Paget-Schroeter Syndrome Is Still Uncommon or Increasingly Common!

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1. Abstract

Case Report

Upper extremity deep venous thrombosis (UEDVT) is a condition characterised by thrombosis in the veins of the upper limb, with an increasing incidence despite remaining less common than lower extremity DVT. A primary etiological factor of UEDVT is Paget-Schroetter syndrome, also referred to as effort thrombosis, which primarily involves the axillary and subclavian veins [1]. Paget-Schroetter syndrome is often concomitant with thoracic outlet syndrome. Early diagnosis and intervention are imperative for favorable clinical outcomes. Point-of-care ultrasound (POCUS), as a bedside diagnostic modality, has demonstrated significant efficacy in the early detection of this condition. Herein, we report a case of a 38-year-old male in which POCUS was pivotal in diagnosing Paget-Schroetter syndrome, thereby enabling expedited therapeutic intervention.

2. Introduction

Upper extremity deep venous thrombosis (UEDVT) is a condition characterised by thrombosis in the veins of the upper limb, with an increasing incidence despite remaining less common than lower extremity DVT. A primary etiological factor of UEDVT is Paget-Schroetter syndrome, also referred to as effort thrombosis, which primarily involves the axillary and subclavian veins [1]. This syndrome is predominantly observed among healthy adults, with a marked male predominance, and is frequently associated with vigorous physical activity. If not promptly addressed, it may progress to severe thromboembolic complications. Paget-Schroetter syndrome is often concomitant with thoracic outlet syndrome. Early diagnosis and intervention are imperative for favorable clinical outcomes. Point-of-care ultrasound (POCUS), as a bedside diagnostic modality, has demonstrated significant efficacy in the early detection of this condition [2]. Herein, we report a case of a 38-year-old male in which POCUS was pivotal in diagnosing Paget-Schroetter syndrome, thereby enabling expedited therapeutic intervention.

3. Case Capsule

A 38-year-old male patient, employed as a gym trainer, presented to the emergency department with a two-week history of swelling in the left upper limb, which initially was painless. Over the past two days, the patient reported a progression in both the swelling and associated pain in the left upper limb. The patient also experienced fever over the last two days, which subsided following the administration

of antipyretics. Additionally, there was a noted history of distended veins in the neck. There was no reported history of respiratory distress or other significant symptoms. The patient's past medical history was unremarkable for type 2 diabetes mellitus, hypertension, bronchial asthma, or bleeding disorders. He reported no previous surgical interventions and maintained normal bowel and bladder function. The patient has a 12-year history of smoking and alcohol consumption. Upon examination, the patient was alert and oriented to time, place, and person. Initial vital signs indicated a blood pressure of 160/90 mmHg, a heart rate of 81 beats per minute, and a respiratory rate of 21 breaths per minute. Systemic examination yielded no abnormalities. Local examination of the left upper limb revealed swelling and distended veins, with a palpable radial pulse. Laboratory investigations, including blood tests and coagulation profile, returned within normal limits. The primary medical team elected to perform a bedside point-of-care ultrasound (POCUS) of the left upper limb. The ultrasound examination revealed the presence of a thrombus within the basilic and cephalic veins, extending into the axillary and subclavian veins on the left side. The veins were non-compressible. Conversely, the right upper limb displayed no evidence of thrombus formation, and the venous structures appeared normal and compressible. The patient was immediately initiated on low molecular weight heparin (LMWH) at a dosage of 0.4 mL subcutaneously, administered twice daily. He was subsequently transferred to a high dependency unit for continuous observation. Contrast venography was performed, revealing thrombus presence in the left axillary and subclavian veins. There was noted nonopacification of the left brachiocephalic trunk, left internal jugular vein, left subclavian vein, and left axillary vein. In contrast, the right side displayed normal opacification without any abnormalities. No compressive masses were identified affecting the vessels. A chest X-ray was conducted to exclude the presence of a cervical rib. Based on consultation with a vascular surgeon, oral rivaroxaban was added to the treatment regimen.

4. Discussion

The presence of deep venous thrombosis (DVT) in the upper limb is relatively uncommon. Primary thrombosis in the upper extremity can occur spontaneously or as a result of thoracic outlet obstruction. A specific variant of thoracic outlet obstruction is Paget-Schroetter syndrome, commonly referred to as effort thrombosis [3]. This condition is predominantly

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observed in young male patients engaged in high-intensity activities, such as gymnastics. The pathogenesis of thrombus formation may arise from an acute single insult or from a chronic insult due to cumulative effects. Anatomical abnormalities also contribute to thrombus formation, such as the presence of a cervical rib, congenital fibrous bands, hypertrophy of the scalenus tendon, or abnormal insertion of the costoclavicular ligament. These anomalies can diminish the costoclavicular space, reduce mobility, and lead to compression and stasis of blood flow. Additionally, increased venous trauma from vigorous hand movements can exacerbate the risk of thrombus [3]. The patient, a professional gym trainer, was diagnosed with Paget-Schroetter syndrome, presenting with acute onset pain and swelling in the upper limb, accompanied by dilated superficial collateral veins, known as Urschel's sign. In this case, point-of-care ultrasound (POCUS) was pivotal in establishing a prompt diagnosis, allowing immediate initiation of treatment in the emergency department [2]. We also conducted investigations to assess both inherited and acquired thrombophilic states. Additionally, a chest X-ray was performed to exclude the presence of a cervical rib.

Management of upper extremity deep venous thrombosis involves a combination of thrombolytic therapy and invasive interventions. For symptoms persisting for less than two weeks, thrombolytic therapy is often sufficient. Catheterdirected thrombolysis using fibrinolytic agents has recently demonstrated increased effectiveness [4]. Following thrombolysis, venography is performed to confirm successful vein recanalization. If the affected vein remains occluded,

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repair, may be indicated. Anticoagulant therapy is recommended for all patients to prevent further thromboembolic events. Common complications associated with Paget-Schroetter syndrome include pulmonary embolism (PE) and postthrombotic syndrome. The incidence of pulmonary embolism in conjunction with Paget-Schroetter syndrome ranges from approximately 10% to 25% [5]. However, these complications can be mitigated with early diagnosis and prompt therapeutic intervention.

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