Massive dilatation of the pulmonary arteries due to acute on chronic thromboembolic pulmonary hypertension

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Case
A 54-year-old woman with a history of multiple pulmonary emboli (PE) and pulmonary hypertension (PH), presented with chest pain and breathlessness. She had stopped taking warfarin 5 days previously. She was tachypneic (24 breaths/min) and tachycardic (124 bpm). ECG revealed right-axis deviation. Chest X-ray (CXR) and CT angiogram of the thorax are shown in Figures 1 and 2. She was diagnosed with an acute PE superimposed on chronic thromboembolic pulmonary hypertension (CTEPH). Enlargement of the right atrium with contrast reflux into the inferior vena cava was also seen on CT.

Discussion
CTEPH (PH WHO Group IV) is the result of incomplete resolution of pulmonary thromboemboli, leading to obstruction of vessels and increased vascular resistance. It is estimated that approximately 3% of patients will present with symptomatic PH one year following an acute pulmonary embolism.

The pathogenesis of CTEPH remains poorly understood. One hypothesis is that persistent obstruction of pulmonary arteries may result in raised pulmonary artery pressures, with development of PH the result of progressive vascular remodelling. Another line of thinking is that pulmonary emboli are the result of a primary arteriopathy of the pulmonary vessels, rather than the initiating event.

The symptoms of CTEPH are non-specific and related to progressive PH, with patients reporting increasing dyspnea on exertion, haemoptysis or signs of right heart failure.

Imaging is important in making the diagnosis. CXR findings may include cardiomegaly, dilatation of the right side of the heart, enlargement of central pulmonary vessels.

Figure 1: CT scout view of thorax (AP) shows enlargement of the proximal pulmonary arteries, (arrow) and cardiomegaly.

Figure 2: CT-Thorax shows enlarged pulmonary arteries (diameters larger than aorta), with eccentric emboli in left pulmonary artery (arrow).
vasculature, pleural effusions, scarring and atelectasis. Echocardiography is often used, with the suggestion that routine echocardiography 6 weeks after pulmonary embolism may help identify patients at risk of developing CTEPH. Diagnostic features of CTEPH on CT scan are as follows:\textsuperscript{5,6}

1. Vascular signs:
   a. Signs of embolism: Complete obstruction, partial filling defects, calcifications, webs
   b. Signs of PH: Asymmetric dilatation of the central pulmonary artery, right ventricular enlargement and hypertrophy, bulging of the ventricular septum, contrast reflux into the inferior vena cava and hepatic veins due to tricuspid regurgitation
   c. Signs of collateral systemic supply: Enlarged bronchial and non-bronchial systemic arteries
2. Lung parenchymal signs: scars, mosaic pattern of attenuation

Patients with persistent PH following an acute pulmonary embolism, despite adequate anticoagulation, have a high risk of mortality from right heart failure if left untreated. Unfortunately, treatment options for patients with CTEPH remain limited. All patients who are potential surgical candidates should be referred to specialised centres for evaluation of the potential for pulmonary endarterectomy. Pulmonary endarterectomy can result in nearly normalised haemodynamics, and substantial improvement in clinical symptoms in selected patients.\textsuperscript{7} Patients with inoperable disease, or with refractory PH after endarterectomy, are treated with pharmacological agents. Riociguat, a soluble guanylate cyclase stimulator, which promotes arterial vasodilatation, is the only pharmacological agent that has been approved in CTEPH. Riociguat has been found to significantly improve exercise capacity and pulmonary vascular resistance in patients with CTEPH.\textsuperscript{8,9}

**Competing interests:** XXXX

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