Chronic thromboembolic pulmonary hypertension – a disease not to be underestimated

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Chronic thromboembolic pulmonary hypertension (CTEPH) is a form of pulmonary hypertension characterised by persistent perfusion defects despite anticoagulation. It is provoked by acute pulmonary embolism that ultimately results in a chronic obstructing microvasculopathy.[1]

An accurate incidence of CTEPH is difficult to quantify as it is frequently underdiagnosed. The incidence after acute pulmonary embolism (PE) is also prone to overestimation. Improving the knowledge and awareness of the risk factors for CTEPH may lead to earlier diagnosis. Untreated, CTEPH is associated with a poor prognosis and substantial risk of mortality. Surgical intervention with pulmonary endarterectomy is potentially curative. However, early diagnosis of CTEPH is a major challenge, with an average diagnostic delay of over a year, resulting in surgical intervention being impossible in up to 40% of patients. This highlights the need for improved practical screening methods.

One of the most common reasons for a delay in diagnosis is the nonspecific clinical presentation of CTEPH. Symptoms of progressive dyspnea and limitations in daily functioning are often misdiagnosed as other cardiopulmonary disease or, often in the elderly population, attributed to age and deconditioning. Guideline recommendations regarding the optimal follow-up of patients with acute PE are lacking.

The greatest challenge is that most patients diagnosed with CTEPH do not have a clear history of a prior pulmonary embolism. Therefore, the current diagnostic rate of CTEPH is likely to be much lower than expected. CTEPH is often identified during diagnostic work-up in patients with unexplained pulmonary hypertension (PH). Durrington *et al*.,[2] recently published the largest study to date on diagnostic rates in patients following an acute episode of PE. They noted a cumulative incidence of 2.1% and that a single follow-up in a PE follow-up clinic at 3 - 4 months after the initial event resulted in identifying patients earlier and with less severe CTEPH. A notable limitation to this study is the significant exclusion of 41% of the cohort due to malignancy or frailty. This may have resulted in a substantial underestimation of the true incidence of CTEPH within the study population.

The study further highlighted the need for a systematic review of patients' computed tomography (CT) scans at the time of pulmonary embolism diagnosis. Twenty percent of patients in their cohort at follow-up may have actually had a diagnosis of CTEPH which was evident on their initial imaging. Features of chronic thromboembolic pulmonary disease (CTEPD) or pulmonary hypertension on initial imaging were reported to be a predictor for the development CTEPH at follow up.

CT signs suggesting chronic thromboembolic pulmonary disease include arterial webs or bands, dilated bronchial arteries, eccentric organised thrombi, total or partial occlusion of vessels and a mosaic parenchymal perfusion pattern.[3] Additional CT signs of pulmonary hypertension include a dilated pulmonary artery trunk ≥30 mm, right ventricle (RV) to left ventricle ratio ≥1 and RV outflow tract hypertrophy ≥6 mm. Evaluation of these is often more difficult than expected and often a lack of experienced radiologists in reporting, limits diagnostics.

In an ideal world, a dedicated PE follow-up clinic will allow for additional benefits in addition to improving CTEPH diagnostics. Decisions can be made on the duration of anticoagulation, patients can be screened for occult malignancy which may have been missed at diagnosis, and patients with pre-existing chronic lung diseases can have optimisation of these conditions with improved quality of life.

With most studies undertaken in first-world settings, the application of screening tools that include expensive imaging tests may be difficult in already resource-constrained health contexts. It is important to remember that the causes for pulmonary hypertension in sub-Saharan Africa are mainly due to left heart disease, lung diseases, tuberculosis, HIV and schistosomiasis.[4] These should not be forgotten in a quest to diagnose a cause for PH with a much lower incidence. Nevertheless, spreading awareness of the importance of screening for symptoms for CTEPH, especially in patients post pulmonary embolus, encouraging collaborative efforts between physicians and radiologists, developing new diagnostic screening methods and conducting inexpensive clinical trials in the field of CTEPH will help improve future diagnostics.

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