Case Report

Primary upper extremity deep vein thrombosis (effort thrombosis)

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Abstract

Primary spontaneous upper extremity deep vein thrombosis is characterised by thrombosis within deep veins draining the upper extremity due to anatomical abnormalities of the thoracic outlet causing axillosubclavian compression and subsequent thrombosis. It is an uncommon condition that typically presents with unilateral arm swelling in a young male following vigorous upper extremity activity. The diagnosis of this condition is usually made by Doppler ultrasound, but other investigations are mandatory to exclude the secondary causes of upper extremity DVT. Different treatment options are available including anticoagulation, thrombolysis, and surgery. We report the case of a young healthy male with athletic physique who presented with pain and swelling of his dominant arm after weightlifting in the gym.

Keywords

Effort thrombosis, Paget-Schroetter syndrome, Upper extremity DVT

Teaching Points

1. Effort thrombosis typically presents with arm swelling following a strenuous activity.
2. Doppler ultrasound is the initial test to confirm the diagnosis of upper extremity DVT.
3. Unlike lower extremity DVT, D-dimer is not a part of the clinical pathway for diagnosis of upper extremity DVT.
4. Effort thrombosis is a diagnosis of exclusion and work up to rule out secondary causes is warranted.
5. DOACs for 3-6 month is acceptable as a treatment for primary UEDVT.

Case presentation:

A 30-year old male presented to the Medical Unit with right arm pain and swelling for 3 weeks. The pain was a dull ache in nature, of moderate severity and extending from the shoulder down to the fingers. There was also mild neck and axillary pain. The pain was reproduced by elevation of the arm above the level of head. He denied any shortness of breath, chest pain, cough, or fever. There was no personal or family history of coagulopathy. The patient enjoyed lifting weights at the gym four times per week and playing badminton twice a week. He had never smoked and consumes alcohol occasionally. On examination,the patient was a muscular young man weighing 90kg. The right arm circumference was 3 cm larger than the left arm measured at the mid-point between the tip of the acromium and the tip of the olecranon. There was no cyanosis or dilated superficial veins. Radial and brachial pulses were easily palpable and neurological examination was normal. Initial investigations including full blood count, biochemical profile, coagulation profile, C-reactive protein and D-dimer were normal (Table 1). Unilateral arm swelling differential diagnosis is wide and includes venous compression or thrombosis, cellulitis and lymphoedema. Cellulitis was unlikely given the absence of fever and normal inflammatory markers. Lymphoedema is usually associated with an antecedent risk factor such as prior axillary lymph node dissection. In this case, despite the normal D dimer, Doppler ultrasound demonstrated a thrombus in the right subclavian vein at the junction with the jugular vein (Figure 1).

Following confirmation of right subclavian thrombosis, chest and abdominal CT imaging were performed to exclude a cervical rib and occult malignancy. An inherited thrombophilia screen and antibody testing for antiphospholipid syndrome were normal. In the absence of a causative iatrogenic factor (central venous cannulation) or a secondary cause for thrombosis (hereditary thrombophilia, antiphospholipid syndrome), a diagnosis of effort thrombosis was reached. The patient was commenced on therapeutic low molecular weight heparin (LMWH) and subsequently received apixaban for a total of three months. He was advised to refrain from strenuous upper extremity activities. The patient remained systemically well, tolerated the treatment well and was discharged to his general practitioner. He completed a three-month period of treatment and did not develop further complications. He did not seek medical attention either from primary or secondary care following this episode.

Discussion

Upper extremity deep vein thrombosis (UEDVT) is the formation of fibrin clots within the deep veins draining the upper extremity (subclavian, axillary, or

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Primary upper extremity deep vein thrombosis (effort thrombosis)

Investigation	value	Units	Reference range
Haemoglobin, blood	148.0	g/L	135.0-175.0
WBC, blood	5.4	x 10 9 /L	
Platelet ,blood	253	x 10 9 /L	
Prothrombin time (PT), blood	11.9	s	9.5-12.0
APTT, blood	29.1	s	20.0-29.0
D-dimer,blood	200	ng/ml	0-500
C-reactive protein,blood	0.5	Mg/L	0.8
Creatinine level, serum	79	umol/L	65-114
Urea level, serum	4.8	mmol/L	3.0-7.6
Alanine aminotransferase level, serum	10	[iU]/L	5-45
Alkaline phosphatase level, serum	45	[iU]/L	30-125
Bilirubin level,serum	16	Umol/L	<21
Total protein level, serum	76	g/L	65-82
Albumin level, serum	48	g/L	36-48
Viscosity, blood	1.68	Mpa s	1.50-1.72
Lupus anticoagulant screen ,blood	Not detected		
Anticardiolipin level, serum	<2	GPLU/ml	0-19
AntiB2-glycoprotein-1 AB level, serum	<1	U/ml	0-19

brachial veins). It accounts for approximately 4-10% of cases of deep vein thrombosis. While the majority of cases of upper extremity deep vein thrombosis are secondary to central venous cannulation (e.g. central line, pacemaker) or prothrombotic states (e.g., thrombophilia, malignancy), Primary UEDVT is rare and is caused by effort thrombosis (Paget-Schroetter Syndrome) or thoracic outlet syndrome.[1]

