MANAGEMENT OF SUBCLAVIAN VEIN THROMBOSIS KNOWN AS PAGET-SCHROETTER SYNDROME

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Summary

Subclavian vein thrombosis, known also as Paget-Schroetter syndrome, is a surgical disease. Conservative treatments with anticoagulants alone, or even with thrombolytics and balloon angioplasties, are inadequate. . Decompression of the thoracic inlet is mandatory in all cases, along with direct repair of the subclavian vein.

1. Definition

Paget-Schroetter syndrome is a condition that involves thrombosis of the subclavian vein. It is also known as subclavian vein "effort thrombosis" or primary subclavian-axillary vein thrombosis. It affects mostly young people active in physical work or sports. It has a sudden occurrence and if not treated promptly, ideally within 2 weeks of the original event, it leads to rapid occlusion of the distal arm veins and evolves into fibrosis and total obliteration of those veins. The process is irreversible unless prompt intervention is undertaken. The consequence, if the treatment is not implemented promptly, is eventual permanent disability of the affected arm.

2. Anatomy of the Obstructing Site.

The obstructing mechanism, which is extrinsic to the vein, is formed by the first rib below, the subclavius muscle tendon in front, and the anterior scalene muscle in the back.

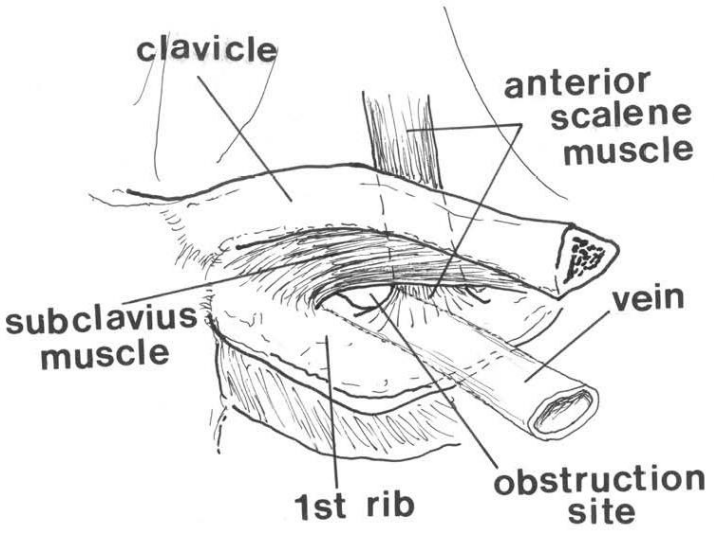

Figure 1. A left sided thoracic inlet illustration shows the tunnel through which the subclavian vein runs into the mediastinum. The three structures causing the compression are the subclavius muscle, the anterior scalene muscle and the 1st rib.

When a sudden or repeated effort is done with the arm, the vein gets crushed between these structures, and the endothelium is injured. Thrombosis occurs which propagates along the distal vein, occluding it totally, leading to severe venous congestion of the arm and edema.

3. Diagnosis

The patient usually presents with a swollen and painful arm, frequently with a bluish discoloration of the skin. If the patient has had recurrent episodes in the past, the arm is not as edematous, but there are multiple enlarged tortuous veins under the skin as collateral circulation develops to circumvent the obstruction site. The initial test of choice is to obtain an ultrasound of the subclavicular area. This test routinely makes the diagnosis showing the lack of flow into the vein and absence of pulsatility of the venous flow.

After confirming the diagnosis with the ultrasound exam the next step is to perform a venogram done by the interventional radiologist.

Occasionally, because of the edema present in the arm, it is difficult to visualize the vein to be punctured. Therefore the radiologist may resort to the use of an ultrasound device to locate the brachial vein and accomplish the puncture with insertion of a catheter to obtain the radiographic image. The venogram is essential to determine the extent of the thrombotic process and access whether an adequate inflow still exists. This is essential to determine whether or not the patient is still a surgical candidate.

