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Case report

Paget-Schroetter syndrome in a non athlete - a case report [☆]

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ABSTRACT

A young male waiter presented with left shoulder pain, with ultrasound showing thrombosis of the left subclavian vein. After exclusion of hypercoagulability, a dynamic MRI upper limb venogram was performed, confirming Paget-Schroetter syndrome (PSS). Our case is unusual as Paget Schroetter syndrome are seldom reported in non athletes. This is also the first report to show dynamic MRI images of PSS.

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Introduction

Paget-Schroetter syndrome, or effort thrombosis of the axillary-subclavian vein, is an uncommon deep vein thrombosis that can lead to significant disability if not diagnosed and treated early. It is classically reported in athletes who perform repetitive shoulder-arm motions. We describe in this article an unusual case of Paget Schroetter syndrome in a young male waiter and his imaging findings. There were limited previous case reports on non athletes developing PSS, this is also the first report to show dynamic MRI images of PSS.

Case report

A 33-year-old waiter with good past health presented with insidious onset of left shoulder pain, left arm redness and swelling. The extremity was neurovascularly intact. His chest and shoulder radiographs were normal. Doppler sonographic study revealed acute subclavian vein thrombosis extending to the axillary vein (Fig. 1).

Hypercoagulability work-up was negative. He denied a history of self medication or intravenous drug use. CT and dynamic MRI venography was performed, showing no cervical rib or space occupying lesion at the thoracic outlet. However,

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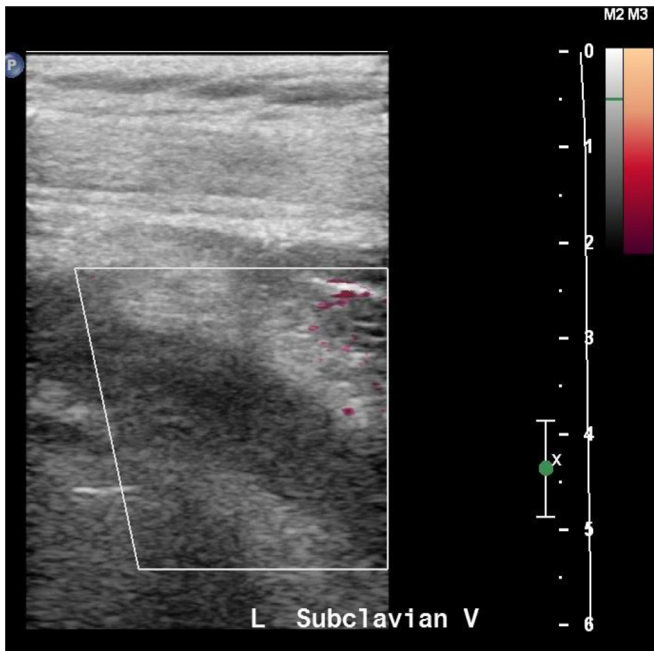


Fig. 1 – Echogenic thrombus and absence of doppler signal in the left subclavian vein, consistent with complete thrombosis.

even though the subclavian veins were not compressed at neutral shoulder position, their central portions were obliterated within the costoclavicular triangles during shoulder abduction (Fig. 2).

The patient volunteered that he had to maintain left shoulder abduction, holding several plates at a time, for hours when he was waiting at work. A diagnosis of Paget-Schroetter syndrome was made.

The patient was started on anticoagulants, and a follow up Doppler sonography 10 months after treatment (Fig. 3) showed chronic thrombosis of the left subclavian vein with interval formation of vascular collaterals. The patient reported persistent intermittent left arm pain and a subjective decrease in muscular endurance, but could manage his waiting job.

Discussion

Paget-Schroetter syndrome (PSS), also known as ‘effort thrombosis’, is a rare primary thrombosis of the axillary and/or subclavian vein. It was first described by Paget in 1875 and von Schroetter in 1884. Hughes later named the disease ‘Paget-Schroetter syndrome’ in 1948 [1]. Its incidence is estimated as one to two per 100,000 persons per year [2].

