

## A 19-year-old man with Paget-Schroetter syndrome

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### Abstract

The Paget-Schroetter syndrome or effort thrombosis of subclavian vein is an uncommon condition that should be better managed by catheter-directed thrombolysis. In developed countries, venography studies by computed tomographic and magnetic resonance have been the most adequate tools for characterization of this diagnosis.

Here we present the case of a 19-year-old man who had pain in the right hemithorax, following a physical effort. The pain was radiating to the right upper limb, associated with cyanosis, and venous dilatation. The arterial pulses and the rest of physical examination were normal. Paget-Schroetter syndrome was diagnosed by images of Doppler echocardiography, which showed occlusion of subclavian vein. The management was conservative, initially with enoxaparin 60 mg twice/day, and further with warfarin 5 mg/day for six months. He was asymptomatic also upon discharge. In developing countries, Doppler ultrasonography can be useful for diagnosis of thrombosis of subclavian vein, and anticoagulation alone may be a good option for its treatment.

**Keywords:** Paget Schroetter syndrome, subclavian vein, vein effort thrombosis.

## Introduction

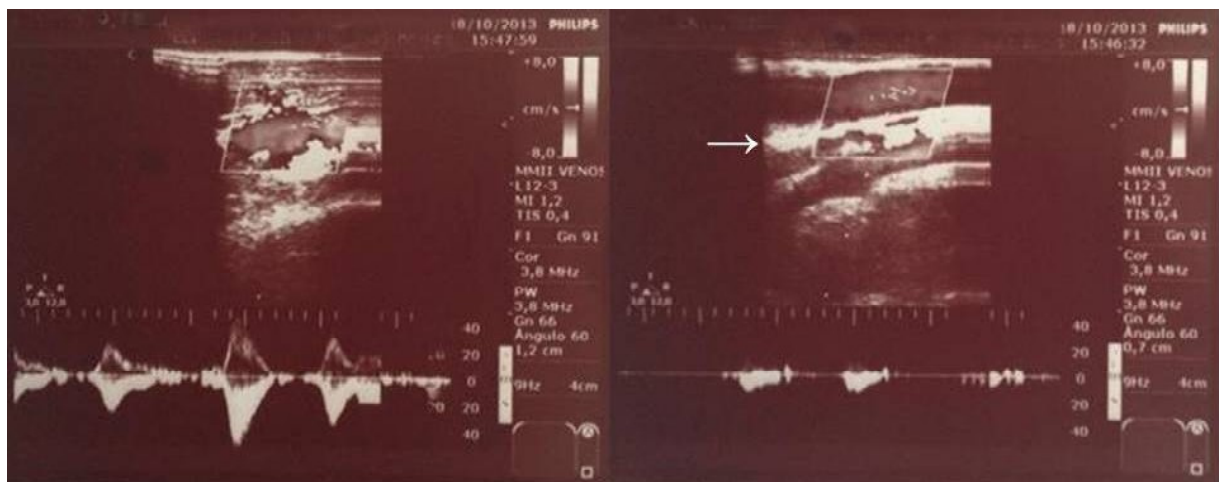
The Paget-Schroetter syndrome is a rare condition which involves thrombosis of subclavian veins associated with muscular efforts of the upper limb, which cause endothelial injuries (1-6). Venography studies by computed tomography and magnetic resonance are growing in use, but images of Doppler ultrasonography (US) are also useful for diagnosis in developing areas (1,2). Catheter-directed thrombolysis is considered essential for optimizing outcomes (1,2,4,6). Anticoagulation alone can also achieve clinical improvement in Paget-Schroetter syndrome (1,3). The aim of this paper is to report a case of this syndrome diagnosed by US and controlled by anticoagulation.

