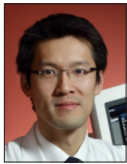


ADVANCES IN HEMATOLOGY

Current Developments in the Management of Hematologic Disorders

Section Editor: Craig M. Kessler, MD

Diagnosis and Treatment of Upper Extremity Deep Vein Thrombosis and Thoracic Outlet Syndrome



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H&O How common is upper extremity deep vein thrombosis (DVT)?

YWL Upper extremity DVT is relatively rare, accounting for no more than 10% to 20% of all cases of DVT. Secondary DVT is more common than primary DVT.

H&O Who is at risk for upper extremity DVT?

YWL Patients with an intravenous device, such as a pacemaker, defibrillator, peripherally inserted central catheter (PICC), or dialysis line, are at increased risk. The larger the diameter of the device, the greater the risk—so a dialysis line, for example, would carry a greater risk than a PICC. People who are in a hypercoagulable state also are at increased risk, as are patients who have a narrowed thoracic outlet.

H&O What are the signs of upper extremity DVT?

YWL Patients present with typical symptoms, such as swelling, congestion, some pain, tightness, and occasionally a darkish blue discoloration in the arm.

H&O How do physicians diagnose upper extremity DVT?

YWL The clinical examination tells us a lot. A D-dimer test can be useful in the outpatient setting, but false-positives are common in patients who recently have undergone surgery or another procedure. The best test for confirming a diagnosis of DVT is duplex ultrasonography.

H&O What are the complications of upper extremity DVT?

YWL Patients whose condition goes unrecognized have a small chance of developing a pulmonary embolism and subsequently post-thrombotic syndrome, although that is rarer in patients with upper extremity DVT than in those with lower extremity DVT. When the condition is diagnosed early and treated, patients have a reduced chance of complications.

H&O In what percentage of cases does embolization to the lung occur?

YWL Probably in fewer than 5% of cases.

H&O Should a patient with a central venous access device receive upper extremity DVT prophylaxis?

YWL This is a controversial question. The American College of Chest Physicians guidelines on antithrombotic therapy for VTE that were published in 2012 recommend that patients not receive DVT prophylaxis, but some physicians prescribe a daily dose of 1 or 2 mg of warfarin, based on some published research. This dose is small enough to have no effect on the international normalized ratio (INR) or prothrombin time, so there is no risk of clinical bleeding. We do not know whether it is effective, however, and I do not recommend using warfarin routinely in this situation.

H&O Could you provide a brief overview of thoracic outlet syndrome (TOS), and how common it is?

YWL TOS is a rare condition affecting the thoracic outlet, which is the area underneath the collarbone and above the first rib. It occurs when the neurovascular structures that pass through the outlet become compressed. Nerve compression develops in approximately 90% to 95% of cases, venous compression in approximately 5%, and arterial compression in approximately 1% to 2%. Because I am a surgeon working at a referral center for TOS, I see a slightly different distribution: approximately 40% to 50% of my patients who undergo TOS surgery have nerve compression, 30% to 40% have venous compression, and 5% to 10% have arterial compression.

H&O Is TOS more frequently bilateral or unilateral?

YWL TOS is more frequently unilateral. Only 10% to 20% of cases are bilateral.

H&O Who is at risk?

YWL We see a fair number of athletes with the condition, especially if their sport involves repetitive, over-the-head arm motion, such as swimming and baseball—pitchers are especially at risk. This motion can result in compression of the thoracic outlet and the collarbone. We occasionally see TOS, especially neurogenic TOS, in people who have experienced trauma, such as a whiplash injury.

Although we understand some of the causes of TOS, most of the time we do not know why it develops in certain patients. Many patients do not have any recognized predisposing factors.

H&O What is the relationship between upper extremity DVT and TOS?

YWL In approximately 5% to 10% of patients in whom venous TOS develops, the amount of blood returning to the heart is decreased because a narrowed thoracic outlet is compressing the subclavian vein. This situation can lead to upper extremity DVT. A milder condition that can occur is McCleery syndrome, in which DVT does not necessarily develop, but the patient experiences swelling and congestion in the arm when engaging in activities that involve an overhead motion, such as swimming. Sometimes, I am able to operate on these patients when they are still at this stage, before DVT develops, but more typically I do not see them until they have DVT.

H&O How is TOS detected?

