Chronic Thromboembolic Pulmonary Hypertension (CTEPH) a Rare Cause of Death in a Woman with Stage 1B Cervical Cancer

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ABSTRACT

Introduction: Chronic thromboembolic pulmonary hypertension (CTEPH) is a rare form of severe pulmonary hypertension. Potentially fatal unless recognised and treated early, it is caused by the chronic occlusion of pulmonary arteries by micro thromboembolic material, ultimately leading to right heart failure. It is typically preceded by an acute thromboembolic event, although this is not always the case. Coupled with the non-specific nature of its symptoms (exertional dyspnoea, fatigue, angina), as well as a lack of awareness amongst the medical community regarding this disease, CTEPH is often misdiagnosed. **Case Description:** A previously well 40-year-old Chinese lady with no history of cardiopulmonary disease was admitted for treatment of a stage IB large cell non-keratinising squamous cell cervical carcinoma. She had an excellent response to radical radiotherapy clinically and radiologically. After the second fraction, she started to experience exertional dyspnoea. A CT pulmonary angiogram (CTPA) showed no evidence of pulmonary embolism (PE), but she was treated empirically for it as well as for a chest infection, and her symptoms improved. Similar episodes occurred intermittently throughout the remaining fractions, however these resolved spontaneously without treatment. Upon successful completion of brachytherapy, she moved overseas, whereby she was reported to have increasingly limited mobility due to persistent exertional dyspnoea (New York Heart Association functional class III – IV). Shortly after her move, she developed an acute attack of shortness of breath and required a hospital admission, where she was investigated for PE yet again. As before, the CTPA returned negative for PE, but she received empirical treatment for a PE and a chest infection. Despite this, she did not improve, and was urgently transferred back to Malaysia for further treatment due to financial reasons. Here, she was reviewed by a pulmonary physician who, based on clinical findings and an echocardiogram, strongly suspected CTEPH. Nonetheless, she deteriorated rapidly, and died of cardiopulmonary failure a few hours later. **Conclusion:** CTEPH is a serious disease requiring early identification and prevention. It is essential that clinicians have a high index of suspicion for CTEPH based on history and clinical examination findings, as there exists potentially curative medical and surgical therapeutic options for CTEPH. Furthermore, it is important to note from this case report that a negative CTPA does not exclude CTEPH. Instead, ventilation/perfusion scintigraphy should be used, as the literature shows that it is the preferred imaging modality for the exclusion of CTEPH.