

Paget-Schroetter syndrome: case report

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Abstract

Introduction: The Paget-Schroetter Syndrome is one in which the compression of the axillary and subclavian veins generates thrombosis, leading to blood stasis in the venous system of the upper limb and its respective complications. First-line treatment focuses on thrombolysis and anticoagulation of patients. **Case presentation:** A 23-year-old female patient attended the emergency department due to edema and erythema of the right thoracic extremity for 1 day, as well as pain in the axillary region when moving the arm and infraclavicular pain on palpation, and paresthesia in the fingers, that started 2 hours before consultation. **Conclusion:** However, the debate continues as to whether the resection of the first rib as a decompression method should be applied to all patients or whether, depending on the characteristics of each patient, it is possible to follow other regimes to avoid the recurrence of symptoms and even subsequent thrombosis episodes.

Keywords: Thrombosis. Anticoagulants. Thrombolysis. Decompression.

Introduction

Paget-Schroetter syndrome is a compression of the subclavian and axillary veins that triggers thrombosis of the affected veins. This is generated due to the decrease in space between the first rib, the clavicle, the scalene muscle, the subclavian muscles, and the costocoracoid ligament^{1,2}. Venous thrombosis in these regions accounts for 2-4% of all cases of venous thrombosis deep, with an incidence of 11 of every 1,000,000 hospital admissions². This syndrome is responsible for 10-20% of deep vein thromboses of the thoracic extremities and 30-40% of spontaneous thromboses of the axillo-subclavian veins^{3,4}.

The pathophysiology is due to an injury to the vascular endothelium, which triggers the coagulation cascade until a thrombus forms within the lumen and the flow is obstructed³⁻⁵. This damage is generated when performing physical effort with the shoulder, which is why it is observed in young, previously healthy, and physically active patients^{1,3-6}. Repeated damage generates perivascular fibrosis, which perpetuates the stenosis of these veins⁴.

The clinical signs observed are a sensation of heaviness of the affected limb, pain, edema, erythema, cyanosis, collateral venous network, and visibly dilated veins in the shoulder (Urschel sign). Most patients present symptoms within the first 24 h after exertion^{1,3,4,6}.

The imaging study to be performed in the first instance is Doppler ultrasound, with a sensitivity of 78-100% and a specificity of 82-100%. However, the gold standard is 1.4 contrast venography. Subsequently, phlebography should be performed to observe thrombosis of the subclavian and axillary veins, as well as the collateral branches, and a provocation maneuver that consists of abducting the limb to observe flow obstruction 1 must be performed. Among the differential diagnoses are found: cellulitis, lymphedema, neoplastic compression, and superficial thrombosis³. It is advisable to evaluate other possible causes of hypercoagulability³.

The first step of treatment consists of anticoagulation followed by catheter-directed intrathrombolysis using alteplase or reteplase within the first 10 days^{1,4,6}. It is suggested that no more than 2 weeks have passed

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from thrombus formation to the procedure since this is associated with a success rate of 75-84%, while if it is performed between 2 and 12 weeks this rate decreases to 29%^{4,7}. Likewise, the patient must be anticoagulated for 3-6 more months with low molecular weight heparin (LMWH) and subsequently with Vitamin K^{1,6} antagonists. More than 90% of patients manage to return to their daily activities.

Resection of the first rib by transaxillary, supra, or infraclavicular resection is recommended by multiple authors, although a claviclectomy can also be performed, since thrombosis after thrombolysis is generated in up to a third of cases^{1,6,7}. Finally, angioplasty is performed and a stent is placed if considered necessary^{1,2}. Complications of thrombosis include pulmonary thromboembolism, post-thrombotic syndrome (associated with conservative treatment with anticoagulants), recurrent thrombosis and perivascular fibrosis, the latter occurring in 10-12% of patients²⁻⁴.

Case report

A 23-year-old female patient attended the emergency department due to edema and erythema of the right thoracic extremity for 1 day, as well as pain in the axillary region when moving the arm and infraclavicular pain on palpation, and paresthesia in the fingers, that started 2 hours before consultation. In previous days, the patient reported having a feeling of heaviness in her right thoracic extremity after lifting weights in the gym.

The patient reports having been lifting weights for 4 years, using an oral contraceptive with estradiol and dienogest for 1 year, changing this method to a dermal patch with ethinyl estradiol and norelgestromin 1 month before the development of symptoms. As a family history, his father developed deep vein thrombosis of the left pelvic extremity after spinal instrumentation.

