Chronic thromboembolic pulmonary hypertension (CTEPH) is defined as pre-capillary pulmonary hypertension (PH) in the presence of chronic thromboembolic disease. CTEPH is the only type of PH that is potentially curable by a surgical procedure: pulmonary thromboendarterectomy (PTE). It is crucial to recognize the importance of early referral of any patient who may have CTEPH, even in the absence of resting pulmonary hypertension, as excellent results can be achieved by reducing exercise-induced PH and restoring pulmonary vascular anatomy.

**Case Report**

The patient is a 28-year-old, previously healthy woman who presented with chest pain and dyspnea on exertion three weeks after a motor vehicle accident and was diagnosed with acute pulmonary embolism. She was anticoagulated with heparin then warfarin, and eventually switched to rivaroxaban due to difficulty maintaining a consistent therapeutic INR on warfarin. Although her chest pain improved, it did not completely resolve, and she had persistent dyspnea with activities of daily living. She also experienced profound fatigue. This persistent dyspnea limited her ability to work at her physically demanding job at the local tree nursery.

Her past medical history was noncontributory. Her family history was positive for deep vein thrombosis in her father. Her coagulation panel showed evidence of lupus anticoagulant.

After her initial evaluation in the pulmonary hypertension clinic, the patient had a screening ventilation/perfusion (V/Q) scan that showed an absence of perfusion in the right middle and lower lobes (Figure 1) and was consistent with filling defects found on CT angiogram (Figure 2). Her echocardiogram showed borderline pulmonary hypertension with an estimated right ventricular systolic pressure 41 mmHg. To confirm the diagnosis, she underwent an exercise right-heart catheterization, which revealed an increase in mean pulmonary artery pressure from 23 mmHg to 39 mmHg at maximal exercise. The patient underwent successful pulmonary thromboendarterectomy and mitral valve repair to remove fibrous vegetations.

Her symptoms have improved postoperatively. Her postoperative echocardiogram showed an estimated pulmonary artery systolic pressure of 28 mmHg. She is currently enrolled in cardiac rehabilitation and improving her exercise capacity weekly. She is maintained on rivaroxaban.

**Discussion**

Pulmonary emboli fail to resolve with anticoagulation in up to 3 percent of patients (1). This case highlights several key points in the presentation, diagnosis, and management of CTEPH. Her presentation was consistent with indolent symptoms in CTEPH (persistent dyspnea on exertion, chest pain, exercise intolerance, and fatigue). Other symptoms include lightheadedness and palpitations. As CTEPH progresses, so do symptoms related to right-heart dysfunction such as edema or syncope (2). While our patient had an insidious progression, people with CTEPH may also experience episodes of acute progression. As for her diagnosis, the first confirmatory study performed was the V/Q scan. This is the test of choice in screening for chronic thromboembolic disease. Although many providers are using a CT angiogram to screen for CTEPH, studies show this test can actually miss chronic disease. It has been reported that CT is not as sensitive for detecting CTEPH as a V/Q scan (51.3 percent sensitivity for CT versus 90–100 percent sensitivity for V/Q) (3). She also had a CT angiogram to define her thromboembolic anatomy for surgical planning. Interestingly, her echocardiogram at rest showed borderline PAH, and due to the fact that she had exercise-induced symptoms, an exercise-induced right-heart catheterization was performed and was crucial in making the diagnosis. Once the diagnosis was made, she was scheduled for the definitive treatment, pulmonary thromboendarterectomy.

She will continue to be followed postoperatively for improvement in her symptoms, and if she has residual pulmonary hypertension, medical therapy will additionally be considered.

**Conclusion**

This case not only highlights the importance of early recognition of CTEPH but also the importance of early referral to a specialist center experienced in the assessment and surgical management of this potentially curative disease. Surgical decision-making involves many factors: comorbid conditions, accessibility of diseased vessels, and the correlation of clot burden to hemodynamic impairment.

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Figure 1. Ventilation/Perfusion (V/Q) scan showing an absence of perfusion in the right, middle, and lower lobes.

Figure 2. CT angiogram showing filling defects.