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EFFORT THROMBOSIS PROVOKED BY SAXOPHONE PERFORMANCE

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☐ **Abstract—Background: Internal jugular venous thrombosis (IJVT) is an uncommon condition rarely diagnosed in the outpatient setting. IJVT carries significant morbidity and mortality and must be considered in the differential diagnosis for new-onset neck pain and swelling, especially in the emergency setting. Paget-Schroetter syndrome (PSS), or primary thrombosis secondary to effort, is an uncommon, likely under-recognized etiology of thrombosis. We report a case of PSS extending from the right subclavian vein into the right internal jugular vein, suspected based upon patient history and physical examination and confirmed by point-of-care ultrasound (POCUS). We then review the presentation, causes, and diagnostic standards for PSS. Case Report: We present a case of a 79-year-old man who presented to the Emergency Department with acute right-sided neck pain 1 day after playing the saxophone for 4 h the prior evening. POCUS confirmed Paget-Schroetter syndrome, or primary effort thrombosis of the internal jugular vein. Why Should an Emergency Physician Be Aware of This?: To our knowledge, this is the first documented case of PSS resulting from venous stasis with prolonged Valsalva maneuver and vascular trauma with activity of playing the saxophone. The significance of this case is the unusual etiology of a rare presentation and the ability to diagnose this condition quickly and accurately with POCUS.** © **2018 Elsevier Inc. All rights reserved.**

☐ **Keywords—point-of-care ultrasound; Paget-Schroetter; effort thrombosis; internal jugular vein; saxophone**

INTRODUCTION

Paget-Schroetter syndrome (PSS), or "effort thrombosis," is defined as primary spontaneous thrombosis of the subclavian vein at the costoclavicular junction (1). Though PSS is a rare condition with an extrapolated incidence of 2.03 per 100,000 per year, it is becoming increasingly common (2,3). PSS comprises approximately 10% of total cases of deep vein thrombosis, which may reflect underdiagnosis (3). It is most commonly seen in young, healthy males (1). Secondary upper-extremity deep vein thrombosis (UEDVT) is more common, and associated with malignancy, catheterization, surgery or trauma to the arm or shoulder, pregnancy, and hormone-induced thrombophilic states (3).

PSS is characterized as a venous variant of thoracic outlet syndrome. Currently, the condition is believed to result from mechanical damage to the vein caused by stretching during the strain of the muscles surrounding the thoracic outlet. Narrowing of the thoracic outlet may be anatomical or physiological. Normal anatomic structure allows compression of the subclavian vein within the costoclavicular space during arm abduction (4). However, in the setting of repetitive arm motion, prolonged hyperabduction, or external rotation, it is thought that chronic compression and endothelial trauma results in inflammation, intimal hypertrophy, and collagen scarring in the musculature surrounding the blood vessels (5).

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Activities associated with PSS include baseball, basketball, swimming, weight lifting, combat training, wrestling, and tennis (6,7). Intermittent compression, partial thrombosis, and recanalization may also contribute to this process, which results in development of collateral vasculature (4,8). Inherited, neoplastic and iatrogenic thrombophilia, associated with central venous catheterization, for instance, may complete Virchow's triad in the pathogenesis of this condition (9).

In 60–80% of PSS cases, patients report having participated in recent vigorous exercise, and in up to 85% of such cases, report symptoms within 24 h of those activities. Patients classically present with an acutely swollen, heavy, painful arm with red-blue discoloration. When occlusion is chronic, venous congestion at collateral vessels may be evident (1).

Duplex ultrasound is the gold standard for diagnosis of PSS, as demonstrated by incompressibility and absence of flow. Sensitivity for identifying UEDVT has been shown to be 97% (90–100% with 95% confidence interval) for compression ultrasonography, 84% (72–97%) for Doppler ultrasonography, 91% (85–97%) for Doppler ultrasonography with compression (10). Contrast venography is the gold standard for diagnosis, but is not as accessible and convenient as duplex ultrasound. Venography and use of such modalities as magnetic resonance and computed tomography is recommended if noninvasive ultrasound is inconclusive, or in anticipation of intervention (1,11).

Treatment strategies include anticoagulation, thrombolysis, mechanical catheter interventions such as thrombectomy, and surgery, though none has emerged as the standard of care (3). As UEDVT is more likely to be associated with malignancy than lower extremity deep vein thrombosis (LEDVT), age-appropriate cancer screening completes the work-up for PSS, as well as screening for hereditary clotting deficiencies (12,13). Without treatment, thrombus progression, recurrence, pulmonary embolism, and postthrombotic syndrome may occur (3). Elderly patients with central venous catheters or pacemakers have the greatest rate of mortality from UEDVT (14).

CASE REPORT

An elderly 79-year-old man with past medical history of middle cerebral artery stroke with residual aphasia, myocardial infarction, and hypertension presented to the Emergency Department (ED) with acute mild right neck pain. The evening prior, the patient had played the saxophone for several hours after a several-year hiatus from regular practice and performance. Later that evening, the patient's wife noted bilateral neck, face, and hand swelling, worse on the right, which gradually worsened over 24 h. At presentation, the patient denied neck stiffness, decreased range of motion, or positional change in pain. The patient also denied fevers, trauma, change in oral secretions, stridor, chest pain, shortness of breath, drug or alcohol use, and smoking. The patient had no personal or family history of blood clots. The patient was compliant with his medication regimen of aspirin, atorvastatin, donepezil, levocetirizine, lisinopril, nifedipine, tamsulosin, and riboflavin.

