Case Report



Beyond Rhabdomyolysis: Uncommon Challenges in Clinical Practice

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Abstract

Paget-Schroetter Syndrome (PSS) is a rare form of upper extremity deep vein thrombosis (UEDVT), commonly associated with repetitive arm movements or anatomical abnormalities of the thoracic outlet. Due to its rarity, it can be overlooked, leading to delayed diagnosis and treatment. The authors present the case of a young healthy male, who developed right upper limb pain and swelling, following an intense calisthenic workout and an 18-hour flight. Initially diagnosed and treated for rhabdomyolysis, further evaluation revealed painful asymmetry in the right arm. A venous Doppler ultrasound and subsequent Computed Tomography (CT) venography confirmed Paget-Schroetter syndrome. This case highlights the importance of considering this rare entity, a subset of thoracic outlet syndrome, in young males presenting with post-exertional upper limb pain, particularly in the dominant arm.

Keywords: Thoracic outlet syndrome, Paget-Schroetter syndrome, CT venography, rhabdomyolysis.

Introduction

The Paget-Schroetter syndrome, also called effort-induced venous thrombosis, is a spontaneous venous thrombosis usually due to repetitive physical activity or strenuous efforts, often involving the upper limb. It commonly affects athletes and is a rare cause of thoracic outlet syndrome. This condition leads to pain, swelling, redness in the arm and, if left untreated, may cause complications such as post-thrombotic syndrome.

Thoracic outlet syndrome (TOS) ^[1] presents a diagnostic challenge in clinical practice. It results from anatomical variations affecting the transition between the thorax and the upper limb, potentially compressing the brachial plexus, subclavian vein, or subclavian artery. The most common form involves neural compression, followed by venous involvement, with arterial compression being the least frequent. Paget-Schroetter syndrome is a subset of the venous group ^[2]. Certain tumors, such as those in the head, neck, or upper limb, and Pancoast syndrome, may mimic this condition, adding to the diagnostic complexity.

The first description of spontaneous axillary-subclavian venous thrombosis was by Cruveilhier in 1816, with a more detailed account provided by James Paget in 1875. In 1894, von Schroetter identified vascular trauma from muscular tension as a possible etiology. The term "Paget-Schroetter Syndrome" (PSS) was introduced in 1948 by Hughes ^[3].

Epidemiologically, PSS accounts for 30-40% of spontaneous axillary-subclavian vein thromboses and 10-20% of all

upper extremity deep vein thromboses (UEDVTs). It predominantly affects young males, with a male-to-female ratio of 2:1, typically presenting around age 30. The dominant limb is involved in most cases, and 60-80% of patients report a precipitating event, commonly related to sports or occupational exertion ^[1].

Case Report

A 34-year-old male, with neither relevant medical history nor regular medication, denied substance or supplement use. He presented to the emergency department (ER) with swelling, redness, warmth, and pain in the right upper limb, with no additional complaints. Five days before, he had engaged in an intense calisthenic workout in California, followed by an 18-hour flight back to Portugal.

Initial laboratory tests revealed increased values of hepatic enzymology and creatine kinase (CK). Rhabdomyolysis was initially diagnosed and managed accordingly. However, worsening asymmetry of the right upper limb and increasing pain with palpation and movement prompted further evaluation.

A third laboratory assessment included D-dimer testing, revealing an elevated level of $551.0 \ \mu g/L$ (Reference: $<230 \ \mu g/L$), alongside a general worsening of laboratory markers except for CK. Given the clinical and laboratory deterioration, vascular surgery consultation was sought for venous thrombosis screening via Doppler ultrasound. Findings included: Tritonal flow in the radial and ulnar arteries; reduced superficial venous system filling; poorly compressible axillary vein; Irregular flow in the subclavian vein.

A contrast-enhanced CT angiography (arterial and venous phases) of the right upper limb was performed. The arterial phase revealed normal vascular caliber and density, while the venous phase

indicated a suspected thrombus in the right subclavian vein (Figure 1 & 2). No signs of pulmonary embolism were detected.



Figure 1: Coronal cut of the CT-Scan showing engorged (thrombus) right subclavian vein (red circle), more than the superior vena cava.



Figure 2: CT axial cut showing marked asymmetry between the two subclavian veins (red arrows).

After multidisciplinary discussion, conservative management was chosen and therapeutic enoxaparin was given, being later transitioned to apixaban according to vascular surgery guidance. During hospitalization, there was a significant clinical and analytical improvement (see Table 1). During this period, an etiological study was conducted and inherited thrombophilias, immunological testing and Antiphospholipid Syndrome yielded negative results.

Table 1: Complement	tarv diagnostic te	sts during hospitalization

Laboratory results	At admission (ER)	Reassessment	1 st day Hospitalization	At discharge
AST (U/L)	745	696	739	22
ALT (U/L)	134	158	206	19
LDH (U/L)	1510	835	555	166
CK (U/L)	44703	37345	26993	91

A thoraco-abdomino-pelvic CT scan ruled out neoplasia, including thoracic tumors, thereby eliminating the need for further invasive studies. The patient was discharged on a six-month course of apixaban and followed up in vascular surgery consultation. A sixmonth follow-up Doppler ultrasound confirmed normal venous patency without residual thrombus or reflux. The patient was advised to use prophylactic enoxaparin (40 mg subcutaneously) before long flights.

Discussion

Effort-induced venous thrombosis, refers to subclavian-axillary vein thrombosis associated with strenuous and repetitive upper limb activities. Key contributing factors include anatomical abnormalities of the thoracic outlet and repetitive trauma to the subclavian vein endothelium. The condition typically follows intense sports or occupational activities involving vigorous, sustained upper limb movements.

Patients commonly present with swelling, pain, redness, warmth, and functional impairment of the affected limb. In severe cases, cyanosis and Urschel's sign (venous engorgement) may be observed. Retroversion, hyperabduction, and extension of the affected arm during strenuous activities may stretch the subclavian vein, leading to endothelial microtrauma and coagulation cascade activation. Evidence suggests anatomical variations in the thoracic outlet (e.g., cervical ribs, congenital fibrous bands, scalene muscle hypertrophy, and abnormal costoclavicular ligament insertion) play a role in disease pathogenesis ^[3].

The most frequent complications of PSS include pulmonary embolism, post-thrombotic syndrome (characterized by pain, heaviness, and limb swelling), and recurrent thrombosis.

The availability of thrombotic agents, combined with prompt surgical neurovascular decompression of the thoracic outlet, has reduced morbidity and necessity for thrombectomy, substantially improving clinical results ^[4].

Conclusion

Although rare, subclavian vein thrombosis should be considered and ruled out in young men presenting with upper limb pain and asymmetry. If confirmed, anticoagulation should be promptly initiated. Referral to vascular surgery is essential, as some patients may require surgical intervention, particularly given the substantial incidence of residual symptoms, functional impairment, and recurrent thrombosis with conservative management.

Declarations

Ethics approval and consent to participate

"Not applicable"

List of abbreviations

CK: Creatine Kinase CT: Computed tomography ER: Emergency department PSS: Paget-Schroetter Syndrome TOS: Thoracic outlet syndrome UEDVT: Upper extremity deep vein thrombosis

Data Availability

"Not applicable"

Conflicts of Interest

"The author(s) declare(s) that there is no conflict of interest regarding the publication of this paper."

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Authors' contributions

PDM, research design, data collection drafting of the paper. TMC, DFS, SMM, RJA, ISG, research, design and data collection. LFV for the intellectual content revision.

All authors read and approved the final manuscript.

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