CLINICAL VIGNETTE

Deep Vein Thrombosis from Pseudotumor

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A 77-year-old female presented to hematology for a new deep venous thrombosis (DVT), pleural effusion and pelvic mass. The patient noted increasing shortness of breath for several months. Her primary care physician obtained a chest radiograph with a large, left pleural effusion. Computed tomography (CT) of the chest confirmed the large left pleural effusion without other abnormalities. Given worsening symptoms, she was admitted to the hospital for expedited evaluations with thoracentesis performed on two separate occasions with negative cytology. Given ongoing concern for malignancy, CT abdomen and pelvis confirmed the pleural effusion and noted a 7.3 centimeters (cm) and cholelithiasis with a dilated 1.3cm common bile duct. Abdominal ultrasound confirmed gallstones with negative Murphy's sign and no indications of infection or inflammation in the biliary region. Given some mild left lower leg swelling, bilateral lower extremity ultrasound noted an acute non-occlusive thrombus in the left femoral and soleal veins and a non-vascular mass in the left inguinal region. She was started on rivaroxaban. Biopsy of the pelvic mass returned non-diagnostic. Discharge magnetic resonance imaging of the pelvis noted a 10.9cm mass consistent with an aseptic lymphocyte-dominant vasculitis-associated lesion (ALVAL)/ pseudotumor anterior to a left hip joint prosthesis which extended into the lower abdominal quadrant and displaced the left external iliac vessels. She was re-admitted for pleural biopsy and pleurodesis. All pathology from this procedure was negative for malignancy.

The patient was quite healthy for her age with mild hypertension, gastric reflux, and Gilbert's disease. She had a prior left total hip arthroplasty (THA) with a complicated postoperative course and required a repeat replacement of the joint. At the time of hematology evaluation, she had no B symptoms, was active, and feeling better after recent pleurodesis. Hypercoagulable testing was negative including Factor V Leiden, prothrombin gene mutation, Protein C and S deficiencies, Antithrombin III deficiency, antiphospholipid syndrome, and homocysteinemia.

ALVAL is a potential complication of total hip arthroplasties.[1,2] It is conjectured that metal ions from the prosthesis are released during normal wear and trigger a type IV hypersensitivity response by forming haptens with native proteins.[1] The metallic remnants of wear can often be detected in nearby soft tissue, lymph nodes and even organs including spleen and liver.[1] Common pathologic findings include necrotic or fibrinous exudate and prominence of macrophages.[1,2] Case reports noted associations with inflammatory arthritis and hyperplasia.[1] A defining feature is a perivascular infiltrate that leads to the name.[1,2] While a type IV sensitivity is a common hypothesis, it is unclear if it results from a direct allergy to the prosthetic material, or rather, a reaction to loosening or other complications from the surgery.[1,2] This is further supported by the fact that the pathologic features are very non-specific.[1] Symptoms usually include pain, due to the pseudotumor that is produced and consequent loosening of the hardware although this symptom is not always present.[1,2] While ALVAL appears to be a fairly rare complication, numbers are likely to increase with growing use of THA and utilization of artificial hips in younger patients leading to more potential years for wearing complications.[1,2] Various terms are used in other case reports including ALVAL, pseudotumor, and metallosis and some believe they all represent a spectrum from local allergic reaction to metallosis and finally pseudotumor.[3] The changes can take months or even years to develop.[2] While standard radiographs can identify many complications associated with failing prostheses, radiographs often misses ALVAL.[2] MRIs may note a periprosthetic mass or fluid collection.[2] Intraoperative evaluation is required and if the diagnosis is confirmed as ALVAL or a pseudotumor, replacement of the prosthetic with non-metal materials is required.[1,3]

Outcomes after revision vary, and in severe cases of ALVAL, prognosis may be poor.[2] In minor cases, symptoms generally resolve.[2] In severe cases, consequent damage to other local structures like bone marrow, muscles, may be extensive.[2] Furthermore, even with thorough debridement, some of the inflammatory makeup of ALVAL may remain and persist even with the new non-metal components.[2]

This patient did not have significant left hip symptoms. The left pleural effusion led to concern for malignancy with further imaging and testing. Multiple assessments confirmed that the fluid was not related to cancer. It is suspected that the inflammatory response related to the ALVAL likely led to the pleural effusion. Similarly, the mass effect of the pseudotumor on local blood vessels caused venous obstruction and consequent DVT. To date, her lower extremity symptoms improved with anticoagulation, and repeat imaging noted resolution of the thrombosis. She was referred back to her orthopedist for management of her ALVAL. Until the ALVAL obstruction is corrected, she has risk of DVT recurrence and remains on anticoagulation.

REFERENCES

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