Paget-Schroetter Syndrome in an Overhead Athlete

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Introduction
Paget-Schroetter syndrome, or “effort-induced” thrombosis, describes venous thrombosis of the axillosubclavian vein in the upper extremity. The hallmark of this syndrome is the formation of thrombus with associated excessive upper extremity activity in the absence of secondary causes. Although this is an unusual presentation for upper extremity venous thrombosis, it has been associated with athletic activity, particularly in athletes who participate in sports with significant overhead activity (e.g., baseball, volleyball). It is important for physicians to keep this disease process in mind when evaluating arm pain, as prompt treatment can significantly reduce future morbidity.

Case Report

History
A 20-year-old collegiate baseball player presented to the training room with 36 hours of swelling and discoloration of his proximal right upper extremity. Presentation was just prior to the beginning of the fall (off-season) practice season. He is a left hand-dominant outfielder. The patient stated that he noted his right arm was “turning blue” and felt “stiff.” He denied recent trauma or illness. He was participating upper body strength training as a part of preseason conditioning, but noted no recent changes in the program. He denied any numbness or weakness in the right arm.

Physical examination
Gross inspection revealed mild circumferential ecchymosis overlying the proximal one fourth of the right upper extremity. There was full active and passive range of motion. There was no point tenderness with palpation. There was no deficit in motor strength. There were no sensory deficits in the involved upper extremity. Radial pulses were full and equal bilaterally. Circumference of the proximal right upper extremity was 34 cm compared with 30 cm on the left. The patient had full range of motion of the neck without pain. Adson’s test was positive. Spurling’s test was negative.

Differential diagnosis
1. Axillosubclavian venous thrombosis
2. Upper arm contusion
3. Allergic reaction
4. Thoracic outlet syndrome
5. Unilateral brachial plexopathy
6. Subclavian artery aneurysm
7. Pott’s syndrome
8. Bony/soft tissue tumor

Tests and results
Ultrasound of the right upper extremity was ordered for the following morning. Results showed venous thrombosis in the right subclavian vein.

Working diagnosis
Subclavian vein thrombosis, likely Paget-Schroetter syndrome (effort-induced axillosubclavian vein thrombosis).

Treatments
1. Patient was admitted for catheter-directed thrombolysis (day 1)
2. Urokinase was infused and resolution of the clot was obtained over a period of approximately 36 hours (day 3)
3. Follow-up venogram after thrombolysis showed persistent compression of the right subclavian vein (consistent with Paget-Schroetter syndrome)
4. Cardiothoracic surgery was consulted and recommended thoracic outlet decompression; risks and benefits of surgery and impact on future athletic participation were discussed prior to surgery.
5. Patient underwent a surgical procedure involving resection of the right first rib, partial resection of the right anterior scalene muscle, and stenting of the right subclavian vein to the right internal jugular vein (axillary vein bypass; day 4) 
6. Patient was switched from intravenous to oral anticoagulation with the plan to continue anticoagulation for a total of three months with a goal International Normalized Ratio (INR) of 2.0–3.0 (day 5) 
7. Patient was discharged from hospital

Outcomes
The athlete is now nearly 8 months postsurgery. He was cleared to return to upper body conditioning and participation in limited drills by 3 weeks after surgery. He completed a 3-month course of warfarin with an INR maintained between 2.0 and 3.0. Approximately 3 months postsurgery, he was given clearance for full athletic participation by his cardiovascular surgeon. He was able to begin preseason practice with the team approximately 4 months after surgery. He is currently participating in baseball and has resumed his position as starting outfielder. He has had no further complications.

Discussion
Paget-Schroetter syndrome was described separately by Sir James Paget in 1858 and Leopold von Schroetter in 1899 [1]. This syndrome describes venous thrombosis in the axillosubclavian venous system of the upper extremity. Thrombosis occurs in the presence of excessive physical activity with no other underlying etiologies (eg, hypercoagulable state, indwelling catheters). Because of the association with activity, Paget-Schroetter syndrome is also referred to as effort-induced thrombosis.

Pathophysiology
There are several anatomic considerations that lead to the development of Paget-Schroetter syndrome. The axillosubclavian vein transverses the thoracic outlet which predisposes to external compression. This vein passes through a tunnel composed of the clavicle and subclavious muscle anteriorly, the anterior scalene muscle laterally, the first rib posteriorly, and the costoclavicular ligament medially. Hypertrophy or distortion of any of these elements can lead to anatomic constriction. Common anatomic variants seen with Paget-Schroetter syndrome include the presence of a cervical rib and/or a hypertrophied anterior scalene muscle. Combined with excessive upper extremity activity, this can lead to external compression [1]. In particular, extremes of abduction and external rotation of the shoulder, motions often seen in overhead athletes, can produce strain in this area [2]. This strain can lead to microtraumatization to the venous intima, resulting in local activation of coagulation and the formation of venous thrombosis [3]. Although Paget-Schroetter syndrome is most commonly seen in the dominant upper extremity, it can occur in the nondominant extremity as well (as illustrated in the case presentation here). In a study by Sheeran et al. [4], four of 14 (29%) patients meeting the criteria for Paget-Schroetter syndrome had occurrence in the nondominant extremity.

Diagnosis
Patients with Paget-Schroetter syndrome generally present with complaints of aching pain in the upper arm and shoulder, usually exacerbated by activity. There may be associated numbness or paresthesias distally. Physical examination generally reveals swelling and discoloration at the involved site along with decreased capillary refill secondary to venous congestion. Decreased sensation to light touch may be noted. Arterial pulses should be unaffected. Occasionally, the venous thrombus can be detected by palpation as a firm, cord-like mass along the inner aspect of the upper arm [5]. A positive Adson’s test suggests compression in the thoracic outlet [1].

Conformation of diagnosis is made by either Doppler flow ultrasonography or venography. Although ultrasound is less sensitive, it is also less invasive and easier to obtain. It is therefore often the first choice for diagnostic imaging [4].

Even with a history suggestive of Paget-Schroetter syndrome, venous thrombosis in the upper extremity should prompt a laboratory evaluation for an inherited hypercoagulable state. Studies should include, but may not be limited to, assays for Factor V Leiden, prothrombin gene mutation, protein C and S, antithrombin III, and homocysteine. Other acquired hypercoagulable states include malignancy, connective tissue disease (eg, lupus), and nephrotic syndrome [5].

Once discovered, treatment for Paget-Schroetter syndrome should be initiated promptly. Although embolization to the lungs is more common with venous thrombosis in the lower extremities, the incidence of pulmonary embolism associated with Paget-Schroetter syndrome has been reported to be as high as 30%. In addition, delay in diagnosis and treatment can lead to long-term morbidity such as chronic pain, edema, and limited range of motion of the involved extremity [4].

Treatment
Treatment for Paget-Schroetter syndrome involves a multidisciplinary approach. Previously, treatment consisted of prolonged anticoagulation with associated rest of the involved extremity [1]. However, with improved technology in the field of interventional radiology, local thrombolytic therapy in conjunction with venography is becoming the mainstay for initial management. This allows for the rapid establishment of patency of the axillosubclavian vein.

Surgical options for the management of Paget-Schroetter syndrome include first rib resection, thoracic outlet decompression, and stenting of the axillosub-