Effort thrombosis is typically seen in young adolescent athletic males and is precipitated by repetitive overarm hyperabduction resulting in microtraumatic endothelial damage. Venous thoracic outlet syndrome occurs due to compression of the subclavian vein at the costoclavicular junction e.g. cervical rib.[2]

Upper extremity DVT can present acutely with swelling of the dominant arm associated with pain worsened by exercise or hyperabduction of the affected arm. There may be vague pain in the neck and shoulder or pain radiating to the fourth and fifth digits indicating thoracic outlet obstruction. Clinical examination may reveal oedema of the affected limb, cyanosis, low-grade fever or dilated subcutaneous collateral veins (Urschel's sign). Of note, pain and oedema may be minimal in patients with partial thrombosis or chronic venous stenosis due to repetitive injury.[1,2] A focussed examination to exclude malignancy should be included in the initial clinica assessment.

Whilst D-dimer is routinely used in the exclusion of low clinical suspicion lower limb DVT due to its high negative predictive value, it does not form part of a clinical pathway for the exclusion of upper limb DVT. A positive value is not diagnostic and does not aid location of the thrombus.[3] In this patient

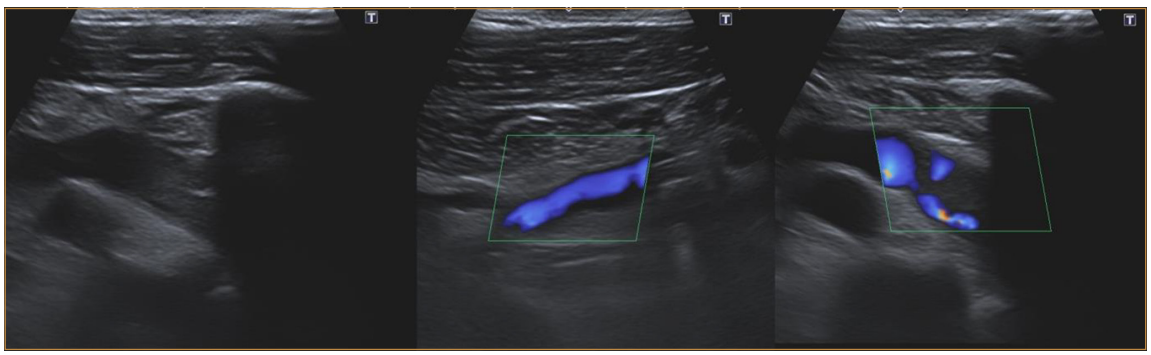

Figure 1. Doppler ultrasound of the right upper limb showing a thrombus in the right subclavian vein at the junction to the jugular vein

plasma D-dimer was normal, likely due to delay in presentation.

Combined modality ultrasound (compression with either Doppler or colour Doppler) is the initial test to diagnose upper extremity DVT. Other non-invasive methods like CT or MR venography are usually reserved for atypical presentations or equivocal US results.[4] While abnormal venous anatomy is best demonstrated by catheter-based venography, the method is an invasive one, and as such does not form the gold standard for diagnosis. The diagnosis in our case was established with combined ultrasonography (Figure 1).

There is debate about the role for thrombophilia screening after UEDVT. Identification of any underlying hypercoagulable state like hereditary thrombophilia and malignancy should be included in the work-up when appropriate. Patients presenting with classic signs of venous outflow obstruction should have a plain chest X-ray to identify bony abnormalities.[1]

Treatment with immediate systemic anticoagulation aims to alleviate symptoms, prevent progression of thrombosis and reduce complications such as pulmonary embolism, recurrence and post-thrombotic syndrome.[1,2] The American College of Chest Physicians recommends anticoagulation for a minimum of three months for all patients with UEDVT,[5] while the British Society of Haematology advocates a period of three to six months.[6] Direct oral anticoagulants appear as effective as vitamin K antagonist for prevention of recurrent VTE and compare favourably for major bleeding, clinically relevant non-major bleeding, and death,[7] and do not requiring therapeutic drug monitoring. Catheter directed thrombolysis may lead to improved outcomes compared with systemic anticoagulation alone if done within 14 days of thrombosis in selected patients with acute (<14 days) severe symptoms, thrombus involving most of the subclavian and axillary veins, and a life expectancy of >1 year.[5,8] Surgical procedures such as thoracic outlet decompression may have a role in selected patients in the appropriate clinical context.[1]

Conclusion

Clinicians should be aware of the association between exercise and development of venous thrombosis in the upper extremities. We re-emphasise the fact that effort thrombosis is a diagnosis of exclusion that warrants initiation of a set of investigations to rule out the secondary causes of DVT. Importantly, the choice of treatment modalities should be individually tailored to each patient depending on the onset of presentation, severity of symptoms, and life expectancy.

Financial and competing interests disclosure

The authors declare that they have no relevant affiliations or financial involvement with any organisation or entity with a financial interest in or financial conflict with the subject matters or materials discussed in this manuscript.

No writing assistance was utilised in the production of this manuscript.

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