Paget Schroetter Syndrome: A case study of the chiropractor's role in recognizing and comanaging an important condition

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Objective: To emphasize the importance for health care professionals to be knowledgeable of a relatively rare form of thoracic outlet syndrome, known as Paget Schroetter syndrome. The etiology, key signs and symptoms, and the importance of immediate referral are highlighted and an introduction to manual therapists' role within a multidisciplinary team is provided.

Clinical Features: *Healthy athletes aged 15-30* are most commonly affected with 60-80% of patients reporting a history of repetitive or vigorous overhead activity prior to symptom onset.

Intervention and Outcome: *Manual therapists* have a role in recognizing, referring, and providing symptomatic relief with soft tissue therapy, correcting abnormal biomechanics, manipulations, mobilizations, and a rehabilitative program, as seen in this case report. Conclusion: Early recognition and referral of Objectif : Mettre l'accent sur l'importance de la connaissance des professionnels des soins de santé quant à une forme relativement rare du syndrome de la traversée thoracobrachiale connu sous le nom de syndrome de Paget-Schroetter. L'article souligne l'étiologie et les signes et symptômes principaux du syndrome, ainsi que l'importance d'une recommandation sans délai. Il donne également un aperçu du rôle du thérapeute manuel au sein d'une équipe multidisciplinaire.

Caractéristiques cliniques : Les athlètes en bonne santé âgés de 15 à 30 ans sont les plus souvent atteints, et 60 à 80 % des patients rapportent des antécédents de mouvements répétitifs ou vigoureux au-dessus de la tête avant l'apparition des symptômes.

Intervention et résultat : *Les thérapeutes manuels* ont un rôle à jouer pour identifier le syndrome, recommander les patients et fournir un soulagement des symptômes par la thérapie des tissus mous, en corrigeant les anomalies biomécaniques, et au moyen de manipulations, de mobilisations et d'un programme de réadaptation, comme l'indique la présente étude de cas.

Conclusion : L'identification précoce du syndrome de Paget-Schroetter et la recommandation rapide des patients qui en sont atteints sont essentielles à leur

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Disclaimer: Written consent was received from the patient for publication of this case report. No funding was received for this report. The authors have no commercial associations that might pose a conflict of interest in connection with the submitted article. ©JCCA 2012 Paget Schroetter syndrome are essential for optimal recovery. Manual therapists may prove to have a role in decreasing the need for surgical decompression and accelerating resumption of regular activities. (JCCA 2012;56(4):256-261)

KEY WORDS: Paget Schroetter syndrome, deep venous thrombosis, upper extremity thrombosis, thoracic outlet syndrome, chiropractic, effort thrombosis rétablissement optimal. Les thérapeutes manuels peuvent avoir un rôle à jouer afin de réduire le besoin pour des décompressions chirurgicales et d'accélérer la reprise des activités régulières. (JCCA 2012;56(4):256-261)

MOTS CLÉS : syndrome de Paget-Schroetter, thrombose veineuse profonde, thrombose des membres supérieurs, syndrome de la traversée thoracobrachiale, chiropratique, thrombose d'effort

Introduction

Although Paget Schroetter Syndrome (PSS) is often regarded as the most common vascular problem in athletes, it is not well known or recognized by many health care practitioners.¹ PSS, also known as "effort thrombosis", is a form of primary upper extremity deep vein thrombosis (UEDVT) involving the axillary-subclavian vein within the costoclavicular space.² It is hypothesized that repetitive overhead activity causes an intermittent and positional compression on the subclavian vein resulting in recurrent endothelial microtrauma and consecutive activation of the coagulation cascade.^{2,3} Approximately 60-80% of patients diagnosed with PSS report a history of repetitive or vigorous overhead activity.4 The most common population affected by this condition is young, healthy athletes between the age of 15-30 years.⁵ UEDVT accounts for 11% of all cases of deep vein thrombosis and PSS represents 15-24% of those cases, however, like many rare conditions, its incidence is likely under-recognized and under-reported.2,4,5

According to *Perlowski and Jaff*, vascular issues are commonly overlooked in athletes for three main reasons, the first being that most athletes are often young and healthy.⁶ The second reason is that their symptoms may be very similar to, and therefore mistaken for, a musculoskeletal injury. Finally, many health care practitioners may not adequately perform a vascular examination and are unaware of PSS as a differential diagnosis.⁶ Currently there are no prospective randomized controlled trials concerning the conservative management of PSS and most treatment plans are guided by case reports, retrospective reviews and/or expert opinion. The purpose of this case report is to underscore the importance for chiropractors to be aware of this condition, to recognize the signs and symptoms for quick referral and to understand how this profession has a role in co-treating patients for symptomatic relief with the treating physician.

