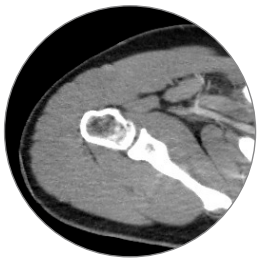

Paget-Schröetter Syndrome. A Case Report

Síndrome de Paget-Schrötter. Presentación de un caso

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Palabras clave (DeCS)
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Trombosis
Adulto joven

Summary

Paget-Schroetter syndrome is defined as a primary deep vein thrombosis of the upper extremities due to obstructive anatomical anomalies in the axillary or subclavian vein that changes venous blood flow or due to intense muscular effort. It usually occurs in young adults with no previous history and its clinical manifestations are pain, spontaneous and severe edema of the upper extremities after stressful physical activity associated with paresthesias, weakness and cyanosis. Diagnosis requires a combination of clinical probability, D-dimer results and ultrasonography. It occurs in approximately 1-2 people per 100,000 inhabitants per year and affects both sexes. The following case is a patient with Paget-Schroetter syndrome which came to the ER at a fourth-level hospital.

Resumen

El síndrome Paget-Schrötter se define como la trombosis venosa profunda primaria de las extremidades superiores, atribuible a anomalías anatómicas obstructivas en la vena axilar o subclavia que alteran el retorno venoso, o por un esfuerzo muscular intenso. Por lo general, se presenta en adultos jóvenes sin antecedentes y se expresa clínicamente como dolor, edema espontáneo y grave de las extremidades superiores luego de una actividad física vigorosa, asociado a parestesias, debilidad y cianosis. Para el diagnóstico se necesita la combinación de la probabilidad clínica, el resultado del dímero D y la ultrasonografía. Su incidencia es aproximadamente de 1 a 2 personas por 100.000 habitantes al año y afecta a ambos sexos. A continuación se presenta el caso de un paciente con síndrome de Paget-Schrötter que consultó al servicio de urgencias en un hospital de tercer nivel.

Introducción

Paget-Schrötter syndrome is defined as primary deep vein thrombosis of the upper extremities secondary to obstructive anatomical abnormalities in the axillary or subclavian vein that alter venous return, or by intense muscular effort. It usually occurs in young adults with no previous history and is clinically expressed as pain, spontaneous and severe edema of the upper extremities after strenuous physical activity, associated with paresthesias, weakness and cyanosis (1).

It is a rare entity, corresponding to 5% of all deep vein thrombosis and occurs in approximately 1-2 persons per 100,000 inhabitants per year, in equal proportion in both sexes. There is a prevalence associated with factor V Leiden, antiphospholipid antibodies and prothrombin gene mutations (1, 2).

The diagnosis requires a combination of clinical probability, D-dimer result and ultrasonography.

Case presentation

A 22-year-old male patient with clinical picture of one day of evolution, characterized by pain in the right upper limb associated with edema and paresthesias. The only relevant history is that he is an individual who practices weightlifting.

Physical examination revealed distal edema extending to the proximal third of the right arm, with slight violaceous coloration, adequate capillary filling, with brachial and radial pulses, sensitivity and arcs of mobility preserved.

With the suspicion of possible venous thrombosis of unclear cause, arterial and venous Doppler of the right upper limb was performed, the examination of the arterial vessels was found within normal limits (Figure 1). Venous Doppler demonstrated acute venous thrombosis of the right subclavian and axillary veins (Figure 2). After ruling out thrombophilia and procoagulant phenomenon associated with SARS-CoV-2 infection, angio-scanography of thoracic vessels was performed, which confirmed acute thrombosis of the right subclavian vein. Due to the history of weightlifting, Paget-Schrötter syndrome was suggested in the differential diagnosis (Figure 3). After this diagnosis, the treating department started full anticoagulation therapy with low molecular weight heparin (enoxaparin) at a dose

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of 1 mg/kg every 12 hours. Subsequently, endovascular treatment was performed by puncture under ultrasound guidance in the brachial vein, with subsequent identification of the site of obstruction by the thrombus, since the thrombus was already obstructing the brachial vein. site of obstruction by the aforementioned thrombus. Once identified, the thromboaspiration system was advanced and the intravenous injection of 5,000 IU of unfractionated heparin was performed, the proximal third of the subclavian vein and the innominate vein were advanced, and angioplasty was performed with an 8 mm balloon in the brachial and axillary veins, and a 10 mm balloon in the brachial, axillary and subclavian veins. Control phlebography confirmed adequate patency of the right brachial and axillary veins, but showed residual thrombus of 20% in the subclavian segment (Figures 4a and 4b).

