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

Abrar Gani ¹, Georgia Lucas,^{2,3} Jonathan Simon Refson,⁴ Awad El-karim⁵

¹Department of Surgery, Princess Alexandra Hospital NHS Trust, Harlow, UK

²Princess Alexandra Hospital, Princess Alexandra Hospital NHS Trust, Harlow, UK

³Department of Paediatrics, North Middlesex University Hospital, London, UK

⁴Department of Vascular Surgery, The Princess Alexandra Hospital, Harlow, UK

⁵Department of Acute Medicine, Princess Alexandra Hospital, Harlow, UK

Correspondence to
Dr Abrar Gani;
abrar.gani1@nhs.net

AG and GL are joint first authors.

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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. In view of the inconclusive 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

Figure 2 Six months after commencement of anti-coagulation. (A) and (B) show minimal swelling of the right arm with mild discoloration.

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 discoloration ([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 discoloration.

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.

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Images in...

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 teams discussion, a decision was made to proceed to surgical intervention due to unsatisfactory resolution of symptoms. Our patient underwent a first rib

Patient's perspective

I would describe the first symptoms as a dull aching in my elbow region, which I would compare to tennis elbow. During the next week, I felt more aching until my whole arm felt extremely heavy and hard to the touch. During the first week where the clot was forming, I also saw the swelling increase. The only other symptom was discolouration which I would describe as 'greyish-blue'. I would not say that my arm ever felt painful, but very uncomfortable.

I presented to A&E and was admitted 3 week after onset of first symptoms. A venogram failed to show the clot, but the clot was found the next day (I was admitted for one night) by means of an ultrasound scan. During this one-night stay in hospital, I would say the symptoms doubled—this includes the swelling, discolouration and feelings of heaviness and hardness. The swelling was so severe that I could not bend my arm at the elbow. By the time I was discharged from hospital 1 day later (with apixaban and a compression sleeve for my arm), the symptoms were at their worst.

I revisited the consultant vascular surgeon after 2 days, and most of the acute swelling and other symptoms had reduced remarkably. After around 1 month of anticoagulation and compression, I was able to resume low intensity exercise such as exercise bike, jogging and other cardio machines. I spent the next 9 months doing low-intensity exercise in this way. I tried to restart other exercises using my arm (including weightlifting, swimming and racket sports), but these caused a small amount of the original symptoms to return. I noticed that these symptoms would disappear within 10 mins of stopping exercise if I held my arm above my head. I believe this is mostly due to the expansion of my collateral veins, which became more visible under my skin around my bicep, shoulder and pectoral muscle as time went on.

About 6 months after developing the clot, I was given a venogram that showed there had been little or no dissolving of the clot, but my collaterals had expanded well. I was not satisfied with this, as I was still getting some symptoms when doing very strenuous exercise involving my arm. Therefore, I requested an operation to improve the flow of blood in my subclavian vein.

On 13 August 2020, I underwent a complete first rib resection and bypass from my subclavian vein to my jugular. The vein was harvested from the saphenous vein. I stayed in hospital for five nights after this. I am now writing this account 2 weeks after the surgery. The pain has now reduced, and I am left with a soreness and stiffness in my shoulder. This will be corrected with the help of physiotherapy over the next couple of months. I have already noticed that the visible collaterals have reduced which I believe indicates a successful bypass. A venogram was conducted during the surgery and showed there was good flow of blood through the bypass.

I hope that in 6 months, the bypass will allow me to resume strenuous upper body exercise without getting any symptoms at all.

Learning points

- ▶ Clinical judgement based on clinical assessment is invaluable. Additional investigations should be utilised to support clinical suspicion.
- ▶ Paget Shroetter Syndrome is an important differential to remember in young healthy athletic individuals presenting with unilateral swelling
- ▶ Treatment of Paget Shroetter syndrome includes pharmacological and percutaneous therapies alongside surgical decompression.

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

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ORCID iD

Abrar Gani <http://orcid.org/0000-0002-6772-3305>

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