Case Presentation

A forty three year old avid female downhill skier presented to her chiropractor with a history of sudden onset right lower neck and chest pain with numbness in the right hand. She described the pain as sharp and rated it 5-6/10 on a numeric pain rating scale (zero indicating "no pain" and 10 equaling the "worst pain ever"). The discomfort began after initiating a new upper body strengthening regimen one week earlier with her personal trainer. She immediately went to her local emergency department where she was assessed and a diagnostic ultrasound (US) of her neck region was performed. The patient was subsequently diagnosed with Thoracic Outlet Syndrome (TOS). The patient had previously seen the chiropractor for various sports injuries, cervicothoracic postural strains and headaches without any complaint of numbness or tingling to the upper extremities. The remainder of her health history was unremarkable.

Upon physical examination, cervical active and passive ranges of motion (ROM) were full with stiffness on the right side during left lateral flexion and rotation. Resisted cervical ROM was pain free and graded 5/5. Cervical Jackson's, Spurling's and maximal foramina compression tests were negative for any radicular pain. Cervical Kemp's test on the right caused pain at C7-T1 and along the right first and second costovertebral joints. Right Adson's test reproduced the numbness in her right hand and her radial pulse was significantly decreased. Right Doorbell and Elevated Arm Stress Test (EAST) also reproduced the numbness in her right hand. Palpation of the right pectoralis minor, anterior scalene, upper trapezius, and levator scapulae muscles revealed tenderness and hypertonicity. The right digits were slightly colder than the left, however, the nail bed perfusion was the same bilaterally. Her vital signs were within normal limits and no swelling or colour changes were noted. Neurological testing of the upper limb was within normal limits. Motion palpation revealed restricted and tender right C7-T1, T3-5 and right first and second costovertebral joints.

The patient was diagnosed with suspected right vascular TOS. She was referred for cervical radiographs to determine the presence of cervical ribs. The radiographs showed mild uncovertebral joint degeneration bilaterally at C6 and on the right at C4, mild degenerative disc disease was present at C5-6 and mild facet joint degeneration from C3-6. There was no evidence of cervical ribs. The patient was encouraged to see her family physician and/or the vascular surgeon who assessed her at the hospital for further consultation and to obtain the result of the diagnostic US. It was also suggested at this time that a Dopper US might be indicated to assess the flow of the arterial and venous systems in the upper extremity.

The patient was treated using acupuncture, myofascial release and spinal manipulation therapy of the thoracic spine 2-3 times per week for three weeks. She reported a reduction in pain and decreased frequency of tingling in the right hand at the end of the three week period. At this point she stated that she would be unable to continue therapy due to personal reasons. Two months later, the patient reported that she had seen a vascular surgeon who sent her for a Doppler US. The results of the US were as follows: "Non-occlusive clot seen in right subclavian vein at the proximal mid and distal portions. Right axillary, cephalic, basilic and brachial veins are patent but very slow flow seen without any clot". The surgeon diagnosed her with effort thrombosis and started her on anticoagulant therapy. Surgical decompression involving a partial first rib resection was booked for later in the week.

Three weeks after the transaxillary right partial first rib resection, the surgeon informed her that the healing was progressing adequately and recommended that she start physical therapy. The patient subsequently attended the office of the same chiropractor with complaints of throbbing in the right chest, arm and forearm, aggravated by working on the computer and during activities involving the right arm. She also reported stiffness in the cervical region, rated 5/10 on the VAS, with intermittent headaches. Upon physical examination, active and passive ROM of the right shoulder were full but revealed tightness of the pectoralis musculature during shoulder abduction, horizontal abduction, and flexion. Palpation revealed tenderness and hypertonicity of the right anterior scalene, upper trapezius, levator scapulae, rhomboids, infraspinatus, subscapularis, pectoralis minor, corocobrachialis, pronator teres and flexor carpi ulnaris muscles. Palpation of the right suboccipital musculature reproduced her headache symptoms. The surgical scar over the right axillary region appeared to be healing adequately. Motion palpation revealed restriction and tenderness at C0-1 on the right, C2-3 on the left, T3-4, and over the third and fourth right costovertebral joints. Following the examination, the patient was diagnosed with myofascial pain secondary to the right first rib resection, as well as, cervicothoracic joint dysfunction and tension headaches. She was treated using acupuncture, myofascial release, spinal manipulation therapy, and was given a stretching regimen. She was treated twice a week for five weeks and reported significant reductions in neck, chest and arm pain with no headaches after this time period. One year later the patient was contacted over the phone and has been continuing with her activities with minimal discomfort but avoiding strenuous upper body workouts.