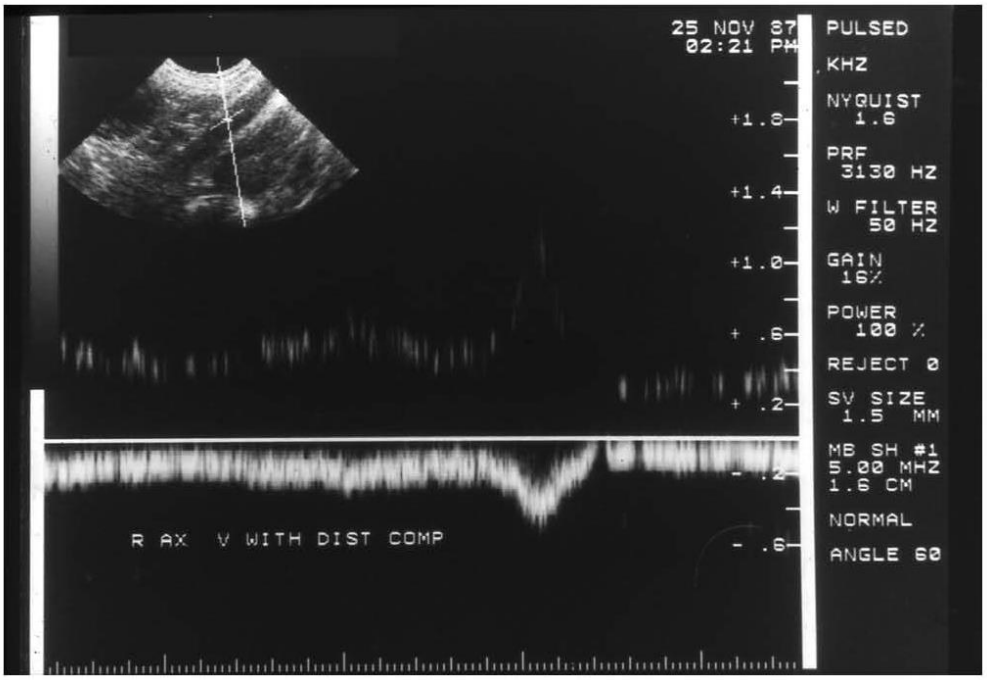

Figure 2. An ultrasound of the subclavian vein is obtained showing complete lack of pulsatility in the tracing with the flat straight line. The only deflection is the result of manual compression of the vein.

