CLINICAL VIGNETTE

Paget-Schroetter Disease and Recurrent Spontaneous Upper Extremity DVTs

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Clinical Case

A 32-year-old female with no significant past medical history presented to the Emergency Department with pain and swelling in left forearm for 2 days. Her pain and swelling started after a rigorous CrossFit exercise class a few days ago. She is a non-smoker, physically active and at a normal weight (BMI=22 kg/m2). She uses a Mirena IUD for birth control. A doppler ultrasound in Emergency department was significant for a left upper extremity subclavian clot. She was started on enoxaparin and transitioned to warfarin which she continued for 3 months. Hematologic evaluation noted normal values for: protein-C, protein-S, prothrombin time, and INR. Anticardiolipin antibodies and beta-2 glycoprotein were normal as well.

Her symptoms improved. However, one year later she returned to the Emergency department with similar right upper extremity pain for the past day. Her pain started after doing a rigorous exercise routine. A doppler ultrasound in the Emergency department revealed an acute thrombus in the brachial vein. She was again started on enoxaparin and transitioned to warfarin which she continued for 6 months. She was eventually diagnosed with Paget-Schroetter Disease.

Discussion

Paget-Schroetter syndrome, otherwise known as primary spontaneous upper extremity deep vein thrombosis, is a rare form of thoracic outlet syndrome that typically presents in young, otherwise healthy individuals with sudden, severe upper extremity pain and swelling following vigorous upper extremity activity. The syndrome is alternatively referred to as "effort" thrombosis.[1] The pathogenesis of Paget-Schroetter is thrombosis of the deep veins draining the upper extremity due to anatomic abnormalities of the thoracic outlet causing axillo-subclavian compression and repeated injury to the underlying axillo-subclavian vein with subsequent thrombosis.[2] Thoracic outlet syndromes have typically attributed to an underlying, congenital or acquired anatomic anomaly that narrows the costoclavicular space predisposing the vein to compression. But under rare circumstances, it appears that no anatomic abnormality is necessary to produce injury to the vein. Extremes in range of motion of the upper extremity can lead to movement of the clavicle relative to the first rib sufficient to cause venous compression. Repetitive overhead arm movements or hyperabduction and external rotation of the shoulder are most often implicated.[3]

Paget-Schroetter usually presents acutely with symptoms and signs of upper extremity deep vein thrombosis. There is always concern for associated pulmonary embolism but it is less common with upper thromboses then with lower thromboses, complicating only about 4-10 percent of cases.[4] Symptoms are often acute onset in nature, however, chronic or intermittent symptoms can be seen in patients with partial thrombosis or those with chronic venous stenosis due to repetitive injury that can cause activity-related obstruction. Such patients also have symptoms that are typically less severe. Swelling or pain may be minimal, and increased venous collateral flow over the chest may be the only clinical sign.[5] Risk factors for both acute and chronic disease include: younger age (including children, typically adolescents); men with athletic or muscular build; strenuous upper extremity activity; repetitive overarm hyperabduction; anatomic abnormalities of the thoracic outlet whether congenital or acquired; and/or underlying thrombophilia.[6]

A diagnosis of Paget-Schroetter may be suspected based upon the clinical presentation but should be confirmed with imaging, typically initially with doppler ultrasound. Once a diagnosis is established, a primary etiology should be sought to identify the underlying anatomic abnormality that is the source of the obstruction. A plain chest radiograph should be completed to identify any obvious bony abnormalities. More advanced imaging may be necessary to demonstrate abnormal muscle attachments or dynamic venous compression for atypical presentations. Screening for thrombophilia should be included in the evaluation due to the high prevalence of an underlying thrombophilia in primary upper extremity deep venous thrombosis.[7]

Treatment of Paget-Schroetter after diagnosis should be immediate to prevent pulmonary embolism and recurrent venous thrombosis. Acute and symptomatic disease is treated with anticoagulation, usually initially with a heparin compound. The decision to proceed with additional treatment of thrombolysis or thoracic outlet decompression is based upon symptom severity and the type of associated anatomic abnormality found. For patients with mild, intermittent, or chronic symptoms, they can be transitioned to oral anti-coagulants and be managed on an outpatient basis.[8] Novel oral anticoagulants, such as dabigatran, can be used and appear as effective as warfarin and offer more convenience for the patient.

Many patients with severe symptoms related to primary upper extremity deep vein thrombosis may require admission to manage symptoms. For these patients, thrombolysis should be considered followed by thoracic outlet decompression. This approach decreases the risk for recurrent thrombosis.[9]

In patients with Paget-Schroetter, anticoagulation should be continued for a minimum of three months following the initial thrombotic event, with a longer duration of therapy indicated for those who have had a recurrent event. Recurrent upper extremity deep vein thrombosis occurs in 2 to 8 percent of patients following treatment.[10]

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