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Upper extremity deep vein thrombosis presenting to a chiropractic clinic: a description of 2 cases

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Abstract

Objective: The purpose of this case series is to describe the presentation of 2 patients who presented to a chiropractic teaching clinic with Paget-Schroetter syndrome (PSS) and to discuss the potential role for conservative therapy in the management of symptoms.

Clinical Features: Two patients presented with a vascular and muscular findings suggesting activity-related upper extremity deep vein thrombosis. One patient presented with recent onset of symptoms (pain in the neck with a "pinched nerve sensation" in the left upper trapezius); and the other presented with chronic, low-grade neck pain of 1 year's duration.

Intervention and Outcome: The initial treatment approach for the patient with acute symptoms included soft tissue therapy. During the second appointment, he was immediately referred for medical evaluation and management because of worsening symptoms. He was diagnosed with thrombus in the left brachial vein, started immediately on a thrombolytic agent, and referred to a thrombosis clinic. Treatment for the second patient with chronic symptoms included soft tissue therapy, spinal manipulative therapy, and active care. Two months after 3 treatments, she reported improved symptoms. She remains under supportive care and has reported continued relief of her symptoms.

Conclusion: Although a rare condition, PSS has the potential to result in significant morbidity and potentially fatal complications; thus, it is critical that practitioners recognize the signs and symptoms to facilitate appropriate and timely referrals. Clinicians should be aware of the presentation and proposed pathogenesis of PSS, and consider this diagnosis in patients with unilateral upper limb and/or neck pain.

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Introduction

Upper extremity deep vein thromboses (UEDVTs) are rare and represent 1% to 4% of all venous thromboses.[1-3] Upper extremity deep vein thromboses are categorized as primary or secondary, and relate to UEDVT affecting the subclavian (18%-69%), axillary (5%-42%), or brachial (4%-13%) veins.[1-7] Approximately one-third of UEDVT s are primary in nature, which include idiopathic thromboses or effort-induced thromboses.[2,4]

Effort or activity-related UEDVT are sometimes referred to as *Paget-Schroetter syndrome* (PSS).[4] The term *PSS* was first used by Hughes in 1949 in his review of 320 cases of subclavian vein thrombosis.[5,8] Sir James Paget identified the first case in 1875 in a patient with acute arm pain and swelling.[5] Independently, van Schroetter reported a similar case in 1884 in a healthy individual after painting.[5] It appeared to the physicians that vascular damage could occur in otherwise healthy individuals following repetitive upper limb stretching.[5]

Paget-Schroetter syndrome has been classified as a disorder with many etiologies[1]; however, it is most commonly induced via activity or physical effort[6] in young and otherwise healthy individuals. Therefore, it most commonly affects the dominant arm.[1,6,7] The purpose of this report of 2 cases is to describe the presentation of PSS to a chiropractic clinic, highlight the importance of prompt referral and imaging, and discuss the potential role for conservative therapy in the management of patients who may experience ongoing symptoms.

Case reports

Case 1

A 44-year-old male bass guitar player and music teacher presented to a chiropractic teaching clinic with left anterior neck pain of 6 days' duration. The complaint originally presented as chest tension while driving and caused the patient to present to his medical doctor. A chest radiograph did not reveal abnormality or pathology, and the patient was diagnosed with gastroesophageal reflux disease and prescribed medication by his medical physician. For the following week, the pain ascended to a location lateral to the esophagus on the left side with associated pain in the left upper trapezius. The intensity of the pain was rated as 7 of 10 on a numeric pain rating scale and was experienced constantly, though worse in the morning. The patient reported that the pain was a "tense" and highly sensitive in the neck, with a "pinched nerve sensation" in the left upper trapezius. The pain was aggravated by lifting, lying prone or supine, swallowing food, deep breathing, and palpation of the neck (specifically lateral to the esophagus), and relieved by self-prescribed stretching and ibuprofen. The patient did not report associated symptoms such as associated cough, fever, or myalgia, and did not report any remarkable findings in his personal or family history. The patient reported being a nonsmoker, consuming approximately 2 alcoholic beverages per week, consuming a healthy diet, and engaging in physical activity 3 to 4 times per week. On initial presentation, Neck Disability Index was scored as 18 of 50, which indicated moderate disability.