The aetiology of PSS is controversial, and is believed to relate to chronic compression of the subclavian vein at the level of the thoracic outlet. Thus, PSS is seen in young patients who use repetitive shoulder-arm motions, most classically vigorous weight-lifters, wrestlers, baseball pitchers or tennis players [3]. Our case is unusual as the patient was not physically active. Such non-athletic patient demographics have been also described in few other case reports (involving a waiter, a mixologist and a TV cameraman) [4,5,6]. These patients have no clear thrombotic risk factors and a negative hypercoagulability workup. Thus a high index of suspicion is needed to work up and prevent future thrombosis.

It is believed that repetitive upper-extremity exertion causes subclavian-axillary vein or perivenous trauma and activates the coagulation cascade [7]. Another theory is that repetitive movements result in scalene muscle hypertrophy (in particular the subclavius muscle), which in turn causes compression of the subclavian vein between the ribs anteriorly, muscle posteriorly, and clavicle superiorly [8].

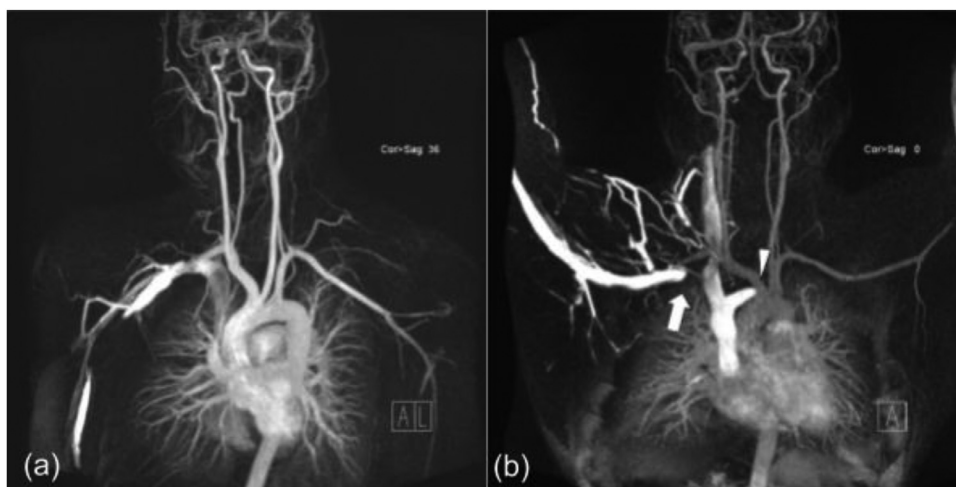


Fig. 2 – MR venogram (time-of-flight images) showing the right subclavian (arrow) being patent in neutral shoulder position (A) and compressed within the costoclavicular triangle during shoulder abduction (B). The left subclavian vein (arrow head) was thrombosed.

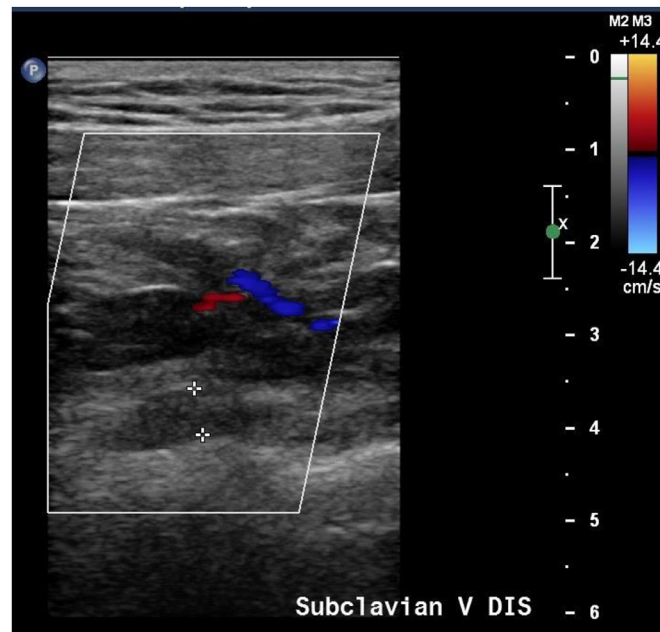


Fig. 3 – Follow up Doppler ultrasound showing chronic thrombosis of subclavian vein and interval development of venous collaterals.

