Case report

Paget-von Schrotter syndrome: Case report

Josias Torres de Siqueira Filho a,*, Railane Lima de Paula b, Arthur César Albuquerque Régis a, Alberto Rubin Figueiredo a, Bruno von Mühlen c, João Victor Fuzeta Peres d

a Department of General Surgery, Getúlio Vargas University Hospital (HUGV), Manaus, Brazil
b Department of Medicine, Nilton Lins University (UNL), Manaus, Brazil
c Department of Urology, Western Regional Hospital, Chapecó, Brazil
d Department of General Surgery, Regional Hospital of Presidente Prudente, São Paulo, Brazil

A R T I C L E   I N F O

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A B S T R A C T

Introduction: Paget-von Schroetter Syndrome is a rare condition, which refers to primary venous thrombosis of the subclavian-axillary bed. It is related to vigorous activities involving the upper limbs, presenting pain, edema and muscle swelling. Its diagnosis involves, besides the clinical suspicion, Doppler ultrasonography and should be performed early to ensure immediate treatment. The therapy is initially made with anticoagulation, but thrombolysis, decompression surgery, venolysis and venoplasty should be considered, which can lead to a better prognosis and lower risk of complications.

Case report: This article refers to a report of a male patient, with a history of physical activity, who developed thrombosis in the left upper limb and evolved, after anticoagulant treatment, to post-thrombotic syndrome.

Discussion: Paget-von Schroetter syndrome can be asymptomatic, but common features include hyperemia, edema, heaviness and pain in the affected arm, usually 24 h after the initial event and may be accompanied by low fever. In diagnostic investigation, imaging exam is essential. Early diagnosis of the syndrome allows the early start of treatment, leading to better results and prognosis.

Conclusion: Therefore, this study has the goal of making the diagnosis more effective and improves the clinical-surgical management, from the increase in the level of medical professionals' suspicion regarding the disease.

1. Introduction

Paget-von Schroetter Syndrome is a rare condition, which refers to primary venous thrombosis of the deep veins that drain the upper extremities of the subclavian-axillary territory, due to intense and repetitive activities of the limb [1–3]. Also known as effort thrombosis, it has an incidence of 2 cases per 100,000 inhabitants [4], with a predominance in young and healthy men, with an average age of 30 years, in a ratio of 2:1 when compared to women, and more frequent in the right territory. In 60% to 80% of patients, there is a history of exercise or vigorous activity involving the upper limbs, with a classic clinical picture of pain, edema and muscle swelling, which may be associated with visible collateral circulation [5].

Due to the high risk of morbidity and mortality and the importance of an early diagnosis and treatment, this study aims to report and discuss the clinical case of a 24-year-old patient with primary venous thrombosis in the left subclavian vein, without a previous personal and family history of venous thrombosis, who developed residual symptoms after completing the treatment. This study has the goal of making the diagnosis more effective and improves the clinical-surgical management, from the increase in the level of medical professionals’ suspicion regarding the disease.

2. Methods

This work was reported according to SCARE 2020 criteria [6].

3. Case report

An 27-year-old man, resident of Manaus, without comorbidities, right-handed, started in December 2016, at the age of 24, the regular and intense practice of weight training, especially in the upper limbs. On November 1st, 2017, he noticed a slight edema of the left upper limb, and asymmetry between the upper limbs. He reports having continued...
the exercises with progression of the edema, which became intense, associated with hyperemia, heaviness and paresthesia (Figs. 1 and 2).

On November 4th, due to signs and symptoms, the patient sought medical care at a local hospital, with significant edema in the left arm, highlighting asymmetry between the limbs, local hyperemia and pulse present without alterations. He performed Doppler ultrasonography (Fig. 3), which showed a thrombus in the left subclavian vein. Laboratory tests were performed, including complete coagulogram and CBC, ruling out a prothrombotic state. It was prescribed the use of compression stockings in the left arm, suspension of weight training for 4 months, and Rivaroxaban 20 mg, for 6 months, concomitantly.