On physical examination, the patient presented edema and erythema of the right thoracic extremity, collateral and dilated veins in the shoulder, pain along the brachial and axillary veins when compressing the forearm, decreased radial pulses, and capillary refill lasting 4 s. She was administered 1.5 mg/kg/24 h of LMWH as the first approach and the next day, a Doppler ultrasound was performed where thrombosis of the axillary and subclavian veins was found. Conservative therapy was started with rivaroxaban 40 mg per single dose and subsequently 20 mg every 24 h, however, the condition deteriorated with the onset of dull pain in the right suprascapular region which gradually increased in

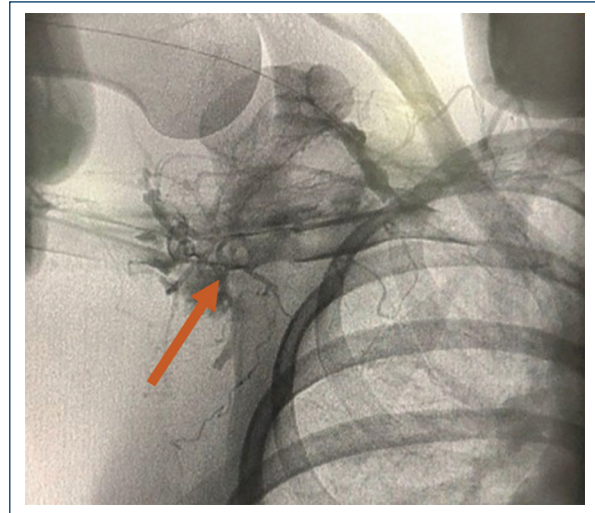


Figure 1. Obstruction of the passage of contrast medium during catheter placement for thrombolysis of axillary and subclavian veins (arrow) due to the presence of thrombus and its passage through the developed collateral circulation.

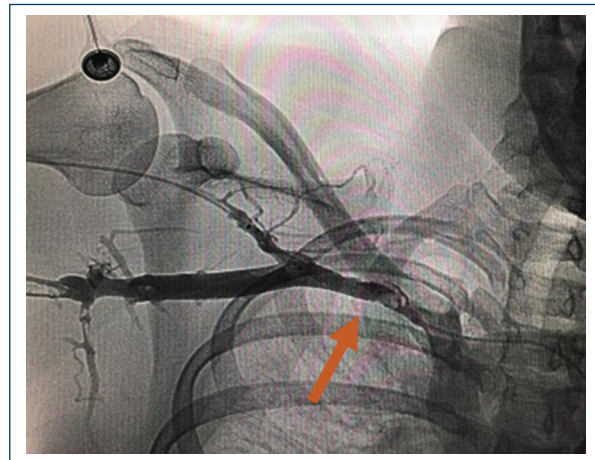


Figure 2. Passage of contrast medium through axillary and subclavian veins free of obstruction after thrombolysis. Tortuosity (arrow) is seen in the path prior to performing balloon angioplasty.

intensity and radiated through the body. The dorsum until it reaches the ipsilateral lung base and becomes transfection, preventing deep inspiration.

A Doppler ultrasound was performed again, which confirmed deep vein thrombosis of the subclavian and axillary veins with data of brachial, ulnar, and radial venous insufficiency. Subsequently, a computed tomography angiography was performed where pulmonary thrombosis of the right posterior basal segmental

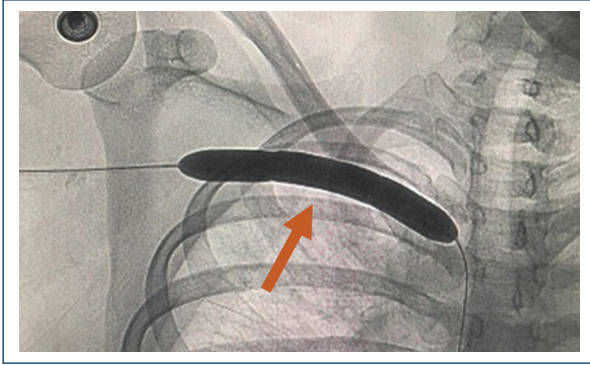


Figure 3. Balloon angioplasty (arrow) of axillary and subclavian veins after thrombolysis and verification of viability of venous circulation by passing contrast medium.

