

Pre Paget-Schroetter syndrome: a case study

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Abstract:

Paget-Schroetter syndrome is an acute subclavian vein thrombosis that usually develops secondary to heavy arm exertion. This case study illustrates a venous anomaly identified in a 26-year-old male with an acutely swollen left arm who was referred to our service for an upper extremity venous duplex examination for possible deep vein thrombosis (DVT). The study was negative for DVT but positive for subclavian vein stenosis and abnormal flow which totally terminated upon raising the arm a mere 10 degrees. This case illustrates how duplex scanning can be an important tool in the identification of venous stenoses and obstructions and abnormal flow patterns during physiologic movement.

Introduction:

Peripheral nerves and blood vessels often course through small apertures, long tunnels, and narrow areas to reach their end points. All along their way those nerves and blood vessels are subject to irritation and compression by the various structures that they traverse. One such area prone to irritation and compression is at the costoclavicular space. The subclavian vein courses through this costoclavicular space where it is bound by the first rib inferiorly, the costoclavicular ligament anteriorly, the clavicle superiorly, and the

anterior scalene muscle posteriorly. Venous compression can occur with extensive movements of the shoulder girdle and as a result of variations in the size and position of the surrounding muscles and ligaments. Over time, repetitive intermittent compression can damage the subclavian vein and may eventually lead to scarring, narrowing, and finally thrombosis. This condition is referred to as Venous Thoracic Outlet syndrome. There may be a neurological component or vascular component or both as well.ⁱ Venous involvement of this condition is referred to as Paget-Schoetter syndrome. If the vein goes on to total thrombosis with typical symptoms of swelling, venous engorgement and discomfort.^{ii,iii} The case study presented below is an example of primary venous obstruction probably resulting from an anatomically narrowed thoracic outlet aggravated by mobility of this area worsened by his line of work which involved moving and stacking boxes repetitively above shoulder level.

Methods:

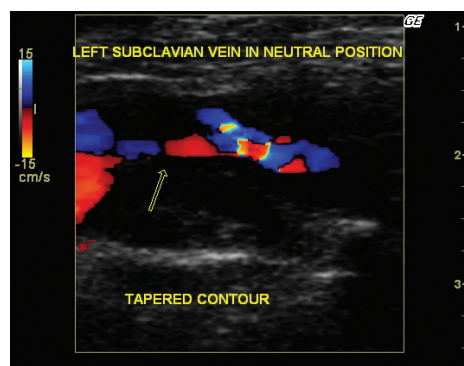
A 26-year-old male with no previous significant medical history presented to the Vascular Lab and Ultrasound Service for acute onset of left upper extremity edema. His right upper arm was noted to



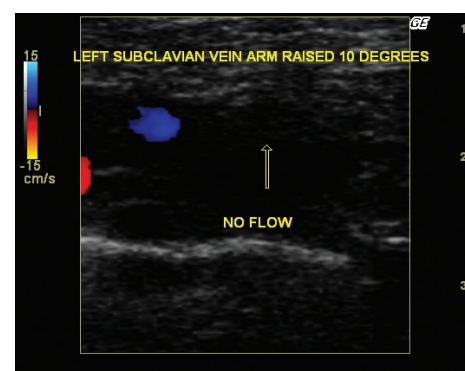
be 31.5 cm in circumference, the left upper arm 36cm, the right forearm 30 cm, and the left forearm measured 32 cm. He initially was seen earlier the same day at a community hospital emergency room where he underwent a CT scan of his chest with intravenous contrast that was found to be normal and he had a left upper extremity venous Doppler study performed that was also found to be normal. He was subsequently discharged and sent to our service for evaluation and treatment. The patient underwent a bilateral upper extremity venous duplex evaluation by a registered vascular technologist in our ICAVL vascular laboratory using a GE Logic 9 ultrasound imager (Milwaukee, WI). A 10Mhz linear transducer was used to evaluate the subclavian, jugular, axillary, brachial, radial and ulnar veins for the presence of DVT bilaterally with the patient in a sitting (neutral) position. Venous Doppler signals were obtained in longitudinal view with Doppler cursor angle parallel to flow. All veins insonated were evaluated for co-aptation in transverse view. The same evaluation was then performed with the arms elevated, and responses were recorded.

