Chronic Thromboembolic Pulmonary Hypertension-Diagnosed on Spiral CT Angiography

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Abstract

Chronic thromboembolic pulmonary hypertension (CTEPH) is a part of the spectrum of venous thromboembolism in which pulmonary thrombus fails to resolve, resulting in occlusion of the major pulmonary artery. Diagnosis of this disease is important as it is potentially curable by pulmonary thromboendarterectomy. A case of CTEPH diagnosed non-invasively on spiral computerized (CT) pulmonary angiography is reported.

INTRODUCTION

Chronic thromboembolic pulmonary hypertension (CTEPH) is more common than generally recognized.1-3 CTEPH is a part of the spectrum of venous thromboembolism in which pulmonary thrombus fails to resolve. Patients usually remain asymptomatic despite extensive pulmonary artery occlusive disease. Eventually all these patients deteriorate and develop severe right heart failure with limited exercise capacity.2

Diagnosis of CTEPH is challenging and is often delayed because of the non-specificity of symptoms and signs. However, diagnosis of this disease is important as it is potentially curable by pulmonary thromboendarterectomy.4 We report a case of CTEPH diagnosed non-invasively on spiral computerized (CT) pulmonary angiography.

CASE REPORT

A 46 years male, non-smoker, presented with history of exertional dyspnoea and dry cough since 12 years, orthopnoea, oedema feet and oliguria since three months. There was no history of chest pain or haemoptysis. Patient had been diagnosed as hypertensive one year back and was under treatment with calcium channel blockers.

Examination revealed presence of bilateral pedal oedema, raised jugular venous pressure, reduced breath sounds over left lower lobe, and loud pulmonary component of second heart sound. There was polycythemia with hemoglobin of 18.9 gm%. Liver functions were slightly deranged but renal functions were normal. Chest radiograph showed presence of cardiomegaly with left sided pleural effusion (Fig. 1). Spirometry revealed a restrictive abnormality. Electrocardiogram showed P-pulmonale with right axis deviation. Arterial blood gas analysis (ABG) revealed hypoxaemia (PaO₂ - 59 mm of Hg) with respiratory alkalosis. 2D-echocardiography (2D ECHO) revealed severe pulmonary hypertension with a pulmonary artery pressure of 106 mm of Hg, right atrial and right ventricular dilatation, small pericardial effusion and normal left ventricular ejection fraction. Venous Doppler of both lower limbs was normal. Ventilation-perfusion (V/Q) scan of the lung showed multiple mismatched ventilation-perfusion defects (Fig. 2). Spiral computerized tomography (CT) angiography with three dimensional virtual reconstruction (3DVRT) imaging showed thromboembolism involving the left pulmonary artery and its branches with an infarct in the left lower lobe. A thrombus was also seen in the right pulmonary artery (Fig. 3). However, the main pulmonary
artery was normal. A diagnosis of CTEPH was made and the patient was referred for surgical pulmonary thromboendarterectomy.

**DISCUSSION**

In CTEPH, pulmonary emboli do not completely resolve instead they sometimes get recanalised and are endothelialised resulting in pulmonary artery obstruction. Thus, CTEPH is caused by occlusion of multiple major pulmonary arteries due to “organized thrombus”. Progression of pulmonary artery thrombosis from in situ thrombus propagation causes marked increase in pulmonary vascular resistance, pulmonary hypertension and eventually severe right heart failure. Impaired endogenous fibrinolysis is thought to be an underlying etiology. Familial thrombophilia (anti-thrombin III, protein C or protein S deficiency) is uncommon among CTEPH patients. Acquired thrombophilia due to a lupus like anticoagulant has been reported in approximately 10% of CTEPH patients. Patients with CTEPH have non-specific complaints such as exertional dyspnoea, chronic fatigue and excessive daytime sleepiness and occasionally chest pain or haemoptysis. Thus, the spectrum may vary from asymptomatic patients to those with severe pulmonary hypertension and right heart failure.

Patients with confirmed CTEPH have at least a segmental or large mismatched ventilation-perfusion defect. CT scan of the chest may show parenchymal abnormalities such as mosaic attenuation, asymmetric artery size and pulmonary infarcts. Pulmonary angiography showing pulmonary artery narrowing or “cut-offs”, bands, webs, pouches or vessel irregularity is diagnostic. Spiral CT pulmonary angiography may obviate the need for invasive pulmonary angiography as in our case.

Spiral computed tomographic (CT) angiography of the pulmonary circulation has emerged recently as a potential useful diagnostic method for the evaluation of the pulmonary circulation. As a non-invasive procedure, it has progressively replaced conventional and digital pulmonary angiography as the standard diagnostic imaging modality of the pulmonary circulation.

Medical measures and thrombolytic agents can neither reverse the effects of CTEPH nor prevent its progression. Diuresis may reduce preload; hydralazine, and nifedipine, may be used to reduce pulmonary vascular resistance. Pulmonary thromboendarterectomy is the treatment of choice. The criteria for considering a patient for surgery include: 1) segmental or more proximal pulmonary artery thrombus, 2) pulmonary vascular resistance >300 dynes-s-cm⁻⁵ and 3) absence of co-morbid disease.

The diagnosis of CTEPH should be considered in all patients with unexplained pulmonary hypertension and right heart failure. Spiral CT pulmonary angiography can be used to confirm the disease non-invasively.

**REFERENCES**