During hospitalization, the patient had an adequate clinical evolution, without postoperative complications, and was discharged with full anticoagulation.

Discussion

Deep venous thrombosis of the subclavian vein is divided into primary and secondary. The primary one is also known as Paget-Schrötter syndrome or "stress thrombosis" and the secondary one, attributable to insertion of intravascular medical devices in most cases, trauma, thrombophilia or neoplastic disease (1).

The subclavian vein is part of the venous vascular structure that drains the upper limb, its course begins at the junction of the axillary and cephalic veins adjacent to the lateral border of the first costal arch; it runs towards the thoracic cavity with the following anatomical relationships: anterior, with the subclavian muscle and costoclavicular ligament; posterior, with the anterior scalene muscle; superior, with the clavicle; and inferior, with the first costal arch (Figure 5), just before joining with the internal jugular vein at the jugulo-subclavian confluent. By having two soft tissue structures between their anatomical relationships (anterior and posterior limits), hypertrophy of the costoclavicular ligament or the anterior scalene muscle can generate compression on the subclavian vein, which decreases its caliber, causes slow flow and, consequently, facilitates thrombus formation (1-3).

Paget-Schrötter syndrome was first described in 1875 by James Paget, followed in 1884 by von Schrötter (2). Among the risk factors are anatomical alterations and excessive muscular exercise leading to hypertrophy of the scalene muscles, mainly the anterior scalene muscle, which is in contact with the posterior border of the subclavian vein in its trajectory before entering the thoracic cavity (Figure 5) (2). The incidence described in the literature is low, 2:100,000 cases per year, with a peak of presentation around 30 years of age and more common in men with a rate of 2:1 with respect to women (1).

The symptomatology of this entity involves edema, changes in skin coloration, collateral circulation in the axillary region and the shoulder of the affected limb (Urschel's sign), which usually appears in the dominant limb. Eighty percent of patients report the onset of symptoms in the first 24 hours after strenuous physical activity (2, 3).

Within the diagnostic imaging modalities, Doppler ultrasound has a supremely valuable value under the color and spectral Doppler modality, with documented sensitivity ranging from 71-100% and specificity of 80-100% (3, 4), when exploring the blood vessels that compose the deep and superficial venous systems of the affected extremity. Findings such as the absence of flow on color Doppler examination, decrease or loss of phasicity of venous spectra, together with an echogenic image inside the vessel, confirm the diagnosis.

When the Doppler is inconclusive due to the posterior acoustic shadow artifact that could be generated by the bony structures of the thoracic operculum or the presence of gas in cases of trauma, angioscanography of vessels in venous phase has a leading role in confirming or ruling out thrombosis, by visualizing the hypodense opacification defect in the vein lumen (2,5). Findings such as collateral circulation or alteration in the density of the subcutaneous cellular tissue of the extremity should be warning signs of suspected thrombosis, whether in the extremities or of central origin (1, 2). Likewise, venous vessel angioresonance has favorable utility because it does not expose the patient to ionizing radiation and also has good anatomical and contrast resolution. Time-of-flight MRI and acquisitions after gadolinium administration have been reported to have a sensitivity and specificity of 71% and 89% for the former and 50% and 80% for the latter, respectively (1).

Phlebography of vessels of the extremity together with cavography can be diagnostic and therapeutic, grouping the visualization of the intraluminal filling defect of the vessel, presence or not of collateral circulation and also as surgical planning immediately before percutaneous treatment (1, 2).

The multidisciplinary treatment of this disease consists of systemic anticoagulation, endovascular management and surgical decompression of the subclavian vein through the thoracic operculum, by means of resection of the first rib together with the anterior scalene and subclavian muscles, and this is the definitive management (6).

