Case 1: Chronic Thromboembolic Pulmonary Hypertension (CTEPH)

Abstract:

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Case Report

A 32 year-old man was referred to the pulmonary hypertension unit with increasing dyspnoea and multiple recent syncopal episodes. He had suffered a pulmonary embolism at the age of 17 and had received an appropriate course of warfarin therapy. Afterwards, he had remained well and in full-time employment until a recent presentation to his local hospital with increasing dyspnoea and syncope occurring episodically over a period of a few months. A computed tomographic pulmonary angiogram (CTPA) performed there was reported as showing evidence of pulmonary embolus.

On review at the pulmonary hypertension unit, he had oxygen saturations of 92% on room air, a raised jugular venous pulse to his earlobe, an S3 gallop and grade 2-3 pitting oedema. He was admitted immediately for further work up. As part of his evaluation he had an ECG, a ventilation/perfusion (V/Q) scan and a further CTPA study (shown). He also had a right heart catherisation that demonstrated pulmonary arterial pressures of 104/46 mmHg, with a mean pulmonary arterial pressure of 65 mmHg, wedge pressure of 5 mmHg and cardiac output of 2 litres/min. Thrombophilia screen was performed and was negative. He was commenced on bosentan, warfarin and diuretics. His 6-minute walk test had improved from 100 m to 305 m prior to discharge.

Figure 1: Axial CT of the thorax below the level of the carina shows a mosaic attenuation pattern (sharply demarcated areas of regional variation in attenuation of the lung parenchyma) that can be seen in the setting of chronic thromboembolic disease. Note the pruning and small calibre of the vessels in the regions of decreased attenuation (i.e. dark areas of the lung).

Figure 2: Axial CT demonstrates enlargement of the pulmonary trunk and pulmonary arteries as well as chronic mural thrombus in the right main pulmonary artery (arrow). Distally, there is a marked calibre reduction in a second order pulmonary artery to the right lower lobe.

Discussion

Pulmonary hypertension is defined as an abnormal elevation of pressure in the pulmonary circulation. The fourth World Symposium on Pulmonary Hypertension most recently updated the clinical classification system for pulmonary hypertension in 2008, called the Dana Point classification system[1]. Irrespective of the underlying aetiology, an elevated mean pulmonary arterial pressure above 25 mmHg is considered abnormal. The term â pulmonary arterial hypertensionâ (PAH) should be reserved for those patients with a haemodynamic profile in which elevated pre-capillary pulmonary resistance causes high pulmonary pressure in the setting of normal venous pressure. PAH can be idiopathic, heritable, drug/toxin induced, associated with connective tissue diseases, HIV infection, portal hypertension, congenital heart disease, schistosomiasis, chronic haemolytic anaemia and persistent pulmonary hypertension in newborns. Other groups in the classification system include pulmonary venous occlusive disease (PVOD) and/or pulmonary capillary haemangiomatosis (PCH), pulmonary hypertension due to left heart disease, pulmonary hypertension due to chronic lung diseases/hypoxaemia, chronic thromboembolic pulmonary hypertension (CTEPH) and pulmonary hypertension with unclear multifactorial mechanisms including haematological, systemic and metabolic disorders.

Chronic thromboembolic pulmonary hypertension (CTEPH) is an important cause of severe pulmonary hypertension usually occurring as a long-term complication of pulmonary embolism. Previously the condition was thought to be rare, but recent studies have estimated that 3.8% of patients with a pulmonary embolus may subsequently develop CTEPH[2]. The severity of the disease depends on the degree of the underlying associated pulmonary hypertension and right heart dysfunction, but overall mortality is considerable. Further non-invasive imaging investigations include ECG, echocardiography, multi detector CT pulmonary angiography (including dual-energy CT) and cardiac magnetic resonance imaging (MRI). Signs of right heart strain on ECG and evidence of right heart chamber enlargement or an increase in tricuspid regurgitation velocities on ECHO are a cause for concern. The V/Q scan typically shows evidence of multiple areas of ventilation-perfusion mismatch, with a low probability scan effectively out ruling the condition. While CTPA findings include parenchymal and vascular signs that are highly suggestive of the cause[3], MRI gives functional assessment of the heart and in particular the right ventricle. CTPA may also be used to assess for any surgical benefit. Recent articles in the literature have described in detail the imaging features of pulmonary hypertension[4], as well as novel CT imaging techniques[5]. Thrombophilia screening should be performed if the cause of the pulmonary emboli is idiopathic. Of note, less than10% of CTEPH patients have an underlying thrombophilia.

When the bulk of the disease process occurs in the proximal vessels, pulmonary endarterectomy may be possible. Although associated with significant (5-10%) mortality surgery is the treatment of choice when available due to the excellent long-term outcomes. Where surgery is not a viable option, heart-lung transplantation and standard pulmonary hypertension medications have been utilised with varying degrees ofsuccess[6-8]. The patient discussed here was referred for pulmonary thromboendarterectomy.

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