Axillary vein thrombosis

Synonyms: Paget-von Schröetter disease, axillary subclavian deep vein thrombosis, upper-extremity deep vein thrombosis, upper-limb deep vein thrombosis, effort-induced thrombosis

Definition

Acute swelling and pain in the upper limb due to occlusion of the axillary and/or subclavian veins by thrombus. This may occur as a primary phenomenon or as a result of secondary factors such as the placement of an indwelling venous catheter, thrombophilia or thoracic outlet syndrome.

See also the separate Deep Vein Thrombosis article.

Epidemiology[1]

- There is a low incidence of about 1 per 100,000 people per year. 4-10% of all deep vein thromboses (DVTs) occur in the upper extremity.
- It is now more common due to the growing use of central venous cannulation in a variety of medical procedures[2]. It occurs in about 25% of patients who undergo prolonged central venous cannulation, although it is often not recognised.
- About 80% of primary cases occur in the dominant arm. It may occur in young, otherwise healthy, individuals who participate in repetitive upper limb exercises[3].

Risk factors[4]

- The presence of a central venous catheter[5].
- Venous compression in the thoracic outlet syndrome.
- Malignancy.
- Congenital thrombophilia, acquired coagulation defects.

- Diabetes mellitus.
- Smoking habit.
- Intense sports activity.

Presentation

Symptoms [6]

Symptoms can be intermittent, or can develop during a period of up to one week.

- Patients tend to present with discomfort and swelling, associated with discolouration of the hand.
- 3-36% of cases may lead to pulmonary embolism with features of pleuritic chest pain, breathlessness and haemoptysis.

Signs

- Physical examination may show low-grade fever due to thrombus formation. Higher fevers are seen with septic thrombophlebitis or in patients with associated malignancy.
- Oedema of the arm and hand - measure the biceps/forearm diameter at a fixed distance from an anatomical landmark.
- Mild-to-moderate cyanosis of the hand.
- Dilated superficial collateral veins may be seen over the chest and upper arm - may be the only indicator in central venous cannulation.
- Fullness in the supraclavicular fossa and even a palpable cord of thrombosed vein.
- Jugular vein may be distended.

Differential diagnosis

- Superficial phlebitis.
- Cellulitis.
- Severe superficial bruising.

- Muscular tear.
- Intramuscular haemorrhage.
- Lymphoedema.
- Occult fracture.
- Superior vena cava obstruction.
- Lymphangitis.
- Localised allergy.
- Gas gangrene.

Investigations[6]

- Ultrasound (compression with either Doppler or colour Doppler) is recommended as the investigation of choice.
- D-dimer testing is less useful than in lower-limb thrombosis, particularly in hospitalised patients with central venous catheters or malignancy.
- In patients with suspected upper-extremity DVT in whom initial ultrasound is negative for thrombosis despite a high clinical suspicion of DVT, CT scan or magnetic resonance phlebography is recommended.
- It is uncertain whether routine thrombophilic screening in patients with this condition is worthwhile. It is probably useful where it occurs idiopathically, with a family history of thrombosis or history of recurrent miscarriage or previous DVT.
- Imaging investigations to detect thoracic outlet syndrome should depend on the degree of clinical suspicion of this cause.
- In idiopathic cases one should consider investigations to look for an occult malignancy or thrombophilia.

Management[7]

- Acute treatment with parenteral anticoagulation (low molecular weight heparin, fondaparinux, intravenous/subcutaneous unfractionated heparin) is recommended. Low molecular weight heparin or fondaparinux are preferred. Rivaroxaban and apixaban can also be used[8].
- Anticoagulant therapy is recommended in preference to thrombolysis. Anticoagulation should be for a minimum of three months.
- For patients who undergo thrombolysis, the same intensity and duration of anticoagulant therapy should be used as for those patients who do not undergo thrombolysis.
- For those patients with upper-extremity DVT that is associated with a central venous catheter, it is recommended that the catheter should not be removed if it is functional and there is an ongoing need for the catheter.
- If the catheter is not removed then anticoagulation should be continued as long as the central venous catheter remains but there should be a minimum of three months of treatment.
- A trial of compression bandages or sleeves to reduce symptoms is recommended for post-thrombotic syndrome of the arm (chronic venous insufficiency that may cause pain, oedema, pigmentation, skin changes and venous ulcers).

Prevention

The use of anticoagulant prophylaxis in patients who are acutely ill and those who undergo central venous catheterisation may prevent upper-extremity DVT. Many patients are also at risk of lower-limb thrombosis, which warrants anticoagulant prophylaxis in its own right[1].

Prognosis

Associated with significant morbidity and mortality due to potential risks of pulmonary embolism, post-thrombotic syndrome and loss of vascular access.

- About 10-20% develop pulmonary embolism [9].
- Reported mortality rates have varied from 15-50%, largely dependent on the underlying cause [10].
- Recurrent thrombosis affects between 2% and 5% of patients [1].
- Post-thrombotic syndrome affects 13% (see 'Complications', below) [10].

Complications

- Pulmonary embolism has been detected on radiological grounds in up to 20% of patients with upper-limb DVT (incidence is highest in untreated/catheter cases).
- Phlegmasia caerulea dolens (PCD) may occur (rarely); there is arterial and venous compromise with a risk of gangrene.
- Compartment syndrome.
- Recurrent thrombosis.
- Post-thrombotic syndrome - chronic upper-limb pain and swelling.
- Stroke following paradoxical embolisation in cases with a patent foramen ovale.
- Right ventricular failure.
- Thoracic duct obstruction.
- Chylous pleural or pericardial effusion.

Further reading

- Stake S, du Breuil AL, Close J; Upper Extremity Deep Vein Thromboses: The Bowler and the Barista. Case Rep Vasc Med. 2016;2016:9631432. doi: 10.1155/2016/9631432. Epub 2016 Oct 9.
- Huang CY, Wu YH, Yeh IJ, et al; Spontaneous bilateral subclavian vein thrombosis in a 40-year-old man: A case report. Medicine (Baltimore). 2018 Apr;97(15):e0327. doi: 10.1097/MD.0000000000010327.

Authored by:	Peer Reviewed by: Dr Laurence Knott	
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