
CASE REPORT**Paget-Schroetter Syndrome in 52-Year-Old Male: An Interesting Case Report**

Nik Kosai^{1*}, Anitha Haniffa¹, Razman Jarmin¹, Srijit Das², Reynu Rajan¹, Hanafiah Harunarashid¹

¹Department of Surgery, ²Department of Anatomy, Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latif Bandar Tun Razak, 56000, Cheras, Kuala Lumpur, Malaysia

Abstract:

Paget-Schroetter syndrome is a form of upper limb deep venous thrombosis and it is commonly seen in young individuals. Paget-Schroetter syndrome is a rare condition and diagnosis becomes more difficult if it occurs in old individuals. There is no clear consensus regarding the exact treatment of Paget's Schroetter syndrome. A high grade of suspicion with early diagnosis and treatment is needed. We here describe the case of Paget-Schroetter syndrome in a 52-year-old male and discuss the important clinical features and treatment modalities.

Keywords: Paget-Schroetter syndrome, Deep Venous Thrombosis

Introduction:

Paget-Schroetter syndrome, also known as effort thrombosis, is a rare condition consisting of axillosubclavian vein thrombosis and Venous Thoracic Outlet Syndrome (VTOS) and it accounts between 1 to 4% of all deep vein thrombosis [1]. Predominantly, it affects the young individuals with the mean age at presentation being the early 30s [1]. It was first described by Paget and von Schroetter in 1875 and 1884, respectively. In true effort thrombosis, the symptoms are persistent and patients classically present with sudden onset pain, bluish discolouration, heaviness and swelling. As the condition is rare, a high grade of suspicion is required to ensure early diagnosis and treatment. We describe Paget-Schroetter syndrome in a 52-year-old male and highlight the important clinical features and treatment modalities.

Case Report:

Patient anonymity was maintained and consent was taken for the case. A 52-year-old man, with no known previous medical illness presented with a 2-week history of sudden onset right arm swelling and pain with no associated discolouration of the skin or neurological deficit. He also complained of heaviness and some discomfort. The patient was a golf enthusiast, and had recently returned after a week-long golf trip with no history of trauma. There was no similar complaint in the past but patient had a history of right elbow fracture and carpal tunnel syndrome.

Upon further examination, the entire upper limb was swollen with prominent superficial veins in the arm. The distal radial pulse was felt with good distal perfusion. The right upper limb was otherwise soft and non tender with full range of movement over all joints. Duplex ultrasonographic examination revealed thrombosis of the right basilic vein up to the axillary vein. He was started on subcutaneous fondaparinux sodium 7.5mg daily for the treatment of deep vein thrombosis. We then proceeded with a Contrast Enhanced Computed Topography (CECT) scan for the right upper limb and thorax which showed no opacification of the right brachial and basilic veins from the mid arm level extending proximally till the right axillary and subclavian veins, the right internal jugular vein was well opacified with no thrombus within and with no filling defect in the right atrium. There was no evidence of any gross structural abnormality that may have contributed to the thoracic outlet

syndrome. The central and peripheral venogram confirmed the right basilic, axillary and subclavian thrombosis (Fig.1) for which we then proceeded with mechanical thrombolysis. Aspirex S 10 was used to fragment the thrombus and to aspirate it from the vessel. Angiographic images post procedure (Fig.2) showed good flow of contrast into the right axillary, subclavian and brachiocephalic veins. The patient was started on warfarin sodium and the limb was kept elevated. Improvement of the symptoms and significant reduction in the size of the arm was witnessed within 2 days. Warfarin sodium was continued to maintain the Internationalized Normal Ratio (INR) within the therapeutic range of 2 to 3. During follow-up, his arm had returned to normal and he has remained asymptomatic.

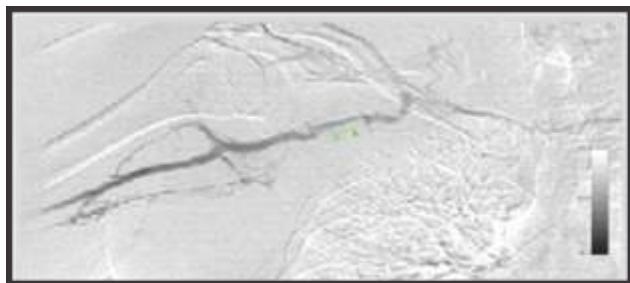


Fig. 1: The Venographic Image done Before the Mechanical Thrombectomy, the Arrows Shows the Filling Defect Indicative of Thrombosis.