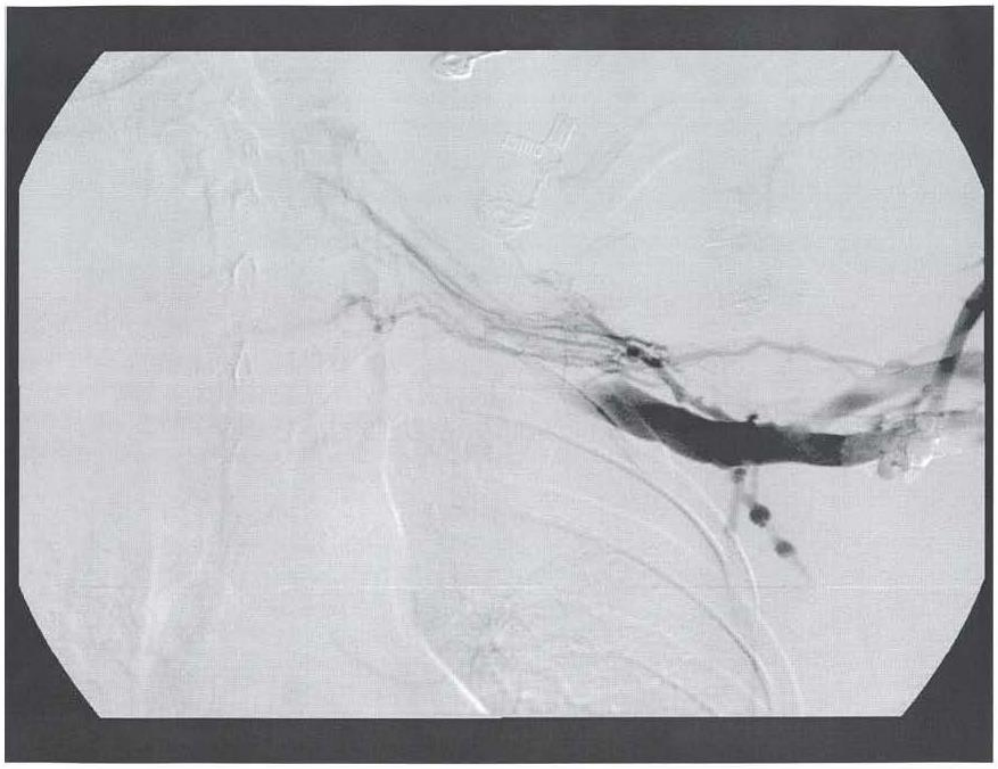

Figure 3. Typical initial venogram showing complete obstruction of the left subclavian vein at the level of the thoracic inlet.

4. Immediate Therapy

4.1. Heparin

Once the diagnosis of venous thrombosis is made by ultrasound, the patient must be started on intravenous Heparin therapy in order to arrest further progression of the thrombotic process and to preclude the occurrence of new thrombus. The infusion of Heparin should be placed on the opposite arm to leave the affected arm free for the manipulations that will be necessary during the course of the patient's treatment. If the patient eventually goes to surgery, the Heparin is usually stopped 4 hours before the time of the operation.

5. Thrombolysis

Immediately upon confirming the diagnosis and the extent of the problem with a venogram, thrombolytic therapy should be started immediately by using the catheter induced infusion technique. The infusion catheter is positioned by the interventional radiologist directly in the thrombus and the infusion of the chosen thrombolytic agent is started using a reliable electrically operated pump set at the appropriate rate. Several thrombolytic drugs have been used. In the 1980's Urokinase (Abbokinase, Abbott Laboratories) was the drug of choice. After 1999, when this drug was removed from the market by the FDA, RTPA thrombolytic agents have been in widespread use. However, in other countries where Urokinase is still available, it can be used at the recommended dose of 2200 units per Kg weight per hour through the infusing catheter. Currently the new thrombolytic agents that have been in use since 1999 are Tenecteplase (TNK, Genentech Laboratories), Reteplase (Retavase, Centrocor Laboratories), and Alteplase (TPA Activase Genentech Laboratories).

TNK is dosed at 0.25 to 0.3 mg/hr or adjusted to 0.05 mg/kg/hr. The Reteplase is used at 0.5 to 1.0 units per hour or at 0.01 units/kg/hr. Alteplase is dosed at 0.05 mg/kg/hr or 0.2 to 10 mg/hr.

The time it takes to dissolve the clot is always less than 24 hours.

The progression is monitored by repeating the venogram at 8 or 12 hours if needed. The thrombolytic agents are infused through the catheter placed inside a sheath, which is usually left in place after the thrombolytic therapy is completed, because it will be used after surgery for a repeated venogram to assess the adequacy of the surgical repair. If indicated, the sheath is also be used to gain access to implant an endovascular stent to correct residual obstruction.

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Biographical Sketch

J. Ernesto Molina, M.D., Ph.D. is Professor of Surgery at the University of Minnesota, Minneapolis, Minnesota, and is board certified in General Surgery and Thoracic Surgery. He received his M.D. from the University of San Carlos de Guatemala in 1960, and a Ph.D. in Surgery from the University of Minnesota in 1978. He is recognized as a leading authority on Thoracic Outlet Syndrome, and has numerous publications on the subject in such journals as Annals of Thoracic Surgery, Journal of Thoracic and Cardiovascular Surgery, the International Journal of Angiology, Journal of Vascular Surgery, and others. Contributor to textbooks of Cardiac and Chest and Vascular Surgery. Author of the textbook: New Techniques for thoracic outlet syndrome. Springer Publisher 2012. He is a member of the American Association for Thoracic Surgery, Society of Thoracic Surgeons, Society for Vascular Surgery, American College of Surgeons, International College of Angiology, and others.