Discussion

When a patient presents with swelling and discomfort without known trauma it is important to consider vascular conditions in the list of differentials (provided in Table 1).^{6,7} PSS often presents with aching, swelling, fa-

Table 1
Differential diagnoses for Paget Schroetter syndrome

DIFFERENTIAL DIAGNOSES FOR PSS		
Pain	Swelling	
 Neurogenic TOS Cervical radiculopathy Brachial plexopathy Rotator cuff tendinopathy Subacromial impingement Scapular dysfunction Biceps tendinopathy Labral tear Instability Quadrilateral space syndrome Long thoracic nerve palsy Suprascapular nerve palsy Peripheral nerve 	 Arterial TOS Compartment syndrome Lymphedema Esophageal or cardiac disease Complex regional pain syndrome Intramuscular hemorrhage Inflammatory disorder Cellulitis Breast cancer Neoplastic compression of mediastinal blood 	
• Suprascapular nerve palsy	• Neoplastic compression of	

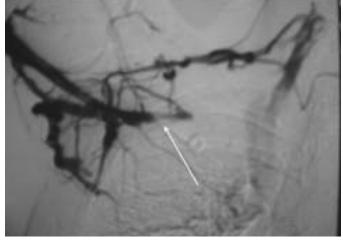


Figure 1: Contrast venogram of the axillary and subclavian vein in the axillary region. A complete occlusion of axillosubclavian vein and significant venous collateralization are depicted in this figure¹

Treat AD, Smith PA, Wen DY, Kinderknecht JJ. Deep vein thrombosis of the subclavian vein in a college volleyball player. Am J Sports Med. 2004;32(2). page 529, copyright ©2004 Sage Publications. Reprinted by permission of Sage Publications.

tigue, heaviness or 'dead arm' sensation and occasionally numbness of the hand or fingers.¹ A history of repetitive or strenuous overhead activity, such as pitching or painting, is commonly reported prior to the onset of symptoms. In this case, the patient started a new workout regimen focusing on the upper extremity prior to the development of her symptoms. Key findings to look for during the physical examination include swelling, erythema, cyanosis, and prominence of the superficial veins.^{8,9} The upper extremity pulses are frequently normal and symmetric during rest, however, thoracic outlet syndrome (TOS) tests may alter the quality of the radial arterial pulse by compressing the neurovascular bundle within the thoracic outlet.² Adson, Wright, EAST, and Eden tests should be used as part of a more complete physical examination and positive results should be considered more significant if the opposite arm is negative.^{10,11} However, these classic TOS provocation tests have been reported to be unreliable and positive for pulse obliteration in up to 90% healthy patients.¹¹ Unfortunately, no study to date has analyzed the specificity, sensitivity and predictability of these tests in relation to the separate categories of TOS.¹¹ In the opinion of the authors, the arm should be placed in many positions, including hyperabduction and external rotation in an attempt to elicit the chief complaint, to recreate distal ischemic signs, edema, or cyanosis of the upper extremity.

