Paget-von Schroetter Syndrome: Upper Extremity Deep Vein Thrombosis after Continuous Lifting of Heavy Weight

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Abstract
Effort-induced axillary and/or subclavian vein thrombosis occurring in otherwise normal individuals is referred to as Paget-von Schroetter Syndrome (PVSS) or primary, effort-induced, upper extremity deep vein thrombosis (UEDVT). A 42-year old right-handed male presented with complaints of swelling over the left shoulder and arm and dull aching pain in the left arm for two days following regular lifting of heavy LPG gas cylinders. Left upper limb venous Doppler study revealed partial thrombus in the distal axillary vein and near total thrombus in the subclavian and basilic vein. He was managed with anticoagulation therapy.

Introduction
Upper extremity deep vein thrombosis (UEDVT), most commonly involving axillary and/or subclavian veins, is an important clinical entity with potential for considerable morbidity. UEDVT is classified as either primary or secondary on the basis of pathogenesis. Primary UEDVT is either effort-induced or is idiopathic. Primary, effort-induced UEDVT is also referred as Paget-von Schroetter syndrome (PVSS). In this case report we describe a case of PVSS caused by repetitive lifting of heavy liquefied petroleum gas (LPG) cylinders.

Case Report
A 42-year old right-handed male presented with complaints of swelling over the left shoulder and arm and dull aching pain in the left arm for two days. He worked in an LPG agency where his job profile required regular lifting of heavy LPG gas cylinders.

There was no history of trauma or injury to his upper extremities. There was no history of chest pain, dyspnoea, cough, fever, bleeding, easy bruising and palpitation. There was no significant history of any past ailment. The patient’s family history was negative for coagulopathies, venous thrombosis, pulmonary embolism or any missed abortions. The patient was a non-smoker and only a social drinker. On examination, he had swelling of left upper limb and shoulder. His blood pressure was mildly elevated at 140/90 mm Hg. Both radial and axillary pulses were palpable with normal volume. Prominent veins were present on his left shoulder and arm (Figures 1 and 2).

Laboratory investigations revealed a normal haemogram, blood sugar, lipid profile, renal, and liver function tests. Haematologic tests for hypercoagulable conditions were all within normal limits. X-ray of the chest, and cervical spine, ECG, and echocardiogram were normal. The left upper limb venous Doppler study revealed partial thrombus in the distal axillary vein and near total thrombus in the subclavian and basilic vein with mild subcutaneous edema in the axillary region (Figure 3). Multislice CT scan of the thorax using non-ionic intravenous contrast revealed venous thrombosis and adjacent soft tissue stranding in the left axillary region (Figure 3). Multislice CT scan of the thorax using non-ionic intravenous contrast revealed venous thrombosis and adjacent soft tissue stranding in the left axillary region with no evidence of pulmonary embolism. Based on the history, clinical examination, laboratory data and imaging results, secondary causes of deep vein thrombosis, such as central venous line placement, pacemaker insertion, hypercoagulable state, underlying malignancy, trauma and recent surgery were ruled out and a diagnosis of primary, effort-induced UEDVT/ PVSS was made. Patient was started initially on low-molecular-weight heparin (enoxaparin) as per the recommended dosage. Simultaneously oral anticoagulant therapy with warfarin was started to achieve an international normalized ratio (INR) between 2 and 3. Low-molecular-weight heparin was stopped when target INR was achieved, while warfarin was continued for six months. Significant improvement in the swelling of the affected limb was observed after a week of institution of anticoagulant therapy.

Discussion
Sir James Paget first described thrombosis of subclavian vein in 1875 but attributed the cause to vasospasm.¹ In 1884, von Schroetter postulated that this syndrome resulted from occlusive thrombosis of subclavian and axillary...
veins. In 1948, Hughes coined the term Paget-von Schroetter syndrome, also referred to as primary, effort-induced, upper extremity deep vein thrombosis (UEDVT), an important clinical entity with potential for considerable morbidity.

UEDVT is classified as either primary or secondary on the basis of pathogenesis. Primary UEDVT is either effort-induced or is idiopathic. Secondary UEDVT often develops in patients with pacemakers, thrombophilic states, central venous catheters or malignancy, and is responsible for majority of the cases of UEDVT. Primary, effort-induced UEDVT is a relatively rare condition, with incidence reported to be about 2.03 per 100,000 people per year by extrapolation from a Swedish Study. However, the incidence of the condition is not known in India. It is likely that this condition in our country is under-recognized and under-reported.

Primary, effort-induced UEDVT is a disorder of the thoracic outlet region, where the subclavian vein passes by the intersection of the clavicle and first rib. In this region, hypertrophied anterior scalene muscle lying posterior to the vein can compress it. The subclavian muscle located anterior to the vein provides the bulk at the costoclavicular junction, can further compress this area. It is however, unclear whether an anatomically smaller costoclavicular space, resulting from either hypertrophied muscle (scalenum anterior or subclavius) or abnormal bone morphology (clavicle or first rib) is required or whether this condition can simply occur without a defined abnormality.

Sixty to 80% of patients with effort-induced UEDVT report a history of vigorous exercise or activity involving repeated hyperabduction and external rotation of arm or backward and outward rotation of shoulder such as rowing, wrestling and lifting heavy weights.

Approximately 85% of patients will have symptoms within 24 hours of the inciting event. Clinical presentation includes complaints of blue, swollen, heavy, painful arm. Patients typically present with sudden onset of an aching discomfort with a feeling of heaviness, swelling, and frequently a reddish-blue discoloration of the affected upper extremity. A prominent venous pattern consisting of dilated superficial collateral veins over the upper arm, base of the neck, and anterior chest wall develops in many patients, especially if occlusion is chronic.

A Duplex colour-Doppler Ultrasonography is diagnostic as it is cheap, non-invasive and highly accurate in experienced hands. However, axial imaging through computed tomography (CT) or magnetic resonance (MR) is not of much value as no gross structural abnormality exists. Contrast venography is both diagnostic and often therapeutic, but has several drawbacks in comparison with duplex ultrasonography, such as being invasive, requirement of contrast agent and contraindicated in pregnancy.

Optimal management of PVSS currently is a subject of intense debate, perhaps disproportionate in volume to the incidence of this clinical entity. Anticoagulant therapy alone is not considered to be effective treatment. Recent data have showed that thrombolytic therapy, surgical decompression of costoclavicular junction, venoplasty, venous bypass, stents, and thrombectomy can be used as the management strategies.

References