On physical examination, the patient showed anterior head carriage and rounded shoulders. His height was reported as 6′3″ and weight as 95.4 kg. Blood pressure was 135/83 mm Hg, pulse rate was 81 beats per minute, and respiratory rate was 15 breaths per minute. Cervical range of motion revealed decreased active and passive extension by 50%, active and passive right lateral flexion by 25%, and right rotation by 10%, all due to pain. Cervical resisted left lateral flexion was graded 4 of 5 and recreated the chief concern. Shoulder range of motion was full and pain-free. Palpation of the left sternocleidomastoid and the region between the thyroid and cricoid cartilage recreated the chief concern; palpation of the cervical spine and cervicothoracic musculature was otherwise unremarkable. Cervical and shoulder orthopedic test results were unremarkable and did not recreate the chief concern. Neurological examination of the upper limb revealed normal sensation (crude touch) in all dermatomes bilaterally; 5 of 5 strength in all myotomes bilaterally; and 2+ C5, C6, and C7 reflexes bilaterally.

At this time, the patient was diagnosed with left cervical muscle hypertonicity associated with gastroesophageal reflux disease; and the plan of management included soft tissue therapy twice a week for 3 weeks. Five days later, the patient returned for a follow-up visit with considerable swelling in the left anterior neck, accompanied by marked vascularity in the left anterior chest and arm and a history of night sweats. He was immediately referred for radiographic imaging; however, the images did not explain the recent swelling in the left supraclavicular space. The patient was referred to the emergency department for further evaluation.

An initial diagnostic ultrasonography did not reveal abnormalities, and the patient was referred for duplex ultrasonography. This imaging study revealed a large

thrombus in the left brachial vein (in the location of his bass guitar strap), and he was started immediately on a thrombolytic agent and referred to a thrombosis clinic. He was treated initially with a daily Lovenox injection (sanofi-aventis Canada Inc., Laval, Quebec, Canada) and 5 mg of warfarin, and his prothrombin time was measured twice weekly to monitor his status. At that time, the diameter of his left forearm and arm was measured at 3.8 cm larger than his right. Following 2 weeks of treatment, the night sweats resolved; and following 6 months of this treatment, duplex ultrasonography revealed that the thrombus had dissolved. However, because of venous scarring, very little blood was able to flow through the left brachial vein. The left arm and forearm remained 1.9 cm larger than the right because of neovascularization.

During his treatment, the patient also underwent genetic testing, which was positive for prothrombin variant (heterozygous). Because of the potential for future incidents given the venous scarring and his genetic variant, the patient elected to continue with warfarin therapy (7 mg/d).

The patient was contacted for follow-up 1 year after presentation and reported no ongoing medical or vascular problems. He had 1 incident of left hand swelling 6 months prior. However, swelling was reduced by the end of the day; and plain film and Doppler ultrasonographic imaging revealed no abnormalities. His international normalized ratio has remained stable at monthly monitoring. He reported that he is less able to do overhead work, is no longer able to lift weights greater than 23 kg, avoids high-impact exercise activity, and feels mildly fatigued; however, he has adapted well to these changes and reports no difficulties coping with his present situation.

Case 2

A 36-year-old female film producer presented to a chiropractic teaching clinic with a history of low-grade right neck, shoulder, and arm pain. The complaint began 1 year prior, after waking up 1 morning with an acute episode of right upper limb swelling and cyanosis. At that time, the patient presented to an emergency department. No chest pain, shortness of breath, or constitutional symptoms were reported; distal pulses and motor strength were normal. She underwent Doppler ultrasonographic examination of her right arm, which revealed an occlusive clot within the right proximal subclavian vein, extending into the cephalic and axillary veins. Sluggish blood flow was observed within the cephalic vein. Results of all other imaging and laboratory studies were normal, and the patient was diagnosed with an UEDVT. She was discharged from the hospital with instructions to visit her family physician to monitor her international normalized ratio while taking warfarin (5 mg orally) during the next 3 months.

A follow-up Doppler study performed 2 months later revealed a small residual but nonocclusive eccentric thrombus within the subclavian vein. At that time, all other veins appeared patent. A final follow-up Doppler study 3 months later revealed no evidence of thrombus in the vascular system of the right upper limb.

Seven months following her initial presentation to the emergency department, the patient underwent a magnetic resonance imaging study to determine the cause of her previous complaint. Imaging of the costoclavicular space with arm abduction revealed extrinsic narrowing of this space bilaterally, with significant resultant compression of the subclavian vein on the right. It was hypothesized that the extrinsic compression was consistent with thoracic outlet syndrome (TOS) and may have contributed to the patient's previously diagnosed condition of PSS.

One year after the initial episode, the patient presented to the chiropractic teaching clinic because of residual local right neck, shoulder, and arm pain that she attributed to PSS and residual TOS. The pain was described as a constant dull ache that varied in intensity depending on activity but averaged 5 of 10 on the numeric pain scale. Prolonged seated postures such as driving and desk and computer work aggravated her symptoms. Her Neck Disability Index was scored as 23 of 50, which indicated moderate disability. She reported relief with deep massage soft tissue therapy, Active Release Techniques, stretching, Tai Chi, Pilates, and yoga.