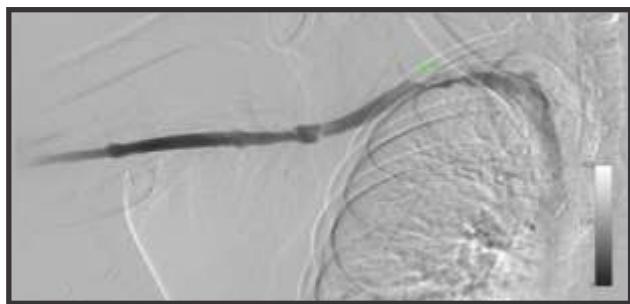


Fig. 2: Image Post of Thrombectomy, the Subclavian and Brachiocephalic Veins Are Opacified

Discussion:

The subclavian vein is in the anterior part of the thoracic outlet, and it passes by the intersection of the first rib and clavicle, making it particularly vulnerable to any injury. The subclavius muscle lying inferior to the clavicle adds bulk and further compresses the subclavian vein. Repetitive arm activity leads to intrinsic damage and extrinsic scar tissue formation, the entrapped vein is less mobile and fixed to the surrounding structures, thus increasing the risk for injury. This phenomenon is witnessed in 40% of individuals, especially with hyperabduction and external rotation [2]. These components combined with the compressive elements of VTOS leads to thrombosis, hence it is also known as “effort” thrombosis.

There is a spectrum of patients that present with venous thoracic outlet syndrome. The most common being catheter and/or dialysis associated secondary thrombosis, effort thrombosis and, the least common being intermittent positional obstruction without thrombosis. The later entity can progress to effort thrombosis if not corrected and usually present with episodic arm discolouration and swelling which is precipitated by either arm abduction or exercise. In all cases, especially if the occlusion is chronic, there will be prominent venous pattern of dilated superficial veins in the anterior chest wall, upper arm and base of the neck. Subclavian vein can be compressed even in normal individuals within the costoclavicular space as a result of abduction of the arm, despite the absence of any anatomical abnormalities [3]. In these patients, the venograms are normal at rest and when arm is abducted, there is evidence of compression and collaterals. There is another subclinical entity where there is recurrent partial thrombosis followed by recanalization of the vein, in which case the patient may or may not be symptomatic. In this instance even if the vein is

not thrombosed, there is evidence of intrinsic defect and strictures within the vein [4]. In all cases there are multiple collateral pathways present which is a diagnostic venographic feature [3]. These collaterals particularly connect the cephalic vein and the profunda branches with the transverse cervical, scapular, and external and internal jugular veins [5]. Historically, effort thrombosis was treated with anticoagulation alone and the results have been disappointing [6]. Catheter directed thrombolysis has shown nearly 100% successes in fresh clots if treatment was initiated within a few days of onset of symptoms, otherwise it has been successful in 62 to 84% of the cases [7]. The rate of re-thrombosis is high as the issue of extrinsic compression has not been addressed [8]. Thus, it has been recommended for the decompression of the anterior thoracic outlet by selective first rib resection to avoid re-thrombosis but the effectiveness of this procedure are yet to be proven

[9]. There is no clear consensus regarding the exact treatment of Paget's Schroetter syndrome. According to reports, it is either combination of anticoagulants and fibrinolitics or it may be even the surgical procedures [10]. As there are no clear guidelines on the management of Paget's Schroetter syndrome and only level 3 evidences is available.

Conclusion:

The reported patient was on anticoagulation therapy for a 6-month duration based on Deep Vein Thrombosis (DVT) clinical practice guidelines. The reported patient had upper limb DVT secondary to effort thrombosis i.e. Paget's Schroetter syndrome. Rate of re-thrombosis is up to 25%, hence the patient requires regular follow-up to rule out recurrent thrombosis so that early treatment can be advocated. The diagnosis of Paget's Schroetter syndrome may be difficult but DVT findings may aid in diagnosis.

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*Author for Correspondence: Dr. Nik Ritza Kosai, Associate Professor, Department of Surgery
Universiti Kebangsaan Malaysia Medical Centre Jalan Yaacob Latif Bandar Tun Razak
56000, Cheras, Kuala Lumpur, Malaysia Email: nikkosai@yahoo.co.uk Tel: +6-03-91455555