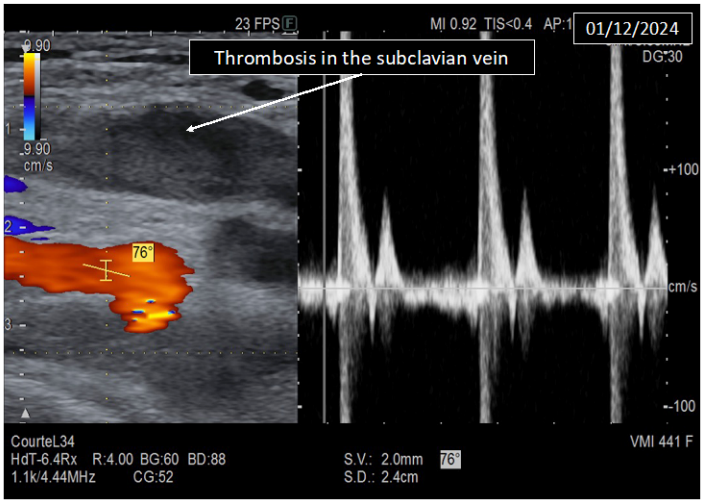

Figure 4: Doppler of the right upper limb showing thrombosis in the right subclavian vein.

The patient underwent a chest CT angiography (Figure 5 and 6), because of the high level of D-dimers, and the recent diagnosis of thrombosis of the subclavian vein, even there was no complain of dyspnea. The CT revealed several small intra-arterial emboli in the subsegmental and segmental regions of the lower pulmonary lobes, and to a lesser extent, in the upper pulmonary lobes.

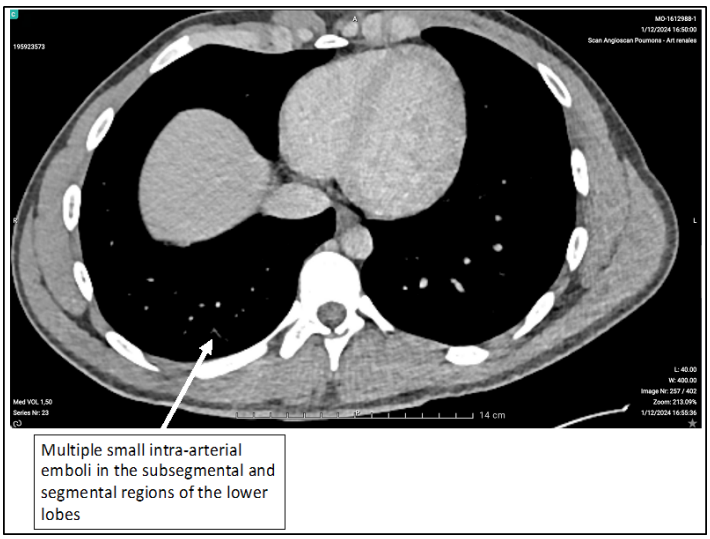

Figure 5: CT angiography showing small intra-arterial emboli of the lower lobes.

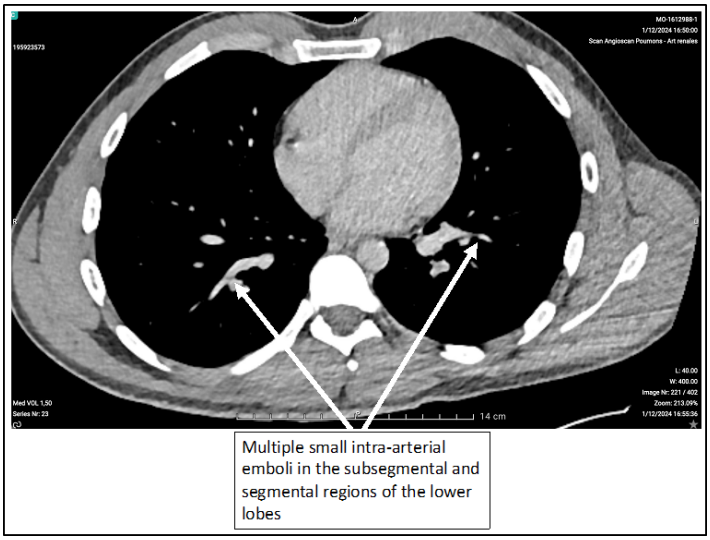

Figure 6: CT angiography showing small intra-arterial emboli of the lower lobes.

The arterial blood gas did not show hypoxemia, and the blood pH was normal. pH: 7.42 (7,35-7,45) pCO2: 38 mm Hg (35-45), pO2: 88 mm Hg (80-105), Oxygen saturation: 97% (95-98), bicarbonates: 25 mmol/L (18-23), lactic acid: 0.51 mmol/L.

So, the diagnosis of Paget Schroetter syndrome (PSS) was confirmed, complicated with a bilateral pulmonary embolism. The vascular surgeon recommended the following treatment: therapeutic oral anticoagulation with Rivaroxaban, wearing an arm sleeve, and raising the arm. The patient was able to return home two days later, after a heart echography that shown no dilatation of the ventricles, and no hearth dysfunction. A follow-up appointment was scheduled for one month later.

The patient was seen one month later (07/01/25) at the hematology consultation. The thrombophilia workup came back negative. The symptoms of pain, swelling and dilatated veins has all involved.

The follow-up Doppler ultrasound performed one month later (08/01/25) (Figure 7) showed an asymmetric thickening of the upper part of the wall of the right subclavian vein. This thickening occupies approximately 50% of the intra-luminal volume. Therefore, the re-canalization was only partial. These wall changes extended over 4 cm in length.

