

Case Study

Differential diagnosis of a rare case of upper limb pain: Paget-Schroetter syndrome in a doner kebab chef

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Abstract. [Purpose] Paget-Schroetter syndrome (PSS) is an uncommon deep vein thrombosis of the axillary and subclavian veins which may occur spontaneously, but is usually caused by excessive upper limb activity. PSS is clinically similar to other upper limb musculoskeletal disorders and soft tissue infections, and this may lead to delay in correct diagnosis in its early stages. The aim of our case report is to discuss this rare condition with reference to the available literature. [Subjects and Methods] Here we report the case of a doner kebab chef who complained of swelling and pain in his right arm around the biceps muscle. The initial diagnosis was biceps tendon rupture, for which the patient underwent magnetic resonance imaging (MRI) of the right arm and shoulder. Since the MRI revealed no pathological findings, right upper limb venous Doppler ultrasound analysis was performed. Subacute thrombosis materials were detected in the subclavian, axillary, and brachial veins. [Results] With rapid anticoagulant therapy, the patient's symptoms quickly improved. [Conclusion] Early diagnosis and treatment of PSS is critical for preventing potentially fatal complications such as pulmonary embolism. Prophylaxis is important for preventing recurrent thrombosis and for avoiding the development of post-thrombotic syndrome. PSS should be considered a possible cause of painful swelling of the upper limbs, especially in young, active patients who use their arms excessively.

Key words: Doner kebab chef, Effort thrombosis, Paget-Schroetter syndrome

(This article was submitted Jun. 5, 2015, and was accepted Jul. 13, 2015)

INTRODUCTION

Primary axillary-subclavian vein thrombosis, first described by Paget in 1875 and Schroetter in 1884, was named Paget-Schroetter syndrome (PSS) by Hughes in 1948¹⁾. Upper extremity effort thrombosis accounts for approximately 1–4% of all episodes of venous thrombosis²⁾. PSS most often develops among young adults who work in occupations that require repeated arm movements which cause axillo-subclavian vein trauma and facilitate the development of deep vein thrombosis (DVT)³⁾. Several sporting activities have been associated with axillary vein thrombosis, including ball games, racket games, and aquatic sports. Due to repetitive extension and hyperabduction movements of the upper limb, this kind of thrombosis is known as effort thrombosis⁴⁾. Hematologic, anatomic, and iatrogenic predisposing factors such as thrombophilia, thoracic outlet syndrome (TOS), and major vein catheterisation are typically underlying

conditions^{5–8)}. Clinical features of PSS include sudden onset of pain, swelling, edema, and cyanosis of the upper limb. Collateral veins are evident around the shoulder and chest. The pain increases with exercise and decreases with rest and elevation of the affected extremity⁹⁾.

Our patient with PSS is a doner kebab chef, or doner chef, a person skilled in marinating meat to be used in a doner kebab. His work involved placing it in a cooker and then cutting and serving it in desired portions. Since slicing the doner with a long, heavy knife involves repeated arm movement, men in this occupation often develop upper extremity problems such as lateral/medial epicondylitis and tendinitis¹⁰⁾. Our patient had previously been diagnosed as having a biceps tendon rupture. When he was finally diagnosed as having PSS, the differential diagnosis of various possible upper limb problems was discussed.

SUBJECT AND METHODS

A 40-year-old male working as a doner kebab chef presented at our outpatient clinic with complaints of experiencing pain, swelling, and edema in his right upper limb (RUL) in the prior few days. This right-handed patient did not describe any trauma, but his occupation requires the continuous use of his right arm, particularly with hyperabduction and extension movements. Physical examination disclosed

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Fig. 1. Significant edema, blue discoloration, and dilatation of the superficial veins in the patient's right upper limb

significant edema, blue discoloration, and dilatation of the superficial veins in the RUL (Fig.1). His range of shoulder motions was restricted, and the movements themselves were painful. He had a positive "Popeye's sign" in the biceps tendon. Neurological examination, Adson's, costoclavicular, and hyperabduction tests were all normal. Brachial, radial, and ulnar pulses were palpable. The cervical, right shoulder, and chest radiographies were normal. The patient underwent magnetic resonance imaging (MRI) of the shoulder. Besides an initial diagnosis of biceps tendon rupture, the MRI revealed no pathological findings, so the patient then underwent RUL venous Doppler ultrasound (VDU). The VDU showed a subacute thrombosis of the subclavian, axillary, and brachial veins.