The patient was 5′4″ tall, 61 kg, and right hand dominant. Mild rubor was observed in the right arm, with no other abnormal findings noted. Vital signs, vascular test results, and neurological examination result of the upper limb were all within normal limits. No range of motion or orthopedic tests (including tests for TOS) aggravated the patient's symptoms; however, palpation revealed tight and tender pectoralis minor, scalenes, and subclavius muscles on the right. Mild joint restrictions were noted in the cervical-thoracic junction and in the midthoracic and lumbar spinal regions.

The patient was diagnosed with postural dysfunction and associated myofascial pain, in addition to the reported resolved PSS. The management plan consisted of soft tissue therapy to affected muscles, spinal manipulation to the thoracic and lumbar spinal areas, postural improvement exercises, and ergonomic sitting advice.

The patient was treated a total of 3 times under the above-stated plan of management. Two months after treatment, she reported that she had found that the treatments helped her symptoms. She remains under supportive care and has reported continued relief of her symptoms.

Discussion

Paget-Schroetter syndrome has been previously reported in the medical and athletic therapy literature; however, to our knowledge, this is the first case report to describe patient presentations to a chiropractic clinic. Furthermore, the first case demonstrated PSS in an atypical patient; he is active, but nonathletic, and may have developed vascular compression through shoulder depression from his guitar strap rather than repeated upper limb movements. The second case demonstrated previously unreported soft tissue sequelae following PSS, which may prompt a patient to present a chiropractic clinic. The first case required identification of a vascular emergency and appropriate referral, whereas the second demonstrated possible long-term sequelae that responded to chiropractic management. Both cases highlight the role a doctor of chiropractic may play in the co-management of this syndrome.

Paget-Schroetter syndrome is a rare condition, affecting 2 in 100 000 persons per year.[4] It occurs predominantly in young patients and affects males more commonly than females.[9,10] It commonly follows repetitive arm motion or extreme exertion; therefore, the dominant limb is most commonly affected.[11]

Anatomy

The upper limb is drained by the basilic and cephalic venous systems.[6] The basilic vein drains the tissues of the upper limb into the axillary vein, which is joined by the cephalic to become the subclavian vein.[6]

The subclavian vein crosses the first rib through a tunnel formed by the clavicle and subclavius anteriorly, the anterior scalene laterally, the first rib posteroinferiorly, and the costoclavicular ligament medially.[6,10] Repetitive mechanical compression is a common mechanism of injury to the vein, as it is frequently compressed while traversing this tunnel.[10,12]

Pathophysiology

Arm abduction, shoulder depression, and extension of the cervical spine may further compress this space and stretch the subclavian vein, which can stimulate the coagulation cascade and predispose the patient to developing symptoms.[1,2,9,13-15] Vascular compression results in repetitive microtrauma to the endothelium of the vessel wall and may cause intimal damage and fibrous tissue formation and facilitate thrombus formation.[1,6,7]

In addition, in cases of PSS, upper limb movements during exercise are believed to contribute to momentary stasis and hypercoagulation.[1] Exercise creates a hypercoagulable state due to transient elevations of factor VIII and von Willebrand factor (coagulation components), which may further predispose a patient to thrombus formation.[6] Athletes and active patients with underlying clotting abnormalities may be predisposed to the development of PSS.[13,16]

Genetic predisposition

Inherited thrombophilia is defined by Franchini et al[17] as "a genetically determined predisposition to the development of thromboembolic complications" and implies that a patient has an increased risk of developing a clot within the blood vessels. In these patients, the clotting process continues unabated and may develop a DVT, pulmonary embolism, myocardial infarction, stroke, recurrent pregnancy loss, or stillbirth.[18]

Inherited disorders can affect any level of the clotting process and can be broken down into 5 main categories, as proposed by Franchini et al.[17] The first category involves the decreased quantity or quality of the processes involved in inhibiting clotting. The second involves an increased quantity or quality of factors that promote clotting. The third group involves hyperhomocysteinemia, the fourth involves defects in the fibrinolytic system, and the fifth involves the abnormal functioning of the platelets (Table 1).