Duplex US is commonly reported as the first line of imaging to be performed when there is a suspicion of PSS.¹² This test is non-invasive, low cost, and demonstrates a high sensitivity and specificity for peripheral UEDVT.¹³ It is important to note, however, that one study reported a false negative rate as high as 30%.⁶ Hence, even if the duplex US is negative but the treating practitioner has a high index of suspicion, a venogram should be performed. Contrast venogram is considered the gold standard for diagnosing PSS and provides excellent visualization of the venous anatomy, as illustrated in Figure 1.² It is, however, more invasive and may lead to complications due to the contrast agent.²

Routine studies including a complete blood count, clotting tests, urinalysis and chest radiographs should be performed to analyze possible causes for the DVT.² Current guidelines from the American College of Chest Physicians (ACCP) recommend initial treatment with low molecular weight heparin or unfractionated heparin.¹⁴ This is typ-

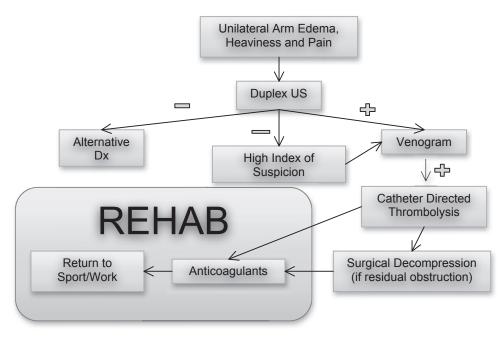


Figure 2: A possible management strategy for Paget Schroetter syndrome from recognition of the signs and symptoms to return to regular activities

ically followed by warfarin or vitamin K antagonists for at least three months.^{2,14} Catheter-directed thrombolysis is also commonly done to restore venous patency, minimize intimal damage and decrease the risk of long term complications.13 This treatment works best when administered within several weeks of onset of symptoms and the successful recanalization approaches 100% when initiated within the first few days.⁴ Currently, there is a great deal of debate about thoracic outlet decompression surgery in the literature; some authors believe that it is an integral part of the management and should be done early whereas others believe a trial of conservative therapy should be done and surgery should be reserved only for those with persistent symptoms.3 ACCP guidelines suggest that surgical decompression may be considered in patients with severe and persistent symptoms who have failed to benefit from medical treatment, as it was in the case presented.¹⁴

Chiropractors and other manual therapists may have a significant role to play in not only recognizing the condition but also administering a trial of conservative therapy in the hopes of eliminating their symptoms and decreasing the need for surgical decompression. Current literature outlines the importance of proper rehabilitation

to return athletes to competition, to allow individuals to go back to work, and to improve quality of life in general.^{9,15,16,17} There are no studies to state which therapy is most beneficial for the treatment of PSS. This stresses the importance of having a strong understanding of the anatomy and pathophysiology of this condition when developing a treatment plan. In the opinion of the authors, soft tissue therapy of the musculature surrounding the costoclavicular space as well as the shoulder girdle may help to decrease the compression of the subclavian vein. Postural education and correction of abnormal shoulder biomechanics through scapular stabilization, manipulation or mobilization of the thoracic spine, costovertebral, sternocalvicular or acromioclavicular joints may also help to prevent the recurrence or persistent symptoms of PSS. General cardiovascular conditioning and strengthening as well as core stability should also be emphasized. If working with athletic populations, sport-specific training is essential before safe return to play. A schematic diagram (Figure 2) is included to summarize the authors' depiction of optimal management for a patient with PSS from recognition of the symptoms to return to regular activities.

Although rare for PSS with an incidence of 5.6%, pul-

monary embolism is a serious complication related to this condition that may result in a fatal outcome, reinforcing the importance of a proper diagnosis.^{8,15} Key signs and symptoms of a pulmonary embolism include dyspnea, chest pain, syncope, low grade fever, and tachycardia.² More commonly, patients may have chronic complications of PSS called post-thrombotic syndrome.² This syndrome can range from mild symptoms of discomfort to significant functional disability and impaired quality of life. Dependent on the specific criteria applied for diagnosing post-thrombotic syndrome the incidence ranges from 7-46%.² Common symptoms of post-thrombotic syndrome include edema, persistent pain in the affected arm, limb fatigue with exertion, distended collateral veins and skin discoloration.² It is also important to be aware that there is a two year cumulative incidence of recurrent venous thromboembolism between 4-8%.²

Conclusions

It is critical that health care practitioners are aware of this rare but possibly fatal condition that presents in young, healthy adults. Early recognition and early treatment are key factors for optimizing recovery. Chiropractors and other manual therapists can have an important role in recognizing the signs and symptoms of PSS and providing an early referral for appropriate medical treatment. Once treatment is initiated, the manual therapist can work in conjunction with the treating physician to co-manage the patient. Correcting abnormal biomechanics and treating the surrounding musculature may prove to be beneficial for decreasing the need for surgical decompression and accelerate the resumption of regular activities.

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