At the end of the treatment, in April 2018, the limb remained with mild edema and angioresonance was performed (Fig. 4), which highlighted a reduction in the caliber at the origin of the left subclavian, with parietal thickening and contrast enhancement (possible fibrosis), it should correspond to chronic recanalized thrombosis. It was decided to extend the use of Rivaroxaban for another 6 months. During this period, he was diagnosed through a new Doppler ultrasonography, with Post-Thrombotic Syndrome, with recanalized thrombosis, noticing a slight irregularity on the internal surface of the vessel, configuring a complication of the pathology.

At the end of 12 months of treatment, with the diagnosis of post-thrombotic syndrome, the patient presented progressive improvement of symptoms, with no other approaches being indicated, such as subclavian vein venogram, venous stent or first rib resection.

4. Discussion

Paget-von Schroetter Syndrome is an uncommon cause of DVT. It occurs mostly in young people, with an average age of 30 years, healthy and athletic, with a higher incidence in men and, in 80% of cases, the dominant arm is affected. The outstanding feature is a lifestyle that involves repetitive and vigorous movements of the upper limbs [3,7–9], mainly abduction, external rotation and hyperextension of the shoulder and neck [4]. In the report presented, the patient is male, 24 years old, and performed such movements, in addition to lifting weights, adding more strength to the exercise, increasing the risk [4].

Currently, it is proposed that its pathogenesis involves mechanisms that develop thrombus after repetitive trauma. In addition to the hypertrophy of the scalene, subclavian and pectoralis minor muscles, which causes extrinsic compression of the vein [7], there is an association with repetitive trauma resulting from exercise, which damages the venous wall. Repetitive endothelial damage results in vascular intimal hyperplasia, inflammation, fibrosis and activation of the coagulation cascade, leading to thrombus formation [9,10].

Paget-von Schroetter syndrome may be asymptomatic, but common features include hyperemia, edema, heaviness and pain in the affected arm, usually 24 h after the initial event and may be accompanied by low-grade fever [8]. As a differential diagnosis one can consider cellulitis, lymphedema, muscle trauma, neoplastic venous compression and superficial venous thrombosis. The presence of dilated collateral veins around the shoulder, known as Urschel’s sign, despite not having been observed in the patient, is characteristic of the syndrome and should raise clinical suspicions of vascular etiology [10].

In the diagnostic investigation, the clinical history, physical
Resection of the first rib has been described by high posterior thoracotomy as one of the 3 pillars of the treatment of Paget-von Schroetter syndrome. Relief of the extrinsic compression of the SV by resecting the first rib forms part of the diagnosis and a hypercoagulable state was suggested. In the present report, erroneously, there was no evolution in the recommended line of treatment and the patient remained symptomatic, being diagnosed with post-thrombotic syndrome [16]. Until the time of the study, he was under clinical supervision and continuous to prevent new episodes.

5. Conclusion

Paget-von Schroetter Syndrome, although rare, should be considered by physicians in healthy patients with complaints in the upper limbs, especially in those with a history of repetitive movement of the upper limbs. After complete anamnesis of the patient with a suspected diagnosis, exams should be performed, with Doppler ultrasonography being the main one, to confirm the diagnosis and exclude other etiologies, in order to guarantee the immediate start of treatment. This must be well applied, according to each patient, for a good recovery and minimal morbidity and mortality.

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Ethical approval

As the manuscript is not a research study, we only have the patient consent for writing and others forms of publication. Also, the ethical approval for this case report has been exempted by our institution.

Informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Research registration

N/A.

Guarantors

JTSF.

Provenance and peer review

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CRediT authorship contribution statement

ACAR and ARF made contributions to conception and the review of literature. JTSF collected the patient details. JTSF and RLP wrote the paper. BVM and JVFP made contributions to patient management. JTSF, RLP and ACAR critically revised the article. All authors read and approved the final manuscript.
Declaration of competing interest

JTSF, RLP, ACAR, ARF, BVM and JVFP declare that don’t have any conflict of interests.

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