Initially, systemic anticoagulation with low molecular weight heparins, oral anticoagulants or vitamin K antagonists should be performed, which according to current evidence, any of the alternatives is equally effective and safe (1-3, 7).

Endovascular treatment aims to provide rapid relief of symptoms, prevent pulmonary embolism, the development of post-thrombotic syndrome, and restore early function of the affected limb.

This management involves pharmacological catheter thrombectomy, in which initial remodeling and removal of the thrombus is performed followed by infusion of thrombolytic agents in order to recanalize the vein lumen. Current evidence shows that this approach is more successful than thrombus aspiration alone, since it achieves venous return in a single session, which reduces hospital stay times and bleeding rates (3, 6).

Conclusion

Paget-Schrötter syndrome is an entity that although it may be infrequent, it is necessary to suspect both primary and secondary causes. Physical examination with edema, changes in skin coloration and collateral circulation of the extremity raise suspicion in addition to the results of paraclinical examinations and diagnostic imaging. Treatment includes anticoagulant therapy, endovascular treatment and even surgical treatment, so multidisciplinary management should be performed. The role of the radiologist is fundamental when this entity is suspected, since it favors the diagnosis and individualized treatment of the patient.

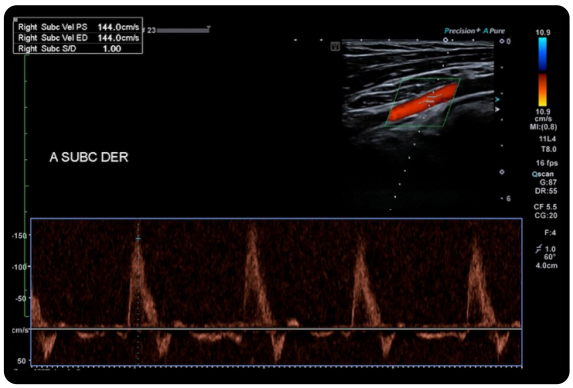

Figure 1. Doppler ultrasound of arterial vessels of the right upper limb. Patent subclavian artery, with three-phase, anterograde, high resistance spectra, within normal limits.

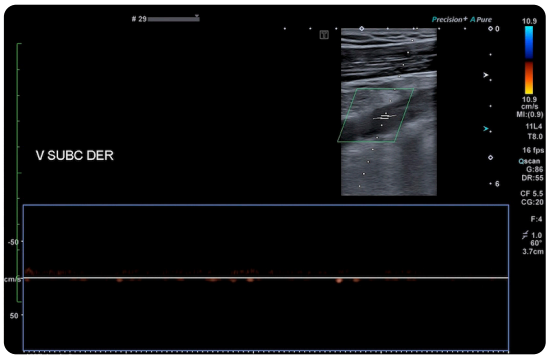

Figure 2. Doppler ultrasound of venous vessels of the right upper limb. Dilatation and occupation by echogenic material within the subclavian vein, extending to the right axillary vein with absence of flow after color and spectral Doppler scanning.

