Case Report

Expanding the Differential for Unilateral Arm Pain in Athletes

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Abstract

Paget-Schroetter syndrome (PSS) is a disease entity characterized by primary thrombosis formation in the axillo-subclavian vein. Repetitive arm movement in athletes causes chronic endothelial trauma leading to thrombus formation. It is critical to include PSS in the differential for athletes presenting with unilateral arm pain as a delay in diagnosis and treatment can lead to significant long-term morbidity. This case report of a division I female basketball player highlights the clinical presentation, pathophysiology, diagnosis algorithm, and treatment options for this rare but should not be missed clinical syndrome. Specifically, this case highlights the need for expert consensus and further research in determining the necessity and timing of decompression surgery and the optimal anticoagulation regimen.

ABBREVIATIONS

PSS: Paget-Schroetter Syndrome

INTRODUCTION

Paget-Schroetter syndrome (PSS) or “effort thrombosis,” although rare, is a disease entity that is crucial to understand and include in the differential for young athletes who present with unilateral arm pain [1,2]. Early diagnosis and treatment of PSS is associated with minimal sequelae; however, diagnosis delay can lead to significant long-term morbidity [3-5]. This case report highlights the clinical presentation, pathophysiology, and treatment options for this rare but should not be missed clinical syndrome.

CASE PRESENTATION

A 19 year female, right-hand dominate, Division-I NCAA female basketball player presented to the training room with a one-day history of right arm pain, described as the “sensation that she was constantly doing bicep curls.” She also noted subjective swelling without inciting injury. After attempting to practice, she noted extreme proximal arm pain resulting in the inability to raise her arm to shoot. Her past medical history is significant for alopecia areata treated with subcutaneous steroid injections. She started taking oral contraceptive pills (OCPs) for menorrhagia three months prior to symptom onset. There was no family history for hypercoagulability. Her physical exam revealed objective edema to the right upper extremity with firmness noted to the right biceps as compared to the left. She had a normal neurovascular exam with brisk capillary refill. She had good strength bilaterally in the upper extremities. A right upper extremity ultrasound was ordered which revealed an occlusive thrombus to the right subclavian vein (Figures 1 & 2). She was admitted to the hospital for emergent venography, which revealed an occlusive thrombus in the axillo subclavian segment. Catheter thrombolysis was initiated for 24 hours. A next day venography revealed a patent axillo subclavian segment but compressive stenosis as the axillary vein crossed the first rib (Figure 3). Percutaneous transluminal angioplasty was performed resulting in good flow. Laboratory work up was notable for heterozygosity to factor V Leiden. The assumption was this was a provoked clot from repetitive shooting, her heterozygosity, and the use of OCPs. Upon discharge there was considerable debate over anticoagulation recommendations. While hematology suggested 3 months of Coumadin, per vascular surgery recommendations, the patient was discharged on 2 weeks of Lovenox with frequent ultrasound monitoring in hopes of allowing the patient to return to basketball this season. A repeat ultrasound one week later revealed recurrence of the mid-distal subclavian DVT. This was followed by a positional venography to more precisely assess the hemodynamics of the compression of the subclavian vein at the site of the first rib and to further identify venous collaterals suggestive of a more chronic compression.

The persistent chronic stenosis identified suggested the need to undergo decompression of the thoracic outlet with first rib resection. The patient was also transitioned to Coumadin for 3 months, inhibiting her ability to return to basketball this season. Ultimately, the patient’s outcome was favorable. She underwent shoulder stabilization physical xtherapy for 6 weeks and remained asymptomatic allowing her to return to noncontact basketball drills. A repeat ultrasound showed just chronic fibrotic tissue formation in the axillo-subclavian vein. Repetitive arm movement in athletes causes chronic endothelial trauma leading to thrombus formation. It is critical to include PSS in the differential for athletes presenting with unilateral arm pain as a delay in diagnosis and treatment can lead to significant long-term morbidity. This case report of a division I female basketball player highlights the clinical presentation, pathophysiology, diagnosis algorithm, and treatment options for this rare but should not be missed clinical syndrome. Specifically, this case highlights the need for expert consensus and further research in determining the necessity and timing of decompression surgery and the optimal anticoagulation regimen.
with no concern for acute thrombosis. Based on these findings and clearance by vascular surgery, the patient returned to play 3 days after termination of Coumadin without further symptoms or complications.

**DISCUSSION**

PSS is a sub-category of venous thoracic outlet syndrome in which a primary thrombosis forms in the subclavian vein as it passes anteriorly in the thoracic outlet at the intersection of the first rib and clavicle [5]. A hypertrophied anterior scalene lying posterior to the vein, or subclavius muscle lying anteriorly to the vein, or smaller than usual costoclavicular space can lead to compressive force and micro-trauma to the subclavian vein thus precipitating thrombosis with repetitive abduction of the shoulder. Rigorous overhead activity alone is thought to cause thrombosis without underlying anatomic defect [5]. Chronic inflammation leads to intimal hypertrophy, and fibrous tissue replaces loose surrounding venous connective tissue. This reduces mobility of the vein and predisposes to stretch and compressive forces [4]. As in our case, patients present with acute onset of arm heaviness, swelling, and an aching discomfort. 85% of patients will describe rigorous upper extremity activity preceding symptoms [5]. In the presence of true effort thrombosis a duplex ultrasound is diagnostic with sensitivity from 78% to 100% and specificity from 82% to 100% [6-9]. In the absence of complete thrombosis excess collateral circulation is suggestive of chronic intermittent positional compression. Venography is typically done after duplex ultrasound and is both diagnostic and therapeutic. There are no randomized control studies to guide treatment, and recommendations are based solely on case series reports and expert opinions. What is well documented is the importance of early treatment with a high morbidity associated with delayed diagnosis or inappropriate treatment. Delayed diagnosis can lead to pulmonary embolism, chronic venous insufficiency, pulmonary hypertension, persistent venous obstruction symptoms and permanent disability [10]. Catheter directed thrombolysis is the standard of treatment with success rates of 84% and nearly 100% when performed within the first 48 hours of onset of symptoms [11,12]. Multiple studies have shown significantly decreased success rates when symptoms are present more than 14 days. Current best practice guidelines support aggressive surgical decompression of the thoracic outlet after thrombolysis in the treatment of primary upper extremity deep vein thrombosis. Without surgical intervention, recurrence is high due to persistent venous stenosis from underlying anatomic

Figure 1 Doppler ultrasound showing distention of the right subclavian vein with intraluminal thrombosis.

Figure 2 Doppler velocity ultrasound revealing lack of blood flow through the subclavian vein.

Figure 3 Intraoperative angiography that highlights a hemodynamically significant stenosis of the subclavian vein as it passes over the first rib.
pathology or chronic inflammatory changes. However, the timing of decompression surgery is under debate. Most current consensus recommendations suggest that early decompression surgery decreases thrombosis recurrence, returns a patient to functional status sooner, and is associated with minimal complications [5]. The final consideration in treatment is the length and choice of anticoagulation. There are no randomized studies to direct anticoagulation type or duration, and decisions are based solely on expert opinion. This was of particular concern in our patient as initially there was conflicting expert recommendation. Current consensus suggests anticoagulation for 3 months with repeat venography [5,13,14]. Overall, PSS is critical to consider early in the differential of young athletes with unilateral arm pain. This case serves as review of the syndrome, but most significantly emphasizes the need for expert consensus and further research in guiding treatment recommendations.

REFERENCES