This case study complied with the ethical standards of the Declaration of Helsinki (1975, revised 1983). The ethics committee of our institution approved the publishing of the case details, and the patient provided his written informed consent.

RESULTS

The patient was referred to the departments of orthopedic, hematology, and cardiovascular surgery. He was started on anticoagulant therapy of enoxaparin (0.6 cm³ twice a day). A compression sleeve for his arm, rest, and elevation of the RUL were recommended. Routine laboratory tests included a coagulation profile: antithrombin-3, protein C, protein S, homocysteine, lupus anticoagulant, anticardiolipin immunoglobulin M and G, antinuclear antibody, and mutations for factor 5 Leiden were all normal. After ten days, the treatment was switched to oral anticoagulant therapy. For the next six months, warfarin (5 mg/day) was given as a prophylaxis. The patient's international normalized ratio (INR) was kept stable at 2–3. He had no complaints after the third month of treatment, and the thrombosis disappeared on control VDU evaluation. Six months later, acetylsalicylic acid (300 mg/day) was commenced. The patient was followed up every 3 months in the first year, and VDU was performed at every visit. He resumed his occupation and is currently continuing his vocation as a doner kebab chef.

DISCUSSION

PSS, or 'effort thrombosis' of the axillary-subclavian vein, is a relatively uncommon condition^{1, 4, 8}. It develops

more frequently in the dominant upper limb, usually after sports activities such as long distance swimming, body building exercises, wrestling, gymnastics, handball, and baseball throwing^{1, 3, 4, 8}. A strain on the subclavian and axillary veins by retroversion or hyperabduction of the arm can cause microtraumatizations of the venous intima, consequently leading to local coagulation⁷. Maharaj and his colleagues also reported a case diagnosed as PSS secondary to exotic dancing⁴. In the present case, we thought that our patient's occupation, one that requires continuous use of the right arm with hyperabduction, flexion, and extension movements, facilitated the development of his condition.

The most common symptoms of PSS are painful congestion and edema of the affected arm¹. These symptoms can also be seen in upper limb musculoskeletal disorders and upper limb soft tissue infections which, as in our patient, may lead to delay in correct diagnosis in the early stages^{1, 5, 6}. Similarly, Oktar and Ergul¹ reported a case which was first diagnosed in another medical center as a soft tissue infection (cellulitis) and treated for ten days with cefuroxime axetil, analgesics, and anti-inflammatory drugs. Because of the lack of response to the antibiotic therapy, the patient was re-evaluated and diagnosed as having PSS. Kidd and Broderick¹¹ also reported a case with a history of oral contraceptive pill use who was initially diagnosed with medial epicondylitis, which was later changed to subclavian thrombosis caused by PSS. Trauma, compressive tumor, cervical radiculopathy, and spinal stenosis that can lead to brachial plexopathy and lymphedema are other causes to be considered during differential diagnosis. In our case, the patient had no fever and no obvious site of infection to suggest cellulitis, he had had no previous surgery or previous episodes of swelling to suggest lymphedema, and his radiographic and MRI evaluations were normal.

Thoracic outlet syndrome (TOS) is a major anatomic disorder that predisposes to PSS. The axillary subclavian vein traverses the tunnel formed by the clavicle and subclavius muscle anteriorly, the scalenus anticus muscle laterally, the first rib posterior-inferiorly, and the costoclavicular ligament medially. Abnormalities of any of these structures may anatomically narrow the tunnel. Ozcahar et al. reported the co-existence of PSS and TOS in their two separate case reports^{6, 7}. Cervical radiography of our patient revealed no cervical rib or subclavian stenosis that would suggest TOS. Hematologic or malignant disorders that lead to thrombophilia predisposes to PSS with intimal injury and increasing blood viscosity^{4-8, 11, 12}. We found no such disorders in our patient's hematologic profiles, nor evidence of malignancy. Venous patency can be effectively restored by means of a multimodal approach, including thrombolytic treatment, anticoagulation and vascular techniques and, in selected cases, surgical thoracic outlet decompression. Treatment choices should be individualized, based on the cause of the thrombosis¹.

In conclusion, early diagnosis and treatment of PSS is critical for preventing potentially fatal complications such as pulmonary embolism. Prophylaxis is important for preventing recurrent thrombosis and for avoiding development of post-thrombotic syndrome. PSS should be considered a possible cause of painful swelling of the upper limb, especially

in young, active patients who use their arms excessively.

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