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# Paget-Schroetter Syndrome: Case Report and Review of the Literature

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## Abstract

Paget-Schroetter syndrome is the spontaneous thrombosis of the axillary/subclavian vein. A 26 years old man presented with acute onset of right upper limb swelling and aching pain after a vigorous game of tennis the day before admission. His right upper limb was swollen and cyanosed. Adson's sign was positive and a venogram confirmed the diagnosis of Paget-Schroetter syndrome. He was started on intravenous thrombolytics followed by oral anticoagulation therapy. His symptoms resolved and he was symptom free at 1 year follow-up. Thrombolytics and anti-coagulation is the most widely accepted first line therapy for this syndrome. Defining any anatomical anomaly as the predisposing factor in this condition is essential in the selection of patient who will benefit from thoracic outlet decompression. Surgery should be selectively offered to patients with definite anatomical evidence of a thoracic outlet obstruction.

## INTRODUCTION

Paget-Schroetter syndrome is the spontaneous, primary thrombosis of the axillary/ subclavian vein. First described independently by Von-Schrotter<sup>1</sup> in 1884 in Vienna and by Paget<sup>2</sup> in 1875 in London, this syndrome is also known as 'effort' thrombosis of the axillary/ subclavian vein because of the frequent association with exertion superimposed on anatomical compressive elements in the thoracic outlet<sup>3, 4</sup>. Optimal management of this condition is still debated. Here we present a case of this condition and examine the differing current opinions on the management of this entity.

## CASE REPORT

A 26 year old, right hand dominant man who was previously well was admitted to our institution complaining of an acute onset of right upper limb swelling, and bluish discoloration over the past one day prior to admission. This was associated with an aching pain over the right axilla. His right arm swelling had started 5 hours after a tennis game. While serving a ball he experienced a sharp pain in his right axilla associated with a snapping sound. On examination, prominent dilated superficial veins, which persisted despite limb elevation, were noted. Pulses were normal and no masses were felt in the axilla or neck. Adson's and hyperabduction signs were positive.

A clinical diagnosis of Paget-Schroetter syndrome was made

and an urgent venogram was done. This confirmed the thrombosis of the axillary-subclavian vein (Fig 1).

Thrombolysis and anticoagulation were initiated. A cervical spine X-ray was done and no cervical rib or any anatomical anomaly was demonstrated. His swelling and symptoms improved over the next few days and a check duplex scan of the subclavian-axillary vein showed that the vein was patent. His coagulation profile was all normal. He refused a magnetic resonance imaging (MRI) scan of his thoracic outlet. He recovered completely and went back to his pre-injury level of activity one year later with no symptoms suggestive of residual chronic thoracic outlet compression.

**Figure 1**

Figure 1: Angiogram done on admission showing a thrombus in the axillary/ subclavian vein of the right upper limb (Arrow).



**DISCUSSION**

The Paget-Schroetter syndrome is one of a collection of syndromes associated with thoracic outlet compression. The subclavian vein rests in the groove anterior to the scalenus anterior with the subclavian artery and brachial plexus lying posterior to it. The clavicle runs in close apposition directly over the neurovascular structures. As the clavicle swings backwards, the vein, artery and nerves become compressed between the clavicle and the first rib, thereby giving rise to a collection of symptoms known as the thoracic outlet compression syndrome.

There is usually a temporal and causal relationship between activities involving hyperabduction of the upper limbs and the genesis of the thrombosis <sup>5, 6</sup>. Hyperabduction and retroversion of the arm, especially with sudden jerky movements can strain the subclavian and axillary artery and vein causing it to be crushed between the clavicle and the first rib <sup>7</sup>. This can cause venous intima damage and initiate venous thrombosis. Mechanical compression of the vein by adjoining bone (cervical rib), muscles or fibrous muscular bands can predispose the veins to mechanical compression and trauma. Roos <sup>8</sup> described nine common anomalies involving fibromuscular bands that are functionally similar to a cervical rib. These muscular and fascial bands when present predispose the neurovascular structures to compression.

A detailed history, thorough appropriate physical examinations are often sufficient to suggest the diagnosis <sup>5</sup>. The onset of symptoms is usually acute, with an antecedent

episode of repetitive or sustained ipsilateral abduction and external rotation. A history of trauma, indwelling catheter and thrombophilic state should also be excluded to distinguish this syndrome from secondary axillary/ subclavian vein thrombosis. The diagnosis is confirmed with a venogram or a duplex ultrasound scan. The differential diagnosis includes lymphangitis, lymphatic obstruction, intramuscular hemorrhage, intermittent venous obstruction without thrombosis and venacaval or innominate vein obstruction.

The optimal management of this entity is the subject of much debate and controversy. There is currently a near universal acceptance of thrombolysis and anticoagulation as the initial forms of therapy <sup>5, 6, 9, 10, 11, 12</sup>. However, the indications for and the timing of surgery are not well defined. Some authors advocate immediate thrombolytic therapy followed by prompt thoracic outlet decompression <sup>5, 11, 12</sup> while others adopt a more selective approach, offering surgery only to patients with persistent symptoms following initial thrombolytic and anticoagulation therapy and a period of close observation <sup>6, 9, 10</sup>.

There are two groups of patients with Paget-Schroetter syndrome. One group of patients has osseous or musculotendinous abnormalities of the thoracic outlet that predisposed them to the development of this syndrome. In their series, Kunkel et al <sup>9</sup> documented osseous or musculotendinous abnormalities in 18 of their 25 patients (72%) presenting with this syndrome. They suggested that repeated compression of the subclavian-axillary vein might cause sequentially fibrosis, stenosis and subsequently thrombosis of the vein. This subgroup of patients may benefit from thoracic outlet decompression. Resection of the first rib or the anterior scalene muscle with excision of any osseous or musculotendinous abnormalities is the commonly performed operation. The second group of patients has no thoracic outlet anatomical abnormalities. The etiology in this group is usually traumatic in nature. A sudden hyperabduction and retroversion of the upper limb (such as during a serve in a tennis game) can crush the subclavian-axillary vein between the clavicle and the first rib. The resulting intimal damage or tear can precipitate the thrombosis of the vein. Conceivably, the risk of recurrence will be less and thoracic outlet decompression may be unnecessary.

Not all patients with Paget-Schroetter syndrome require surgical intervention <sup>6, 9, 10, 12</sup>. Surgery should be reserved

from patients with persistent symptoms of thoracic outlet compression and venous obstruction after an initial period of thrombolysis and close observation. These are usually patients with some degree of extrinsic compression that would benefit from decompressive surgery. The use of adjuncts such as a cervical spine X-ray, computed tomography (CT) scan and magnetic resonance imaging (MRI) of the thoracic outlet may have a valuable role. The demonstration of osseous and musculotendinous abnormalities may be used for selection of patients for early thoracic outlet decompression as advocated by some authors.

Stenting of the axillary/ subclavian vein as a modality of treatment for this condition is contraindicated<sup>5</sup>. Many patients that develop this syndrome have an underlying narrowing of the thoracic outlet (osseous or fibromuscular). A stent inserted under these conditions may not deploy properly because of the external compression and may induce further thrombosis and therefore further compromising the already tenuous venous outflow.

## **CONCLUSION**

In summary, thrombolytics and anti-coagulation is the most widely accepted first line therapy for this syndrome. Surgery however should be selectively offered to patients with definite anatomical evidence of a thoracic outlet obstruction.

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