Results:

No evidence of DVT was found in either upper extremity by duplex scan. However, significant luminal tapering was noted in the left subclavian vein when compared to the contralateral subclavian vein (Fig 1).



The left subclavian Doppler signal was abnormal and had continuous flow that did not change in response to respirations or augmentation when compared to the contralateral subclavian vein flow. The patient was subsequently seen by our cardiothoracic surgeon for consultation due to the abnormal findings on duplex exam. With the surgeon in attendance, the patient's subclavian veins were scanned again to further evaluate response to arm maneuvers. With only a 10-degree elevation of the left arm, the patient developed total occlusion of the proximal portion of the left subclavian vein (Fig 2). At 90 degrees of elevation the right subclavian vein occluded in a similar fashion.



The loss of venous flow with minimal arm maneuvers and the stenosis of the subclavian vein in this patient are consistent with that of "pre Paget-Schroetter syndrome". We believe that this case demonstrates the intermediate stage in the evolution of Paget-Schroetter syndrome: the subclavian vein stenosis that was identified by duplex represented the stage somewhere between the initial injury and complete thrombosis of the subclavian vein.

Discussion:

Concordant with other study populations with Paget-Schroetter syndrome, this patient uses his upper extremities for repetitive lifting in his job as a clerk where he stacks boxes. Patients who are weight lifters, who perform repeated lift-

ing, and / or wear heavy backpacks with straps over the shoulders are at higher risk for developing complications related to repetitive injury of the subclavian vasculature from repeated mechanical compression of the vessels between clavicle, 1st rib and the costoclavicular ligament.^{iv} Patients with intermittent venous occlusion without thrombosis often present with subtle onset of intermittent pain and swelling caused by partial or acute occlusion at the level of the costoclavicular ligament. The degree of venous collateralization in the presence of complete thrombosis may make the severity of symptoms vary. Pain and swelling of the arm is typically aggravated by exercise or work with the arm and are usually relieved with rest.^v This patient presented to our institution before his subclavian vein had thrombosed. There was no evidence intraluminal thrombus nor was there evidence of significant venous collateral flow noted on duplex evaluation.

Historically, the primary diagnostic tools for the diagnosis of Paget-Schroetter syndrome were clinical history, physical examination, and venography.^{vi} As demonstrated in this case, venous duplex scanning may also be used to detect venous obstruction in the axillary – subclavian veins. However, the patient had a previous venous duplex scan at an outside institution that was found to be normal. In contrast, the venous duplex study that was repeated in our institution the same day found abnormal left subclavian vein flow and that the vein looked abnormally tapered in appearance when compared to the contralateral subclavian vein. The disparity in the findings may be related to several factors, one being that identification of veins in the thoracic outlet by duplex can be limited by bony structures in the neck and shoulder.^{vii} Other reasons include, but are not limited to: the presence of large collaterals that could be mistaken for the occluded main channel, mistaking recanalized veins for normal veins, and that partial or

intermittent obstructions may be difficult to evaluate with duplex scanning.^{viii} Both venous duplex scans were in complete agreement with the CT scan that there was no evidence of DVT by duplex in either upper extremity. This particular case study demonstrates that by comparing the appearance and hemodynamics of the vein of the right extremity to the left extremity, a venous stenosis was identified. Furthermore, by abducting the arm after a thorough exam was performed in neutral position, we were able to reproduce and directly visualize the mechanical compression of the subclavian vein. From this duplex information, the clinical decision making pathway was expedited and the patient treated in a timely fashion.

Typically, after diagnosis of Paget-Schroetter syndrome is made, treatment options for subclavian vein thrombosis are catheter directed thrombolysis followed by surgical thoracic outlet decompression (first rib resection) in selected patients.^{ix, x, xi} The desired goals for these patients are to restore patients to normal function, maintain venous patency, and to prevent recurrent thrombosis.

In summary, the case illustrates the efficacy of a complete duplex evaluation to aid in the identification and diagnosis of patients with pre Paget-Schroetter syndrome, allowing intervention prior to the development of thrombosis.

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