



PAGET-SCHROETTER SYNDROME A.K.A. EFFORT THROMBOSIS (UPPER EXTREMITY DEEP VEIN THROMBOSIS)

General Medicine

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ABSTRACT

Deep vein thrombosis according to etiology classified into primary or secondary and according to location upper limb DVT and lower limb DVT. Incidence of secondary DVT are high and seen mostly in hospitalized patient etiology being indwelling catheter, post surgery, immobilization in sick patients due to neurological disorder, orthopaedic interventions. OCPs, pregnancy, cancer, smoking, HRT, genetic disorders are the other causes for secondary DVT. The most common sites for these venous thromboses are leg and pelvic veins. Primary subclavian vein thrombosis also called effort thrombosis is relatively an uncommon condition with an incidence of 1 in 100000 per year. It was first described by Paget in 1875, and Schroetter in 1884 independently. Hughes, while reviewing his 320 cases of subclavian vein thrombosis, gave its name as Paget-Schroetter Syndrome. We report below a case of chronic primary upper limb DVT (Paget-Schroetter syndrome).

KEYWORDS

INTRODUCTION

Incidence of provoked DVT are high and seen mostly in hospitalized patient etiology being indwelling catheter, post surgery, immobilization in sick patients due to neurological disorder, orthopaedic interventions, surgery. OCPs, pregnancy, cancer, smoking, HRT, genetic disorders are the risk factors for DVT. The most common sites for these venous thromboses are leg and pelvic veins. Primary thrombosis of the subclavian vein at the costoclavicular junctions is referred as Paget-Schroetter syndrome, relatively an uncommon condition with an incidence of 1 in 100000¹ per year, associated with strenuous and repetitive activity of the upper extremities. The earliest description of spontaneous Axillary Subclavian VT was by Cruveilhier in 1816, and the first elaborate account was provided by James Paget in 1875.^{2,3} In 1894, von Schroetter was the first to identify vascular trauma from muscle strain as a potential etiologic factor.⁴ In 1948, Hughes coined the term Paget-Schroetter Syndrome (PSS) and published the first review.^{5,6}

CASE REPORT

A thirty seven year old male patient was admitted with chief complaint of prominent veins on the right side of the chest and right arm since a period of 6 months. It started with the patient having to carry a heavy weight on his right shoulder after which he developed painful swelling of the right arm. After a few days, swelling and pain has resolved but patient noticed dilated, prominent veins on the chest and right arm. There was no wound or obvious injury of the right hand. There was no cough, chest pain or breathlessness. The patient was non-diabetic, non hypertensive, non smoker.

On examination: There were dilated engorged veins on the right arm, anterior chest wall mainly on the right side and extending to the neck [Fig. 1], the direction of flow being below upwards towards the neck his right radial pulse rate was 75/minute, regular, good volume; all peripheral pulsations were well felt. B.P. in the both upper limb was 120/80 mm Hg. There was no lymphadenopathy or Horner's syndrome. The examination of respiratory, cardiovascular and abdominal systems was within normal limits. Routine investigations, coagulation profile and X-ray of the chest were within normal limits. A Doppler ultrasound of the right upper limb showed a thrombosis of the right subclavian vein {Fig.2,3}. As the disease was six month old, and there was no active extension of thrombus or phlebitis as evident by absence of signs of inflammation and normal ESR, thrombolytic agents were not tried. The patient was managed conservatively with sub-cutaneous low molecular weight heparin followed by oral Warfarin. Patient's INR was repeated every other day initially for 10days once INR was in therapeutic range for 2 consecutive days INR was repeated every 5th day for three weeks. After 30 days of treatment venous Doppler was reviewed, it was showing decreased thrombus size with recanalization of obstructed vein and same treatment was continued. Patient is being followed up every 3 months Doppler ultrasonography was performed at every visit. The patient's right arm and chest venous dilation improved gradually there was no evidence of post-thrombotic syndrome.



Figure 1. Right sided prominent veins due to collaterals.

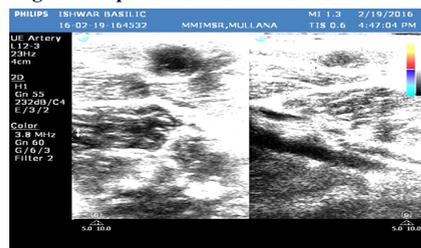


Figure 2. Non compressible subclavian vein.

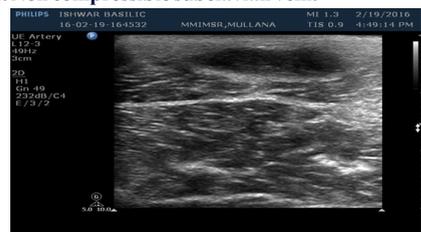


Figure 3. Non compressible subclavian vein.

DISCUSSION

The clinical presentation can easily be confirmed by Doppler ultrasonography, But gold standard for determination of anatomy (location, size and level of obstruction) and for planning active interventions (catheter directed thrombolysis and percutaneous transluminal angioplasty) is contrast venogram (PTA).⁷ In Paget-Schroetter syndrome, earlier the diagnosis and treatment, better the results.⁸ The treatment goals are to relieve the acute symptoms of venous occlusion, prevent pulmonary embolism, reduce the likelihood of recurrent thrombosis, and avoid development of the post-thrombotic syndrome. Thrombolysis and anticoagulation is the mainstay of treatment. It has been suggested that the optimal period for thrombolytic treatment is within 6 weeks of the thrombosis.⁹ Urokinase or recombinant tissue plasminogen activator are recommended thrombolytic agents for DVT.⁷ Anticoagulants are used

to prevent further deposition of thrombus, allowing an established thrombus to stabilise and to undergo endogenous lysis, reducing the risk of recurrent thrombosis.¹² Decompression of the thoracic outlet can be performed later, using various techniques. Transaxillary resection of the first rib is the most popular surgical intervention. After achieving successful thrombolysis, rib resection and neurovascular decompression is safe and effective, suggested by Urschel and Razzuk⁸. If patency is not achieved by rib resection and there is some residual arm disability, surgical axillary-subclavian vein revascularisation can provide good mid-term results.⁸ Various vascular procedures including percutaneous transluminal angioplasty, 'Roto-rooter' techniques, and venous stenting have also been used to treat this syndrome. The 'Roto-rooter' technique is a minimally invasive endovascular procedure used for mechanical removal of the thrombus or the plaque from the arterial or venous walls by using a rotational catheter. Urschel et al^{8,9} reported that thrombolysis or 'Roto-rooter' techniques have not been successful after 3 months. Post-thrombotic syndrome is a common complication of upper limb deep vein thrombosis with a frequency ranging from 7 to 46% (weighted mean, 15%)¹⁰. There is currently no validated, standardised scale for assessing upper limb post-thrombotic syndrome, and little consensus on the optimal management of this condition. In conclusion, Paget-Schroetter syndrome can present as chronic form with pain, swelling and prominent veins of the upper limb, particularly in young and active patients with excessive arm activity.

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