YWL Patients with the neurogenic type of TOS experience numbness and tingling in the upper extremity, and those with the venous type may notice swelling of their arm and present with DVT. Arterial compression can lead to arterial thromboembolism, including blockage in the subclavian artery, and to distal embolization resulting in blockage of the axillary, brachial, radial, or ulnar artery. Frequently, a subclavian artery aneurysm is also present at the site of the arterial compression. As for diagnosis, we usually can get a lot of information by performing compression ultrasound while the patient is positioned with the arms down, overhead, and at 90 degrees. Some physicians will order a computed tomographic scan, which can be helpful but usually is not necessary, except in cases of arterial TOS. We have the option of using invasive venography to confirm a diagnosis of venous TOS, but this also is usually not necessary unless the patient requires thrombolysis for an acute presentation of DVT in the upper extremity.

H&O Should fibrinolytic therapy be used at the time of TOS diagnosis?

YWL The standard treatment is to administer catheter-directed fibrinolytic therapy to a patient with venous TOS during the acute presentation. Here at Hopkins, however, we have observed retrospectively in our surgical patients that fibrinolytic therapy makes very little difference overall, as discussed in a study by Guzzo and colleagues published in the *Journal of Vascular Surgery*. I still think fibrinolytic therapy should be considered, especially for patients who have a lot of swelling and other symptoms of DVT. There is no way to know in advance whether it will be effective, but sometimes it does work. If one of my patients does not receive thrombolytic therapy during the initial, acute presentation, or if the therapy fails to work and thrombosis recurs, I reassure the patient that this is not cause for worry.

H&O How should physicians treat upper extremity DVT?

YWL All patients need anticoagulation, whether with low-molecular-weight heparin, warfarin, or a novel oral anticoagulant. The novel oral anticoagulants have been shown to be at least as effective as warfarin in the treatment for DVT, with no real increase in risk for bleeding. They also can be more convenient to administer. The downsides are their increased cost and the lack of antidotes for them at this point.

The other treatments used depend on the cause of the DVT. Patients with primary DVT that may be related to venous TOS should be referred to a specialist with experi-

ence in the treatment of this condition; surgery often can reduce the recurrence of DVT and improve quality of life in terms of reducing recurrent swelling. I usually recommend surgical decompression after talking to the patient about all of the risks and benefits. We perform surgical decompression by taking out the rib and muscles around the subclavian vein to alleviate the compression. At 1 or 2 weeks after surgical decompression, we use venography to visualize the subclavian vein and use angioplasty to reopen it. I never place a stent in this location because the patients usually are very young, and I worry about possible fracture of the stent in the future. I occasionally must perform an open vein reconstruction, but that is rarely necessary. In most cases, the patient is left with nothing more than a tiny scar beneath the armpit.

For patients with secondary DVT due to a dialysis line or a PICC, the guidelines in *Chest* discuss the different scenarios, which vary according to whether the line is still needed.

H&O What is the success rate for surgery to reverse TOS?

YWL More than 95% of patients have great results, with a full return of arm function. I have operated on many

athletes who do well, including a Division 1 baseball player who went back to pitching at 95 to 100 miles per hour.

H&O Should patients with TOS receive genetic testing for hypercoagulable states?

YWL Some institutions do this routinely in patients with a first episode of upper extremity DVT due to TOS. In my opinion, it adds very little value in this situation—I have found that the results usually are negative in patients who have DVT due to TOS. In addition, the implications of a positive result are quite significant because patients may be committed to lifelong anticoagulation if any significant abnormalities are identified.

Suggested Readings

Grunebach H, Arnold MW, Lum YW. Thoracic outlet syndrome. *Vasc Med.* 2015;20(5):493-495.

Guzzo JL, Chang K, Demos J, Black JH, Freischlag JA. Preoperative thrombolysis and venoplasty affords no benefit in patency following first rib resection and scalenectomy for subacute and chronic subclavian vein thrombosis. *J Vasc Surg.* 2010;52(3):658-662.

Kearon C, Akl EA, Comerota AJ, et al; American College of Chest Physicians. Antithrombotic therapy for VTE disease: antithrombotic therapy and prevention of thrombosis, 9th ed: American College of Chest Physicians Evidence-Based Clinical Practice Guidelines. *Chest.* 2012;141(2 suppl):e419S-e494S.

Moore R, Wei Lum Y. Venous thoracic outlet syndrome. *Vasc Med.* 2015;20(2):182-189.

Erratum

An interview with Elias J. Jabbour, MD, that appeared in the December 2015 issue, “Use of PCR Testing in Chronic Myeloid Leukemia,” had 2 errors that were spotted by a reader. The sentence “The results are expressed as a reduction of the protein from the baseline measurement” has been changed to “The results are expressed as a reduction in the mRNA transcripts from the baseline measurement,” and the table heading “International Scale Percentage of Cells With BCR-ABL1” has been changed to “BCR-ABL/Control Gene Ratio (according to the International Scale).” Readers are advised to download the corrected version at www.hematologyandoncology.net.