Upper Extremity Thrombosis Associated with Overactivity: A Comprehensive review article on Paget-Schroetter Syndrome

Balaji M S[1], Darshan Sohi[2]

[1]Ph.D Scholar, Himalayan University, Itanagar, Arunachal Pradesh, India,[2]Reseach Supervisaor Himalayan University, Itanagar, Arunachal Pradesh.

Abstract

Paget–Schroetter disease, is a form of deep vein thrombosis (DVT) in upper extremity, in which blood clots occurs in the deep veins of the arms. Paget–Schroetter disease syndrome is accounts for 30–40% of spontaneous axillary-subclavian vein thrombosis (ASVT) and for 10–20% of all upper extremity deep venous thrombosis (UEDVT). Important predisposing factors for UEDVT include indwelling hardware, occult or overt malignancy and other thrombophilic states. Clinical manifestations include heaviness, cyanosis, redness, and dilated, visible veins across the upper arm and shoulder. It is diagnosed by compression ultrasonography with the help of colour Doppler. Thrombosis was managed conservatively with limb elevation and anticoagulation therapy. Hence, there is a need for increasing health workers awareness about risk factors, etiology and the management of this unique and relatively uncommon disorder.

Key words: *Deep Vein Thrombosis, ASVT, UEDVT & Thoracic Outlet Decompression*

Introduction

Paget-Schroetter syndrome (PSS) or "effort" thrombosis of the axillary-subclavian vein is a rare cause of deep vein thrombosis (DVT) seen in physically active (sportsmen's) and otherwise healthy individuals. It was first described in 1875 by Paget and Von Schroetter in 1884 and was termed as "Paget-Schroetter syndrome" by Hughes in 1949.[1] Paget-Schroetter syndrome accounts for 30–40% of spontaneous axillary-subclavian vein thrombosis (ASVT) and for 10–20% of all upper extremity deep venous thrombosis (UEDVT).[2] PSS is diagnosed with the help of Doppler ultrasound, computed tomography, and magnetic resonance venography. Oral anticoagulation alone is insufficient and catheter-directed thrombolysis (CDT) is usually performed during management.[3]

Corresponding Author:
Balaji M S,
Ph.D Scholar, Himalayan University, Itanagar,
Arunachal Pradesh, India
Email Id : bforusurroorr@gmail.com

Definition: Paget–Schroetter disease, is a form of deep vein thrombosis (DVT) in upper extremity, in which blood clots occurs in the deep veins of the arms. These DVTs typically occur in the axillary and/or subclavian veins.[4]

Causes

Crucial predisposing factors for UEDVT include indwelling hardware (central venous catheter, ports, and pacemakers), occult or overt malignancy and other thrombophilic states.[5] Effort thrombosis usually follows repeated strenuous sporting or physical activities, such as wrestling, playing ball, swimming and gymnastics, which involves vigorous and sustained upper extremity movements. It is believed that retroversion, hyperabduction and extension of the arms involved with these activities impose undue strain on the subclavian vein leading to microtrauma of the endothelium and activation of the coagulation cascade. Substantial evidence now supports that the role of anatomical abnormalities involving the thoracic outlet in the pathogenesis of effort thrombosis.[6,7]

Signs & Symptoms:

Unfortunately, effort thrombosis is more common in young adults and otherwise healthy men. It preferentially occurs in dominant arm due to repeated use. Those with UEDVT secondary to central venous catheters, patients with effort thrombosis are usually symptomatic. Swelling and arm discomfort are the most frequent evolving problems. Other symptoms include heaviness, cyanosis, redness of arm, and dilated, visible veins across the shoulder and upper arm. Complications this disease include pulmonary embolism, post-thrombotic syndrome and recurrent thrombosis. Post-thrombotic is more frequent in effort thrombosis, compared to secondary UEDVT, and is the major contributor to the morbidity associated with disease.[7]

Diagnostic Evaluation:

Notably, the clinical features of UEDVT have poor evidence, and less than 50% of those with suggestive symptoms actually have deep venous thrombosis (DVT).[7] Compression ultrasonography with colour Doppler by virtue of its ease, availability, portability and low cost is currently the preferred initial test in the evaluation of suspected UEDVT.[8] Radionuclide, magnetic resonance imaging and computed tomographic venography are advanced than ultrasonography. Magnetic resonance venography has the highest sensitivity and specificity among all the non-invasive diagnostic techniques.[9] Though venography is not necessary for diagnosis, it is almost always done as a part of multimodal treatment strategy to deliver catheter-directed thrombolysis and plan thoracic outlet decompression surgery.[7]

Treatment:

Thrombosis were managed traditionally and conservatively with limb elevation and anticoagulation therapy. However, subsequent long-term data demonstrated an unacceptably high incidence of residual symptoms, disability and recurrent thrombosis with this conservative method.[7] This has prompted clinicians to evaluate aggressive management strategies involving thrombolysis, thrombectomy, percutaneous and surgical venoplasty, venous bypass and stents. Systemic fibrinolysis is superior to anticoagulation in achieving vein patency but is associated with increased rates of complications such as intracranial hemorrhage.[10] Therapy directed at thoracic outlet decompression (TOD) involves resection of the first rib, division of the scalenus muscles and or infraclavicular approach. [11]

Conclusion:

Thrombosis is a complex and uncommon condition with a distinct pathological property. Most health workers unfamiliar with effort thrombosis manage it likely to classic lower extremity DVT. Instead, effort thrombosis is ideally managed using a multimodal approach consisting of routine catheter-directed thrombolysis, early TOD in appropriate patients and physiotherapy and occupational therapy. Increasing awareness among primary care and emergency health care workers will ensure early recognition, timely thrombolysis, and prompt referral to a treatment options like thoracic or vascular surgeon. Future scientific research needs to focus on describing the benefit of thrombolytic therapy in patients presenting delayed, identifying factors that predict ineffectiveness of thrombolysis and need for surgical intervention. Other avenues for research include assessment of the need for and duration of anticoagulation following TOD, and cost benefit analysis of the various treatment modalities.

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