Paget-Schroetter Syndrome

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Summary
Paget-Schroetter Syndrome is a primary, effort vein thrombosis of the axillo-subclavian level. It occurs predominantly after repetitive sport movements in the dominant limb in young healthy male individuals. Patients refer with pain, swelling and blue discoloration in the affected upper extremity.

We present a 33-years-old woman with primary thrombosis of left subclavian vein, with no history of repetitive sport exercises or trauma. The patient’s profession requiring leaning against the desk for hours daily could be a trigger factor for vein thrombosis in presented case. Doppler ultrasonography and venography showed complete occlusion of left subclavian vein. Computed tomography showed no evidence for compression at thoracic outlet level. Patient was successfully treated with anticoagulants and followed for 21 months with repeated duplex ultrasound examinations. The symptoms had resolved. Long time left subclavian vein patency and good quality of life have been achieved.

Upper extremity deep vein thrombosis (UEDVT) divides into a primary and secondary form. Primary UEDVT occurs spontaneously in young and otherwise healthy individuals, without apparent risk factors, after strenuous repetitive activity involving the upper extremity. Whereas secondary UEDVT occurs in elder patients with severe comorbidities – cancer and central venous catheter (9, 17).

The Paget-Schroetter Syndrome (PSS) is a primary axillo-subclavian vein thrombosis due to compression in costo-clavicular space at the thoracic outlet. James Paget described spontaneous axillo-subclavian vein thrombosis in 1875 (2). Vascular trauma as a potential etiologic factor was defined 9 years later by Leopold von Schroetter (3). PSS pathogenesis is distinct from other venous thromboembolic disorders. PSS is a relatively rare disorder, with incidence of 1–2 per 100,000 populations, compared with UEDVT in general and lower extremity deep vein thrombosis, with incidence of 16 and 91 per 100,000, respectively. PSS occurs in young healthy individuals who use strenuous shoulder-arm motion causing repetitive trauma. Patients suffering from PSS refer with pain, swelling, blue discoloration in the affected upper extremity (1, 7, 17).

Anatomical abnormalities at the thoracic outlet is an additional, relevant factor indicating and progressing effort thrombosis (1, 7). The primary complications of PSS are chronic venous hypertension and post-thrombotic syndrome.

Duplex ultrasonography, despite lack of high-quality validation data, is a first-line imaging technique to prove presence of thrombus. Computed tomography (CT) and magnetic resonance may be applied in case of not diagnostic ultrasonography (1). Venography enables both: diagnosis (visualization of thrombus) and treatment (catheter-directed thrombolysis and planning surgery). Thus, it remains a gold standard (1, 16).

Management consists of anticoagulation, thrombolysis, operative decompression and endovascular interventions (12, 16).

Case presentation
A 33-years-old woman with no medical history, referred with swelling and pinky blue discolouration of left upper extremity lasting over 2 months. She had right-hand dominance. Past medical history was not
significant, without strenuous, competitive sport activity. When detailed history was taken we noted that the patient was jeweller by trade. She leaned against the desk for hours each day with her left hand holding a piece of jewellery and right hand making small, precise movements. This could explain affecting a non-dominant limb. She had no surgical history.

At the time of presentation her left upper extremity was edematous, radial pulse was palpable, capillary refill was brisk, extensive dilated subcutaneous collaterals could be easily seen on left infraclavicular and deltoid region.

Complete blood cell count and coagulation profiles including protein C, protein S, homocysteine, and antithrombin III were normal. Rheumatologic markers and tumor markers were within normal ranges. CT scanning did not reveal any occult malignancy.

Duplex venous ultrasound showed patent left brachial and axillary veins. The left subclavian vein was patent laterally, with thrombus observed in proximal part. Multiple collateral veins were also observed.

Left upper extremity venogram (Fig. 1 a, b) showed patent axillary and subclavian veins laterally from costo-clavicular space, at the site of the thoracic outlet. Total proximal occlusion of the subclavian vein and prominent collateral circulation bypassing the thrombus through jugular vein have been revealed. CT with tridimensional reconstruction (Fig. 2) showed no evidence for compression at the thoracic outlet level.