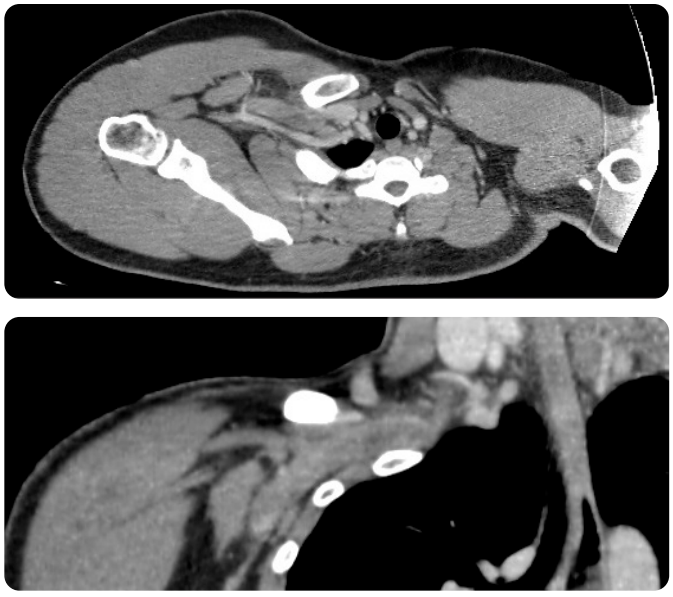

Figure 3. Angioscanography of venous vessels of the right upper limb. A helical volumetric acquisition was performed after intravenous administration of non-ionic water-soluble iodinated contrast medium, with the patient in supine decubitus and arms positioned on both sides of the patient. a) Soft tissue window, post-procedural multiplanar reconstruction in the axis of the subclavian vein and b) coronal reconstruction. Dilatation and absence of opacification of the subclavian vein along its entire course, as well as the axillary vein after administration of intravenous contrast medium. There is adequate patency of the distal veins.

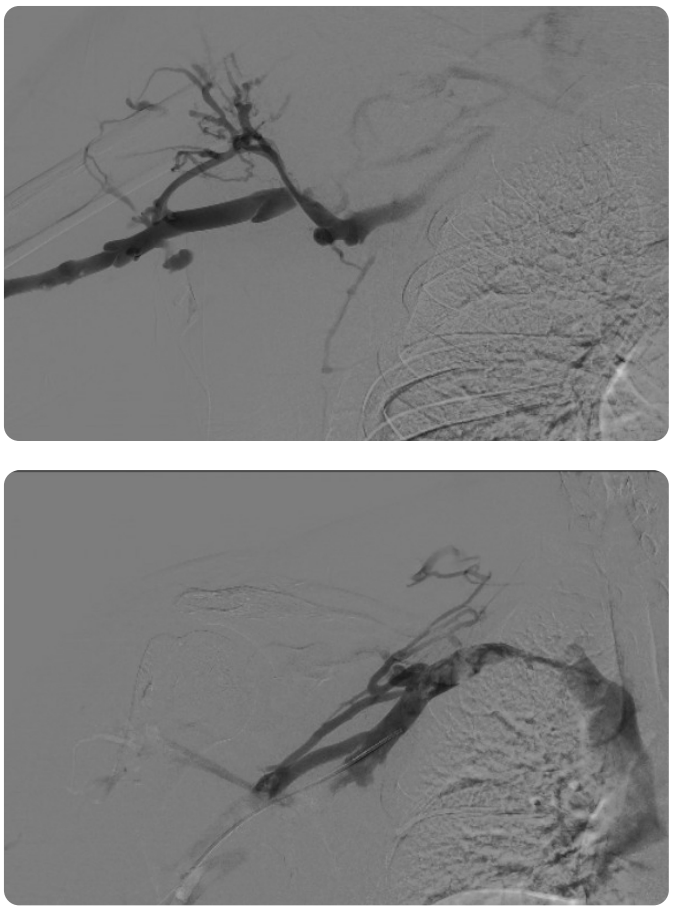

Figure 4. Phlebography of the right upper limb. a) Absence of axillary and subclavian vein opacification. b) The patient underwent thrombolysis and mechanical thrombectomy to achieve patency of the brachial and axillary segment with residual thrombus of 20%.

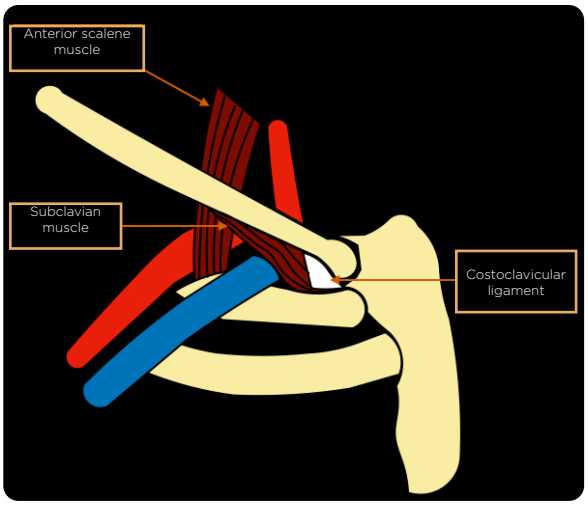

Figure 5. Anatomy. Schematic of the anatomical relationships of the subclavian vein.
Source: Own elaboration.

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