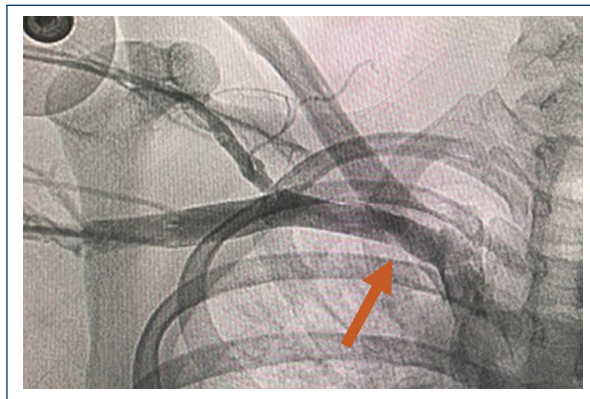


Figure 4. Axillary and subclavian veins after thrombolysis and angioplasty where complete viability (arrow) is observed when contrast medium is used.

branch was found, an area of condensation in the right posterobasal segment probably related to pulmonary infarction and posterobasal linear atelectasis.

As a treatment, catheter-directed intrathrombo thrombolysis with alteplase as seen in [figure 1](#), was used. The veins affected were tortuous and narrow, as can be appreciated in [figure 2](#). Subsequently, balloon angioplasty ([Fig. 3](#)) was performed where the absence of obstruction within the veins was confirmed, as shown in [figure 4](#). Due to the favorable evolution of the treatment, she was discharged from hospitalization and a follow-up appointment was scheduled one month after the procedures. During this time, he did not present obstructive symptoms, and when a Doppler ultrasound was performed 30 days after hospital discharge, compressible veins were observed without compromised flow, so it was not necessary to perform decompressive surgery.

Discussion

Silverberg et al. determined that the exclusive use of oral anticoagulant therapy is associated with post-thrombotic syndrome in 44% of Cases⁸. In this retrospective study, 18 patients who did not receive surgical treatment to resect the first rib were followed, and determined residual symptoms using the villalta scoring system, in which it was observed that 94% had minor symptoms. Likewise, the Quick-DASH system was used to evaluate the impact on quality of life, where 83% of the patients did not present any alteration in their quality of life and all managed to return to their work activities, and an ultrasound was performed to evaluate the viability of the brachial, axillary and subclavian veins, demonstrating that 89% of patients maintained the patency of the affected vein, even years after thrombolysis⁸.

An athlete presented thrombosis on two occasions treated with anticoagulation, due to having a factor V Leiden and PT20210 mutation. Anticoagulant treatment was maintained for 6-12 months in patients without thrombophilias and for life in those with a related condition. Therefore, they determined that the majority of patients who do not receive surgical treatment remain asymptomatic, although slight circulatory changes may be observed. It is important to consider that surgical decompression can promote brachial palsy, chronic pain, chylothorax, phrenic nerve paralysis, arterial injury, and hemorrhage in 5-15% of patients⁸.

Lee et al. formulated an algorithm where only those patients who remained symptomatic upon subsequent visits received surgery. In their study, 23% of patients who did not receive surgical treatment developed thrombosis, but the time they remained anticoagulated was only 5 months, compared to the patients studied by Silverberg et al., where the average treatment time was 26 months^{8,9}.

Lee et al. conducted another study where he found that of 22 patients with upper limb thrombosis (only nine remained without surgical treatment), eight of these had minimal symptoms after anticoagulant treatment and only one persisted with moderate symptoms although he did not present obstruction. Eleven of 13 patients who underwent decompressive surgery continued to partially present the initial symptoms to a lesser extent and two other patients remained with the same degree of symptoms. Lee and his group point out that not all patients should be treated surgically and that they should be followed up while they are undergoing treatment to determine if decompressive surgery is required, as it exposes the patient to multiple risks¹⁰.

However, in a meta-analysis carried out by Karaolanis et al., where the majority of patients received surgical intervention for resection of the first rib, complications associated with surgery were $\leq 6\%$, and are related to a greater rate of vascular viability and lower symptoms compared to patients who received anticoagulation and thrombolysis, which is why it is recommended in patients with low or intermediate surgical risk¹¹.

Likewise, Hange et al. determined that anticoagulation is less effective than thrombolysis and decompressive surgery. The results of thrombolysis are superior if it is performed within the first 2 weeks, but without mechanical decompression, there may be a relapse in a third of patients, which is why surgical treatment is recommended in patients who are candidates⁷.

Conclusion

Despite the great advances regarding the definitive treatment of thoracic limb thrombosis, it is still necessary to carry out more studies with a large number of participants that compare the effectiveness of the long-term use of oral anticoagulants and resection of the first rib after thrombolysis. Likewise, a study is considered necessary to evaluate different variables that patients may present, such as antiphospholipid syndrome, mutations, or protein deficiencies that generate procoagulant states, where the resolution of Paget-Schroetter syndrome also involves and considers the management of these states, to avoid surgical risks and use the advantages of oral anticoagulation.

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Conflicts of interest

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Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patient mentioned in the article. The corresponding author is in possession of this document.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript nor for the creation of images, graphics, tables, or their corresponding captions.

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