Paget-Schroetter syndrome in a young fitness enthusiast with a negative D-dimer: highlighting the balance between clinical suspicion and diagnostic modalities

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DESCRIPTION
A 22-year-old man presented to the emergency department with a 3-week history of spontaneous right arm ache and swelling. He reported that his symptoms were exacerbated by exercise. There was no history of trauma, weight loss or fever. He had no previous medical history. He was a right-handed sportsman playing regular racket sports.

On examination, he had an oedematous right arm with a dusky discolouration (figure 1), with distended neck veins. The right arm had good volume pulses peripherally, with a capillary refill time of <2s. Sensory and motor function was preserved with no deficit bilaterally.

Routine baseline serum blood test and haemophilia screen were within normal range. Of note, his D-dimer was not raised at <150 ng/mL (normal range 0–230 ng/mL). He underwent CT neck/thorax/abdomen/pelvis with upper limb venogram. This demonstrated no evidence of lymph node enlargement or compressive pathology. Furthermore, the CT revealed good opacification of the neck and thoracic veins with adequate opacification of the arteries. Therefore, it effectively excluded both vascular compression and occlusive insult. Routine baseline serum blood tests and imaging results, we proposed alternative imaging modality. Doppler ultrasound sonography revealed a right distal subclavian vein thrombus with extension into the right brachiocephalic vein.

The patient was discussed in the vascular multidisciplinary meeting to discuss optimal revascularisation options. The joint discussion and case review confirmed the diagnosis of Paget-Schroetter syndrome (PSS). Given the subacute nature of the presentation, neither surgical decompression nor thrombolysis was deemed appropriate at this stage. Our patient was started and managed conservatively on oral anticoagulation (apixaban).

At 4 weeks follow-up, our patient had significant improvement of his right upper limb oedema and discolouration (figure 2). At this stage, elective surgical decompression was deferred pending further re-evaluation with dedicated imaging. On the second follow-up, 6 months postdischarge, although the patient continued to demonstrate improvement of symptoms, the improvement was insufficient. Our patient was both a competitive badminton player and a swimmer. He found his arms fatigued and oedematous within the early stages of a badminton game. Additionally, the biomechanics during the catch phase rotation in

Figure 1 On initial presentation to the hospital. (A) and (B) show significant swelling of the right arm with dusky discolouration.

Figure 2 Six months after commencement of anticoagulation. (A) and (B) show minimal swelling of the right arm with mild discolouration.

Video 1 Venogram demonstrating numerous venous collaterals in the right shoulder joint, with a short segment chronic occlusion of the right subclavian vein as it crosses the first rib with delayed enhancement and drainage of the right brachiocephalic vein and SVC.
freestyle swimming resulted in repetitive compression of venous outflow and eventually recurrence of symptoms.

Repeat right upper limb venogram revealed numerous venous collaterals in the right shoulder joint, with a short segment chronic occlusion of the right subclavian vein (video 1). Following a multidisciplinary team discussion, a decision was made to proceed to surgical intervention due to unsatisfactory resolution of symptoms. Our patient underwent a first rib resection with a right subclavian vein bypass using the saphenous as conduit. A combined approach was used due to the extent of proximal occlusion. Initially an infraclavicular approach was used to split pectoralis fibres and expose subclavian vein. Supraclavicular approach was next used to expose internal jugular vein to achieve end to side veno-veno bypass from the patent axillary vein to internal jugular vein. We performed first rib resection to safely tunnel the bypass under the clavicle. Our patient had a good outcome postoperatively and was commenced on oral anticoagulation for a further year to maintain patency of venous graft.

There is limited data in current literature regarding adjunctive therapy postsurgical decompression in PSS. Fairman et al found no benefit for long-term anticoagulation as adjunctive treatment postsurgical decompression.1 However, in our case, anticoagulation was continued for an additional year to maintain graft patency.

PSS, also known as effort thrombosis, is a form of upper extremity deep vein thrombosis. Aetiology of PSS is a combination of anatomical abnormalities of the thoracic outlet combined with repetitive movements of the upper limb, resulting in thrombus formation and subsequent venous occlusion. PSS is rare with an incident rate of around 1–2/100 000 population and is more common in patients in their 30s. PSS is diagnosed using clinical assessment, serum markers and imaging modalities.2

Diagnosis of PSS, in this case, was challenging due to a false-negative D-dimer and CT venogram. D-dimer has a high sensitivity of 96% for venous thromboembolism, however, the false-negative result in our case may be due to the chronicity of the culprit thrombotic event.3,4 D-dimer is also less sensitive in upper extremity deep vein thrombosis (UEDVT).5 Diagnosis of PSS in our case was confirmed on doppler USS, which should remain the first-line radiological investigation in suspected PSS. There is a lack of consensus on optimal management for PSS. Management can be conservative or surgical and our case demonstrates both management options and their outcomes.6

Contributors To whom it may concern, I would like to declare myself AG and GL as joint first authors. We have equally contributed in the case write-up and image production for this case AE-k was the lead medical consultant who alongside ISR the operating consultant reviewed our submission and provided invaluable advice and support to prepare for submission. Regards, AG.

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