

http://dx.doi.org/10.1016/j.jemermed.2012.07.075



A CASE REPORT: A YOUNG WAITER WITH PAGET-SCHROETTER SYNDROME

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□ Abstract—Background: Paget-Schroetter syndrome (PSS) is a rare presentation of primary axillary subclavian vein thrombosis that classically occurs in young men with a degree of underlying thoracic outlet syndrome after a period of upper extremity exertion. The primary complication of PSS is post-thrombotic syndrome, a result of chronic venous hypertension. Objectives: To educate Emergency Physicians on this condition to potentiate timely diagnosis and appropriate disposition. Case Report: A 29-year-old right-handed restaurant waiter presented with 3 days of non-painful, gradual-onset right upper extremity swelling with normal vital signs. The patient's history was otherwise notable for subjective fevers and a right forearm abrasion. Upon examination, the right upper extremity was neurovascularly intact and remarkable for uniform edema and ervthema extending distally from the level of the midhumerus. The primary differential diagnoses were deep venous thrombosis (DVT) vs. soft tissue infection. Venous phase contrast computed tomography did not reveal evidence of underlying soft tissue infection and was inconclusive regarding a DVT. Ultrasound demonstrated a right subclavian vein DVT. The patient was admitted and underwent thrombolysis, venolysis, and first rib resection and initiation of warfarin. Conclusion: PSS is a rare presentation of upper-extremity DVT occurring classically in patients without commonly recognized pro-thrombotic risk factors. PSS carries the potential of significant morbidity in the form of post-thrombotic syndrome and pulmonary embolism. Current literature suggests that optimal outcomes are achieved when treatment is initiated within 6 weeks of onset. The treatment paradigm calls for thrombolysis and, frequently, a first rib resection. © 2013 Elsevier Inc.

□ Keywords—Paget-Schroetter syndrome; effort thrombosis; deep venous thrombosis; thrombolysis; rib resection; thoracic outlet syndrome

INTRODUCTION

Paget-Schroetter syndrome (PSS) is a rare presentation of primary axillary-subclavian vein thrombosis first described independently by Paget in 1875 and von Schroetter in 1884 (1,2). In 1948, Hughes coined the name "Paget-Schroetter syndrome" (3,4). If not recognized and treated within an appropriate timeframe, PSS carries significant morbidity, primarily in the form of post-thrombotic syndrome or pulmonary embolism. It is important that Emergency Physicians be aware of PSS to avoid a missed diagnosis and a potential poor outcome.

CASE REPORT

A 29-year-old right-handed restaurant waiter presented to our Emergency Department (ED) with a complaint of gradual-onset right upper-extremity redness and swelling for 3 days that extended distally from the level of his midhumerus. The extremity was neurovascularly intact and without pain. He reported subjective fevers and noted that several days before the onset of symptoms he had sustained a superficial scratch on his right forearm from his girlfriend. Otherwise, he denied any other associated symptoms. His past medical, surgical, and family history were all unremarkable. He took no medications or herbal

Received: 24 January 2012; Final submission received: 14 May 2012; Accepted: 30 July 2012

or holistic remedies, and had no known allergies. The patient denied any history of intravenous drug use, but offered that he occasionally used alcohol and marijuana. He was afebrile with normal vital signs, both on presentation and throughout his ED course. On physical examination, he was well developed and in no distress. Cardiac, pulmonary, abdominal, neurologic, lowerextremity, and genitourinary examinations were all unremarkable.

Examination of the patient's upper extremities revealed that his right side was significantly swollen as compared with the left. Extending from the midhumerus distally, the right upper extremity was uniformly edematous and erythematous without pitting. At the level of the mid-humerus, a circumferential band of superficial dilatated veins was appreciated. He had a well-healed superficial 5-cm abrasion on his right forearm. No axillary or cervical lymphadenopathy was noted. Both radial and ulnar pulses were strong, and sensation and motor function were intact in all distributions. The extremity was non-tender, and no crepitus or cords were appreciated.

At this juncture, the differential diagnosis included soft tissue infection, or allergic reaction, and venous or lymphatic obstructive process, likely secondary to deep venous thrombosis (DVT). Given the lack of pruritis, the time course, and no history of precipitating exposures, an allergic reaction was felt to be unlikely. An infectious etiology was strongly considered, in part due to the history of minor skin trauma and subjective fevers. A DVT was felt to be likely, given the presentation, however, our patient was initially not recognized to have any risk factors for venothromboembolic disease.

Routine blood work and plain radiographs of the chest and right upper extremity were performed. Laboratory analysis was significant only for a white blood cell count of 17,000 per cubic millimeter (94% neutrophils). Right upper-extremity radiographs were negative for any evidence of gas in underlying tissues or fracture. Chest radiograph did not demonstrate any evidence of compressive or obstructive masses in the chest.

Treatment for possible soft tissue infection was initiated with intravenous ceftriaxone and clindamycin, pending evaluation for DVT. Because it would be several hours before an ultrasound could be performed at our institution, and due to the potential for soft tissue infection involving deep spaces, the decision was made to perform a right upper-extremity computed tomography (CT) scan with venous phase contrast. The CT scan revealed soft tissue edema, but otherwise did not show any evidence of soft tissue infection. Furthermore, it was inconclusive regarding evidence of DVT. At this point, the patient was sent for ultrasound examination, which revealed acute deep venous thrombosis in the right subclavian vein extending to the right brachial and basilic veins (Figures 1, 2).

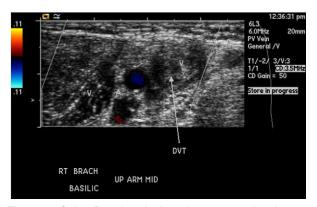


Figure 1. Color Doppler duplex ultrasonography demonstrates right brachial and basilic vein deep venous thrombosis (DVT).

The patient was then admitted to the vascular surgery service, where he underwent three treatments of intravenous tissue plasminogen activator on 3 consecutive days in conjunction with thrombectomy to remove the majority of the clot burden. Venography demonstrated "persistent stenosis with clot at the subclavian/SVC [superior vena cava] junction." The interventional radiology report stated, "This finding is consistent with Paget-Schroetter syndrome." Subsequently, the patient underwent right first rib resection, venolysis, and takedown of the right subclavian muscle. The post-operative course was unremarkable and the patient was bridged to warfarin therapy and discharged from the hospital. A hypercoagulability work-up was negative.

DISCUSSION

PSS, also known as "effort thrombosis," is a rare manifestation of primary upper-extremity deep venous thrombosis, with an incidence of approximately one to two per 100,000 persons per year (5–8). PSS is characterized by upper-extremity DVT resulting from repetitive trauma

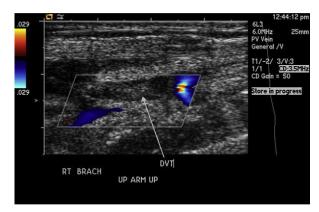


Figure 2. Color Doppler duplex ultrasonography demonstrates right brachial vein deep venous thrombosis (DVT).

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