The patient in case 1 was diagnosed with a prothrombin variation or gene mutation. This type of gene mutation was discovered in 1996, and studies have shown that it affects 2% to 5% of the general population and up to 7% to 18% of those with spontaneous DVT.[17,22] Prothrombin gene mutation falls into the second category of inherited disorders, as it promotes thrombosis formation and there is an increased amount of functionally normal prothrombin available.[17] It has been suggested that the presence of the mutation is not adequate to alone cause a DVT; however, it can occur when coupled with another or environmental risk factor.[17,23] The patient in case 1 was a bass player who placed the strap of his guitar on his left side in the area of the DVT, and this relative

Table 1 Types of inherited thrombophilia and their prevalence[17,19-21]

Proposed category system	Type of syndrome	% of general population	% of those with thrombosis
1. Qualitative and quantitative defects of those factors inhibiting clotting	Antithrombin deficiency	0.2	1-3
	Protein C deficiency	0.2-0.4	3-5
	Protein S deficiency	0.03-0.1	1-5
	Heparin cofactor II deficiency	~1	~1
	Tissue factor pathway inhibitor deficiency	[a]	[a]
	Thrombomodulin deficiency	[a]	[a]
2. Qualitative and quantitative defects of those factors promoting clotting	Activated protein C resistance and factor V Leiden	5	10-50
	Prothrombin gene mutation	2-5	6-18
	Dysfibrinogenemia and hyperfibrinogenemia	<1	<1
	Elevated levels of clotting factors VIII	11	25
	Elevated levels of clotting factors VII, IX, XI, and XII	[a]	[a]
3. Hyperhomocysteinemia		5	10
4. Defects of the fibrinolytic system	Plasminogen	[a]	[a]
	Tissue plasminogen activator	[a]	[a]
	Thrombin-activatable fibrinolysis	[a]	[a]
	Factor XIII	[a]	[a]
	Lipoprotein (a)	[a]	[a]
5. Altered platelet function	Platelet glycoprotein GPIb-IX	[a]	[a]
	GPIa-IIa	[a]	[a]
	GPIIb-IIIa	[a]	[a]

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[a] Percentages unknown or not reported.

immobilization (environmental risk factor) may have contributed to his clot.

Imaging

Imaging plays an integral role in the diagnosis of PSS. The criterion standard for the diagnosis of venous thrombosis is venography; however, venous ultrasonography has become the most commonly used modality given its noninvasive nature.[24] Duplex ultrasonography is the most widely used, primary diagnostic modality for venous thromboses, as it combines grayscale or B-mode ultrasonography to visualize the vessel wall and color Doppler ultrasonography to visualize the blood flow through the vessel.[1,2,6,13,24] It has high specificity (94%) and sensitivity (97%) to allow for appropriate and immediate treatment when indicated.[24]

Clinical picture

As noted above, PSS is a rare condition; however, it is regarded as the most common vascular disorder to affect athletes.[11] Compression of the venous system by anatomical structures commonly occurs in positions of extreme abduction and/or external rotation,[1,6,7,13,14,16] as demonstrated by overhead athletes.[11] It has also been hypothesized that exercise may contribute to transient stasis or hypercoagulation, creating an ideal milieu for thrombus formation.[1,6]

It must be noted however that PSS is not a condition exclusive to young male athletes. Cases have been published of PSS occurring in backpackers,[25,26] and numerous authors have suggested that depression of the shoulder narrows the costoclavicular space and may be a mechanism for the development of PSS.[1,6,9,12,12,14,27] It is suspected that this mechanism was responsible for the development of PSS in these case studies. Particularly, the first patient may have experienced shoulder depression and narrowing of the costoclavicular space because the strap for his bass rests on his left shoulder for many hours daily.

The symptoms of PSS are generally nonspecific and can range in severity.[11] The most common presentation involves a “heavy” sensation in the affected arm with accompanying pain in the ipsilateral neck, shoulder, axilla, and/or arm.[11,16] As the syndrome progresses, patients may develop edema in the upper limb, ecchymosis and skin discoloration, and distension of

the cutaneous veins in the involved areas.[11] It is important to note that patients will rarely report a history of trauma or recent illness or infection that may be considered to explain the acute swelling. In addition, patients will be afebrile.[16]

Conservative management

Although there is no available literature to indicate a need for conservative care following PSS, the second patient in this case reported relief with stretching and soft tissue therapy. It is possible that reactive muscular tension or postural alterations may result as a compensatory mechanism in these patients; and as such, they may experience relief of any associated musculoskeletal complaints with appropriate conservative care after ruling out ongoing vascular abnormalities. Future prospective studies regarding specific interventions and appropriate outcome measures may provide more information regarding the need for such management.

Limitations

This is a description of the management of 2 cases and thus is limited in application to other conditions and the general population. The improvement of the second patient may have occurred because of factors not related to chiropractic care, so it is unknown how much the chiropractic treatment contributed to her improvement.

Conclusion

Clinicians should be aware of the clinical presentation and proposed pathogenesis of PSS, and consider this diagnosis in athletic and nonathletic patients with unilateral upper limb and/or neck pain. Although a rare condition, it has the potential to result in significant morbidity and potentially fatal complications; thus, it is critical that practitioners can recognize the signs and symptoms to facilitate appropriate and timely referrals.

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