A characteristic symptom triad is venous distension, arm swelling and aching pain in the affected extremity. Other signs include subcutaneous venous collaterals in the shoulder (Urschel's sign) and bluish discoloration.

Doppler ultrasound is the first-line imaging modality of choice, with a sensitivity of 78%–100% and specificity of 82%–100% [9]. Computed tomography and magnetic resonance venography are useful in patients with negative ultrasound results but a high index of suspicion for PSS, or to exclude a structural obstructive cause such as a cervical rib [7]. The MRI images in our case demonstrate the dynamic compression of the subclavian veins at abducted shoulder posture but not during a neutral shoulder position.

Our patient's thrombosis is likely related to his occupation, and he was advised of frequent postural changes in prevention of recurrence or contralateral involvement of PSS. He was treated with oral anticoagulation, which is the mainstay of treatment, but residual deep venous thrombosis and chronic disability has been described in up to 40%–90% PSS patients [3]. Recurrent pulmonary embolism or chronic thrombo-embolic pulmonary embolism can also result [2]. Some centers advocate for intravascular thrombectomy or thrombolysis, but there is no consensus on its superiority over the anticoagulant only regimen [8]. Some authors also described surgical decompression of the thoracic outlet through first-rib resection in cases of thoracic outlet syndrome. Decompression techniques include first-rib and/or clavicle resection, with transaxillary resection of the first rib being the most popular surgical intervention. Urschel and Razzuk suggested that prompt transaxillary first-rib resection and neurovascular decompression can be safely and effectively performed after thrombolytic treatment [10]. Urschel however recommended against intravenous stent-

ing for PSS, describing it to add no benefits compared to optimal thrombolysis and prompt resection of the first rib [11].

Conclusion

In conclusion, we hope to illustrate through this case that Paget-Schroetter syndrome is not an entity limited to athletes. PSS should be considered when one encounters spontaneous venous thrombosis of upper limbs. A focused history taking on the presence of excessive or repetitive arm activities would be suggestive. Dynamic imaging with patients posed in their habitual postures could further substantiate the diagnosis, and help prevent progressive and/ or contralateral thrombosis.

REFERENCES

- [1] Hughes ES. Venous obstruction in the upper extremity. *Br J Surg* 1948;36:155–63.
- [2] Joffe HV, Kucher N, Tapson VF, Goldhaber SZ. Upper-extremity deep venous thrombosis: a prospective registry of 592 patients. *Circulation* 2004;110:1605–11.
- [3] Kucher N. Deep-vein thrombosis of the upper extremities. *N Engl J Med* 2011;364:861–9.
- [4] Drakos N, Gausche-Hill M. A case report: a young waiter with Paget-Schroetter syndrome. *J Emerg Med* 2013;44:e291–4.
- [5] Daya A, Gillenwater S, Woytanowski JR, Hadeh A, Shaken Martinez N. Not stirred: deep vein thrombosis due to paget-schroetter syndrome from tending bar. *Am J Respir Crit Care Med* 2020;201:A7327.

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- [6] Beasley R, Braithwaite I, Evans R. Upper extremity deep vein thrombosis in a TV cameraman. *Occup Med (Lond)* 2015;65:337–9.
- [7] Alla VM, Natarajan N, Kaushik M, Warriar R, Nair CK. Paget-Schroetter syndrome: review of pathogenesis and treatment of effort thrombosis. *West J Emerg Med* 2010;11:358–62.
- [8] Illig KA, Doyle AJ. A comprehensive review of Paget-Schroetter syndrome. *J Vasc Surg* 2010;51:1538–47.
- [9] Baarslag HJ, van Beek EJR, Koopman MM, Reekers JA. Prospective study of duplex ultrasonography compared with contrast venography in patients suspected of having deep venous thrombosis of the upper extremities. *Ann Intern Med* 2002;136:865–72.
- [10] Urschel HC Jr, Razzuk MA. Paget-Schroetter syndrome: what is the best management? *Ann Thorac Surg* 2000;69:1663–9.
- [11] Urschel HC Jr, Patel AN. Paget-Schroetter syndrome therapy: failure of intravenous stents. *Ann Thorac Surg* 2003;75:1693–6.