

Paget-Schroetter Syndrome

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Case prompt:

23 year old male presents to the Emergency Department with complaints of right sided neck pain, right arm pain and swelling. Patient states that the symptoms have been ongoing for the past 9 months. He is a pitcher for the local college team. He denies trauma or injury to his neck, shoulder or arm. The pain and swelling in the right upper extremity have been slowly getting worse over time, his neck pain has started recently. He states his arm feels heavy. He denies numbness in the right upper extremity, palpitations, headaches, dyspnea. He has no other medical problems and does not take any medications. He denies illicit drug, alcohol and tobacco use.

His vital signs are normal. His exam is pertinent for swollen right upper extremity that appears slightly larger than left upper extremity. Patient has full range of motion, no muscle weakness or instability on testing. No midline neck tenderness or exacerbation of pain on Spurlings or neck range of motion. Patient is neurologically intact and has 2+ radial and ulnar pulses. What is the most likely diagnosis?

- A. Superior Vena Cava Syndrome
- B. Cervical Radiculopathy
- C. Upper Extremity Lymphedema
- D. Paget-Schroetter Syndrome
- E. Upper Extremity Deep Vein Thrombosis

Answer: Paget-Schroetter Syndrome (PSS)

Background:

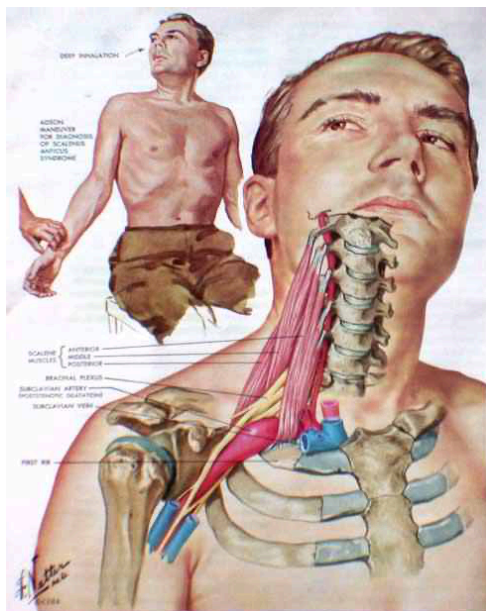
- Paget-Schroetter Syndrome is an effort-based thrombosis of the Axillary and Subclavian vein associated with compression of the Subclavian vein at the thoracic outlet.^{1,2,3}
 - Venous variant of Thoracic Outlet Syndrome (TOS)
- Seen in younger adults (20-30s) with a 2:1 M:F ratio¹¹
 - Most of these are athletes who perform repetitive overhead movements
- Venous Thoracic Outlet Syndrome is the rarest of the etiologies of Thoracic Outlet Syndrome¹¹
 - 1-2 per 100,000 are reported

Pathophysiology:

- Subclavian vein traverses between the first rib, clavicle, and the anterior scalene muscle⁵
 - Repetitive physical activity and compression can lead to compression and subsequent venous injury/thrombosis
 - This can lead to decreased venous return and extremity edema and pain
- Overtime, collateral venous flow will bypass the area of thrombosis¹¹
 - Inflammation from the thrombosis can lead to vein collapse and scar tissue formation

Clinical Presentation:

- Patients will present with worsening upper extremity edema and pain
 - Can be acute, subacute or chronic during presentation
- Typically, an inciting, strenuous exercise will precede symptoms and can be elicited from History of Present Illness
- On exam, affected arm will be edematous and appear enlarged in comparison to contralateral side
 - Can appear tense and cyanotic
 - Urschel's sign may be present
 - Venous collaterals present around the shoulder and chest¹⁰
- Arm will feel heavy to the patient, but range of motion and muscle strength will be intact
- Confirm neurovascular status of affected extremity



Anatomy of Thoracic Outlet



Urschel's Sign in Paget Schroeder Syndrome
Lowless, Sean M. et al.
The American Journal of Medicine, Volume 130, Issue 12, e537

Diagnosis:

- Confirm presence of coagulation disorders in family history or past medical history
- Upper Extremity ultrasound can assess for Deep Vein Thrombosis (DVT), collapsed veins and chronic clot burden¹¹
 - Computerized Tomography (CT) Venography and Magnetic Resonance (MR) Venography is better suited for eval of central venous system
- Laboratory workup should include thrombophilia panel (Proteins C&S, antithrombin levels, Factor V Leiden mutation testing, etc) along with D-Dimer levels¹¹
- If patient has signs and symptoms of Pulmonary Embolism (tachycardia, tachypnea, hypoxia), include a CT Angiogram of the chest

Management:

- Surgical decompression along with anticoagulation has 90% successful treatment ^{4,9,11}
 - First Rib resection along with optional resection of surrounding structures to decompress the thoracic outlet⁷
- Anticoagulation can be utilized as well ^{6,8}
 - Intravenous Heparin preoperatively and short course dual oral anticoagulant post-op

DDx:

- Lymphedema, Deep Vein Thrombosis, Acute Arterial Ischemia, Congestive Heart Failure, Superior Vena Cava syndrome, Neurogenic Claudication, Cervical Radiculopathy, Thoracic Malignancy

Clinical Pearls:

- Thoracic Outlet Syndrome (TOS) affects many athletes, especially ones with repetitive motion in their sport
 - Venous variants are less prevalent than neurogenic and arterial TOS
- Patient will present with more insidious onset including heaviness, pain and edema
 - Always assess neurovascular status
- During initial workup, evaluate for thrombophilia and rule out signs and symptoms of a Pulmonary Embolism
- Gold standard treatment is surgical decompression but oral anticoagulation can aid in management

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