The treatment was initiated with intravenous unfractionated heparin anticoagulation. The left upper extremity was elevated and compressed. Due to long period from symptoms onset, what is contraindication for catheter-directed thrombolysis, patient was treated with anticoagulation and compression alone. The swelling partially resolved and the patient was discharged from the hospital with subcutaneous low molecular weight heparin. Four weeks later, repeat venography (Fig. 3) showed persistently occluded left subclavian vein.

The patient was followed for 21 months with duplex scans repeated each 3 months. After 6 month of anticoagulation the duplex ultrasound revealed patent brachial and axillary veins, post-thromboticaly stenosed, but patent subclavian vein and collateral veins. At this time symptoms had partially resolved. The left forearm and arm remained slightly edematous, with 2–3 cm increased girths. The administration of anticoagulants was prolonged for next 4 months.

At the 21-months follow-up symptoms had resolved, with no swelling, redness or pain. The left forearm and arm girths were not increased. The dilatation of collateral veins on left infraclavicular and deltoid region had significantly diminished. The repeated duplex ultrasound scanning revealed stenosed but still patent left subclavian vein. Both long time subclavian vein patency and good quality of life have been achieved.

Discussion

PSS preferentially affects young, healthy men, predominantly in dominant limb.
Precipitating, usually sport-related event can be defined by majority of patients. Thrombosis usually follows forced arm abduction, retroversion and extension. Despite PSS is usually associated with sporting activities and is mostly seen in young athletes such as tennis players, baseball pitchers, weight lifters, swimmers (1, 17), in described case of right-handed female suffering from left-side thrombosis. The sport related effort as a risk factor of thrombosis was excluded. When detailed trauma history was taken we noted that the patient employed as jeweller worked leaned against the desk for hours each day. In our opinion it could be a trigger factor for subclavian vein thrombosis in non-dominant limb.

The most common symptoms are swelling, arm discomfort, heaviness, red discoloration, cyanosis, dilated visibly veins across the shoulder and upper arm (1, 17), what was also found at the time of our patient’s presentation. Symptoms are usually acute (1), but chronic symptoms, as in our case, are possible as well.

The subclavian vein crosses between clavicle, subclavius muscle, anterior scalenus muscle, first rib and costo-clavicular ligament. Vein is frequently compressed while traversing this tunnel, especially when the subclavius muscle is hypertroph. Repetitive mechanical compression between first rib and the lower border of the clavicle is a common mechanism of injury to the vein (13, 14).

Anatomical abnormalities such as cervical rib, congenital bands, hypertrophy of scalenus tendon, abnormal insertion of costo-clavicular ligament are additional risk factors for costo-clavicular compression. They lead to repetitive endothelial trauma. Narrow costo-clavicular space crowds the vein and restricts its mobility, what reduces vein resistance to repetitive trauma and leads to intimal hyperplasia, inflammation, fibrosis and extensive collateral formation (1). Thoracic outlet syndrome consists of venous, arterial and neurological symptoms single or coexisting (13). Prior to thrombosis in most cases thoracic outlet obstruction is asymptomatic. But if it occurs, in over 90% neurological symptoms coexist, due to compression of brachial plexus (9, 10). In presented case CT scanning with tridimensional reconstruction (Fig. 2) did not reveal compression at the level of thoracic outlet and anatomical abnormalities in this region were excluded.

Ultrasonography is the preferred initial test evaluating suspected thrombosis. It is non-invasive, repeatable examination with a sensitivity of 97% and specificity of 96% for the diagnosis of UEDVT. However, small thrombotic segments can be missed on ultrasound, especially when they are located just beneath the clavicle (1, 9). We choose the duplex ultrasound scanning for the initial examination and follow-up the patient due to its non-invasiveness and high sensitivity and specificity. The patient underwent duplex scanning each 3 months during the 21-months long follow-up.