## Case Report

In October 2013, a 19-year-old right-handed man felt severe chest pain in the right hemithorax, soon after performing a moderate physical effort. The pain was throbbing and respiratory-dependent, radiating from the shoulder to the hand, and was

associated with paresis, erythrocyanosis, and regional venous dilatation. The arterial pulses and the rest of physical examination were unremarkable. The pain was partially alleviated by paracetamol; however, he evolved with edema and cyanosis. On admission, the main hypothesis was Paget-Schroetter syndrome, which was confirmed by images of US, showing a conspicuous venous occlusion due to subacute deep venous thrombosis (DVT) affecting the right subclavia (Figure 1). The axillary, radial and ulnar veins appeared permeable, as well the cephalic and basilica veins. On admission, the results of the routine laboratory determinations were considered unremarkable. With diagnosis of Paget-Schroetter syndrome, the patient underwent a conservative management, beginning with subcutaneous enoxaparin 60 mg (1 mg/kg) twice daily, further changed for oral warfarin (5 mg/day), which was utilized during six months. The patient was asymptomatic on the sixth day of admission, and with INR of control 2.5, he was discharged to outpatient long-term follow-up.

**Figure 1. Echo-Doppler study: Image of the axillary vein appearing permeable, thin-walled and with a normal diameter; the subclavian vein is showed practically without blood flow, and presenting an image of occlusive thrombus (arrow)**



## Discussion

The subclavian vein is the extension of the axillary vein after the lateral edge of the first rib, and follows the lower clavicular surface to join the

internal jugular and form the brachiocephalic vein. This vein is vulnerable to injury at the junction of the first rib and clavicle in thoracic outlet (1,2).

Occlusions of subclavian vein are most often iatrogenic but may be related to malignancies (1). The previously healthy young student herein reported denied repetitive efforts or antecedent trauma; and skeletal abnormalities at the thoracic outlet were not detected by imaging studies (1). Moreover, predisposing factors as iatrogenic factors, thrombophilia, and malignancy were ruled out (1). He did not exert gymnastics, martial arts, ball sports (volley, basket, golf), or swimming (1). Therefore, spontaneous thrombosis might have occurred as described by Cruveilhier in 1816 (7). Estimated incidence of the syndrome is about 1-2 patients per 100,000 people per year, and is more frequent in the right limb of males (2:1), with mean age at presentation in the early 30s (2). Clinical features are often non-specific like pain in the arm, shoulder or neck, paresthesias, and skin changes (3,5); only few patients present with vascular and/or muscular indicative data of DVT (5). Because of the usual diagnostic challenges related to Paget-Schroetter syndrome, late diagnoses frequently occur and prevent the prompt beginning of treatment, with worsening of outcomes (5). Elevated clinical suspicion might favor the appropriate imaging studies and the timely referrals (5). Chiropractic therapy, anticoagulation, catheter-directed thrombolysis, and pharmacomechanical thrombectomy are useful tools for management of individuals with Paget-Schroetter syndrome (1-6). Although yet controversial, invasive procedures are considered gold standard for treatment, including rib or costoclavicular ligament excision, scalenectomy, and radical external venolysis (2). Kidd and Brodrick reported a

previously healthy 32-year-old right-handed woman with bilateral obstruction of the subclavian veins confirmed by imaging studies. Interestingly, clinical findings of the Paget-Schroetter syndrome were exclusively observed on her left upper limb, and anticoagulation was the initial therapy, while the patient decided about bilateral rib resection (3). Stainby et al. reported two patients with the syndrome and emphasized the role played by doctors of chiropractic in the co-management of this condition. One was a 44-year-old man with recent onset of pain in the neck like pinched nerve sensation in his left trapezius, which had initial care of chiropractic clinic, but thrombolytic agent and anticoagulation were further utilized (3). The other, a 36-year-old woman with chronic right neck, shoulder and arm pain, and US images detected clot occlusions in the subclavian cephalic and axillary veins. She underwent anticoagulation plus chiropractic soft tissue and spinal manipulative therapy (3). It seems necessary to increase the awareness about this relatively rare entity (1,5), and perform prospective trials to clarify its risk factors, etiology and non-entirely consensual management (3).

## Conclusion

Although considered uncommon, this potentially fatal clinical condition, which can involve significant morbidity rates, actually may be underestimated, under diagnosed and under reported. In developing countries, US can be a useful option to diagnose the subclavian vein thrombosis, and anticoagulation alone may be a reliable option for the case management.

**Conflicts of interest:** None declared.

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