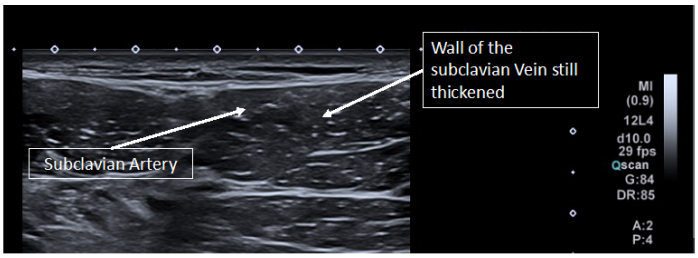

Figure 7: Asymmetric thickening(A) of the upper part of the wall of the subclavian vein.

The patient was seen again at the vascular surgery consultation on 01/16/2025. The anamnesis did not reveal any prior complaints suggestive of thoracic outlet syndrome. The physical examination showed: A warm hand, a capillary refill time of less than three seconds. Palpable humeral, radial, and ulnar pulses, with no changes upon arm elevation. No visible abnormalities in venous return.

The chosen management approach was to continue oral anticoagulation for a minimum total duration of six months, given the context of pulmonary embolism, and to wear elastic arm compression throughout the treatment period. The patient was also given explanations regarding sports activities and the risks associated with thoracic outlet syndrome.

Discussion

Spontaneous Upper Extremity Deep Vein Thrombosis induced by effort, also known as Paget-Schroetter Syndrome (PSS), is a rare condition first described by Cruveilhier in 1816, with a more detailed account provided by James Paget in 1875 [4].

Upper extremity deep vein thromboses (UEDVT) are classified as primary or secondary based on the underlying etiological cause. Paget-Schroetter Syndrome (PSS), also known as 'effort thrombosis,' is a form of primary thrombosis affecting the subclavian vein at the costoclavicular junction. The incidence of effort-induced upper extremity venous thrombosis is 1 to 2 cases per 100,000 individuals per year, occurring primarily in young

adults [1].

Secondary UEDVTs are classified as such due to the presence of a predisposing factor that can cause turbulence in laminar flow, venous compression, or a hypercoagulable state. These factors may include intravascular devices such as implanted medical ports or central venous catheters; however, tumors and other space-occupying lesions have also been implicated in increasing the risk of UEDVT. Most cases of secondary UEDVT occur in elderly patients with pre-existing medical conditions [5].

Physiopathology

Anatomical abnormalities at the thoracic outlet and repetitive trauma to the endothelium of the subclavian vein are key factors in PSS initiation and progression [6]. Repeated endothelial injuries lead to vascular intimal hyperplasia, inflammation, fibrosis, and activation of the coagulation cascade, resulting in thrombus formation [7].

The role of hereditary and acquired thrombophilic disorders remains unresolved and need further investigations. Effort thrombosis preferentially affects the dominant arm [6].

On literature review, majority of case reports show an association between PSS and activities that involve repetitive or prolonged hyper-abduction or external rotation of the shoulder joint, such as: baseball (pitching), violin playing, heavy lifting (TV), horse riding, photography (cameraman), video gaming [7].

Investigation

The symptoms and clinical signs of Upper Extremity Deep Vein Thrombosis (UEDVT) have low specificity (pain, swelling of the upper limb…). Therefore, it is crucial to perform confirmatory tests after a presumptive clinical diagnosis [6]. An important sign to look for is the presence of dilated superficial veins (Urschel's sign) (Figure 2).

Doppler ultrasound is the first-line examination and is also a useful tool for emergency physicians to make a rapid diagnosis if positive signs of venous thrombosis are observed. However, ultrasound can yield false-negative results due to the difficulty of accessing non-compressible areas of the subclavian vein just beneath the clavicle.

Therefore, it is important to perform magnetic resonance venography (MRV) or computed tomography (CT) if Doppler ultrasound is negative in cases with a high index of suspicion [8].

Contrast venography remains the gold standard for diagnosis. However, due to its invasive nature and exposure to ionizing radiation, alternative imaging tests are preferred [9].

Complications

A delay in diagnosis and treatment can lead to serious complications such as post-thrombotic syndrome (chronic pain, venous hypertension, swelling, and ulceration of the limb), recurrent thrombosis, septic thrombophlebitis, and pulmonary embolism [10].

Management

Effort thrombosis should ideally be managed with a multimodal approach, including routine catheter-directed thrombolysis, early thoracic outlet decompression in appropriate patients, as well as physical and occupational therapy [6].

Conclusion

Paget-Schroetter Syndrome is a particularly timely topic, given the recent case of French NBA player Victor Wembanyama, aged 21 and standing 2.22 meters tall. Playing for the San Antonio Spurs, he announced on February 21, 2025, that he would be sidelined for the remainder of the season due to a deep vein thrombosis (DVT) in his right upper limb. The standard treatment, which typically involves anticoagulant therapy, prohibits participation in contact sports like basketball. This isn't the first time a basketball player has faced such an issue; in 2019, American player Brandon Ingram also suffered from a deep vein thrombosis in his right shoulder [11].

A conservative management approach with anticoagulation alone is insufficient and results in significant residual disability. An aggressive multimodal treatment strategy, including catheter-directed thrombolysis with or without early thoracic outlet decompression, is essential to optimize outcomes. Despite a better understanding of its pathogenesis and therapeutic advances, a significant number of patients with Effort Thrombosis continue to receive suboptimal treatment [6].

Conflict of Interest: The authors declare that they have no competing interests.

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