Despite that venography is not always necessary for diagnosis and is invasive, it remains a gold standard for PSS diagnosis. It enables detection of thrombus, catheter-directed thrombolysis as well as detection of compression and planning surgery (1, 16). Our patient underwent venography twice. The initial venography (Fig. 1) revealed a patent left axillary and subclavian veins laterally from costo-clavicular space, total proximal occlusion of the subclavian vein and prominent collateral circulation bypassing the thrombus through jugular vein. Four weeks later repeat venography (Fig. 3) showed persistently occluded left subclavian vein.

CT venography is superior to ultrasonography and available, but the risk of radio-contrast administration cannot be forgotten. Highest sensitivity (100%) and specificity (97%) has MRI venography. Although MRI venography are increasingly used, the availability is limited and costs are high (1).

Conservative management for PSS are limb elevation, compression and anticoagulation. There are significant incidence of residual symptoms, limb disability and recurrent thrombosis with this management alone (1). Thus aggressive multimodal treatment strategies with catheter-directed thrombolysis, thrombectomy, percutaneous venoplasty and stents, surgical venoplasty, venous bypass as well as thoracic outlet decompression are essential (1, 4–6). Catheter-directed thrombolysis, compared to systemic thrombolysis has the therapeutic value without systemic side effects. It is recommended for patients presenting early, within 2 weeks of symptom onset, although some successful outcomes were reported with 4–6 weeks delay.

However, the time from symptom onset to thrombolysis diminishes the success of thrombolysis (1, 9, 16). The reported patient was treated initially with intravenous unfractionated heparin, limb elevation and compression. She did not undergo thrombolysis due to the delay from symptoms onset and symptoms resolving following conservative management. The patient was kept on anticoagulants for 10 months, what is reported to be a suitable period (15). The repeated duplex ultrasound examinations revealed stenosed, but patent left subclavian vein. In a 21-month follow-up the left subclavian vein remains patent. The patient could not be treated with a elastic compression sleeve because this precluded her manual work as jeweller.

Surgical decompression of thoracic outlet is performed if thoracic outlet obstruction is present. In case of repeated residual venous stenosis venography angioplasty should be performed. This management first involves rib resection, scalenectomy and division of the costo-clavicular ligament (1, 12).

There are no statements regarding the role of decompression. Some authors reserve thoracic outlet decompression only for patients whose symptoms are persistent or recurrent after thrombolysis. Others rec-
ommend early and routine decompression, due to suboptimal results, higher incidence of recurrent thrombosis and residual symptoms with delayed surgery (4–6, 8).

There are two approaches: transaxillary introduced by Roos in 1966 (11) and subclavicular anterior route, which seems to be a approach of choice (10), although both approaches have limitations. The transaxillary approach does not allow direct repair of the vein which is frequently needed (10). As postoperative complications nerve, vein or artery injury, bleeding or pneumothorax were noted. The balance between risks and benefits of surgery is crucial (1, 8, 10). Anticoagulation is also required after surgical decompression. Although there are no statements, long term anticoagulation should be recommend in case of suboptimal result after surgery or late presentation as well as coexistent thrombophilia (1).

Vein stenting, surgical thrombectomy, balloon venoplasty, patch venoplasty and venous bypass have limited use and poor long-term patency rate. They are recommended in case of residual stenosis after decompressive surgery. Due to risk of stent crushing, subclavian vein stenting is permissible after surgical decompression (4–6, 10).

Although described in the literature (1), during the 21-months follow-up PSS complications such as pulmonary embolism, post-thrombotic syndrome or prolonged shoulder disability did not reveal.

Conflict of interest
The authors declare no conflict of interest. This research received no specific grants from any funding agency in the public, commercial, or not-for-profit sectors.

Ethical guidelines
Informed patient consent has been obtained. The case report was conducted accoding to the recent Declaration of Helsinki and the national guidelines.

References