The patient had a heart rate of 56 beats/min, oral temperature of 36.8°C, respiratory rate of 17 breaths/min,

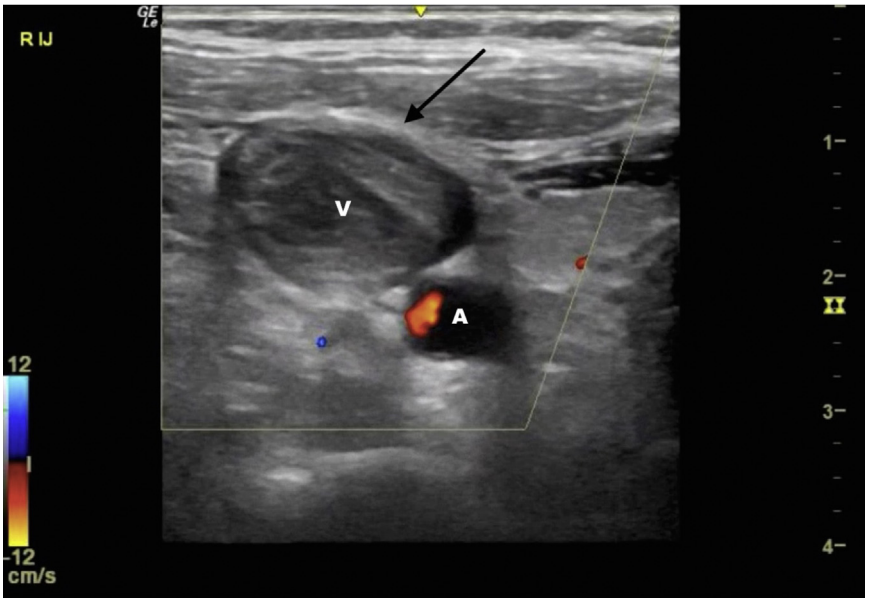

Figure 1. Hyperechoic thrombus visualized in the right internal jugular vein (black arrow). Color Doppler demonstrates flow in right carotid artery (A) and lack of flow in the right internal jugular vein (V).

oxygen saturation of 98% on room air, and was noted to be hypertensive to 159/85 mm Hg. On examination, the patient was in no acute distress. His posterior pharynx had no exudate or asymmetry. Warmth and fullness was noted in the neck bilaterally without defined masses or crepitus. Mild swelling of the face, right upper extremity, and right hand was noted without any erythema or discoloration. Pulmonary and cardiac examination were within normal limits. Venous stasis and peripheral edema were present in bilateral lower extremities without calf tenderness. His neurological examination was unremarkable.

Head and neck x-ray studies were unremarkable. Cardiac troponin-I was elevated but similar to prior studies. Coagulation testing results were within normal limits. His initial electrocardiogram showed inverted T-waves in V3, V4, V5, V6, II, III, aVF, aVL and I, which was unchanged from prior.

Bedside duplex point-of-care ultrasound using a linear probe was conducted by the emergency physician, which demonstrated noncompressibility and absence of flow of the right internal jugular vein and subclavian vein (Figure 1). Follow-up vascular laboratory ultrasound confirmed acute deep venous thrombosis in the internal jugular and subclavian veins.

The patient was admitted to the hospital, where heparin was begun as he was a poor surgical candidate for thrombectomy. Electrophysiology cardiology consultants did not suspect the patient's dual-chamber pacemaker wires were the nidus of the thrombus as they were on the contralateral side, and elected not to remove the device. The patient was diagnosed with primary spontaneous venous thrombosis of the subclavian and internal jugular veins. He was discharged on apixaban.

DISCUSSION

The saxophone is a metal woodwind instrument used in jazz and classical music. The performer generates sound by exhaling at a controlled speed, which vibrates the reed attached to the mouthpiece. While playing the instrument from a seated or standing position, the performer maintains the Valsalva maneuver for adequate breath support. Notes are played by changing finger positions. The instrument is supported using the right thumb, which is placed behind the instrument with the right arm flexed at a 15° angle with the shoulder slightly abducted and internally rotated. The instrument rests at the right side of the performer. Traditionally, the performer uses a neck strap that loops around the posterior surface of the base of the neck (15).

To our knowledge, this is the first reported presentation of PSS in the setting of a patient playing the saxophone. We suspect the thrombus developed as a result of endothelial microtrauma and venous stasis with decreased venous return from the head and upper extremities as a result of increased intrathoracic pressure required to play the saxophone (16). The Valsalva maneuver is associated with initially increased blood flow toward the head and upper extremities through the internal jugular vein as well as valve incompetence, which increased venous stasis over the prolonged practice session (17). We suspect the intermittent obstruction and positional venous blood stagnation while playing the saxophone and wearing the neck strap was sufficiently thrombogenic to promote clot formation from the right subclavian vein to the right internal jugular vein.

We acknowledge that the presence of the pacemaker may have contributed to the formation of the thrombus, given the absence of musculoskeletal abnormalities, indwelling catheter, or prior surgeries in that area. The patient also lacked other risk factors for PSS such as recent hospitalization and personal or familial history of thrombophilia. However, we recognize that although the patient had no history of cancer at the time of presentation, there exists uncertainty as to whether the patient had yet undiagnosed cancer or another condition contributing to coagulopathy (1,3,9,14).

WHY SHOULD AN EMERGENCY PHYSICIAN BE AWARE OF THIS?

PSS is an uncommon etiology of UEDVT. However, the condition is associated with a broad range of strenuous physical activities, which may include musical performance, such as in this case. The variety of patient histories associated with this condition should give emergency medicine practitioners a low threshold for utilizing the duplex ultrasound to rule out this condition. Practitioners skilled in point-of-care ultrasound may quickly and easily visualize implicated vessels at the bedside to minimize morbidity and mortality.

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