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CASE REPORT

Chronic Thromboembolic Pulmonary Hypertension

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ABSTRACT

The characteristic imaging features of chronic thromboembolic pulmonary hypertension are described. This is a very rare disorder; prompt surgical treatment affords good results. The complementary use of the ventilation/perfusion scan and CT pulmonary angiogram in diagnosis, and exclusion of other differential diagnoses, are emphasised.

Key Words: *Computed tomography , Hypertension, pulmonary, Pulmonary thromboembolism, Thromboendarterectomy, Ventilation/perfusion scan*

INTRODUCTION

Chronic thromboembolic pulmonary hypertension (CTEPH) is a very rare disorder, with an incidence of less than 0.5% of all thromboembolic diseases.[1] The majority of patients have no predisposing factors.[2] Dyspnoea on exertion may be the only symptom in the early stage.[3] Typical clinical features of acute pulmonary embolism are usually absent in patients with CTEPH.[3] The diagnosis is made by ventilation/perfusion (V/Q) scan and enhanced CT of the thorax. A preoperative pulmonary angiogram may be necessary.[4] Pulmonary thromboendarterectomy provides good treatment results.[5]

CASE REPORT

A young woman aged 16 years with previously good past health presented with flu-like symptoms in February 2000. Over the next 10 months, she developed progressive exertional dyspnoea, without symptoms suggestive of acute pulmonary embolism. Her chest radiograph at first presentation showed clear lung parenchyma except for a diminutive right hilum (Figure 1). Lung volumes were normal bilaterally. The electrocardiogram (ECG) showed a pattern of right heart strain. An echocardiogram was performed to discern evidence of congenital heart disease, but showed pulmonary hypertension only, with an estimated pressure of about 70 mm Hg (her expected normal pressure limit was 20 to 30 mm Hg). She was initially thought to have idiopathic or primary pulmonary arterial hypertension, and a transoesophageal echocardiogram

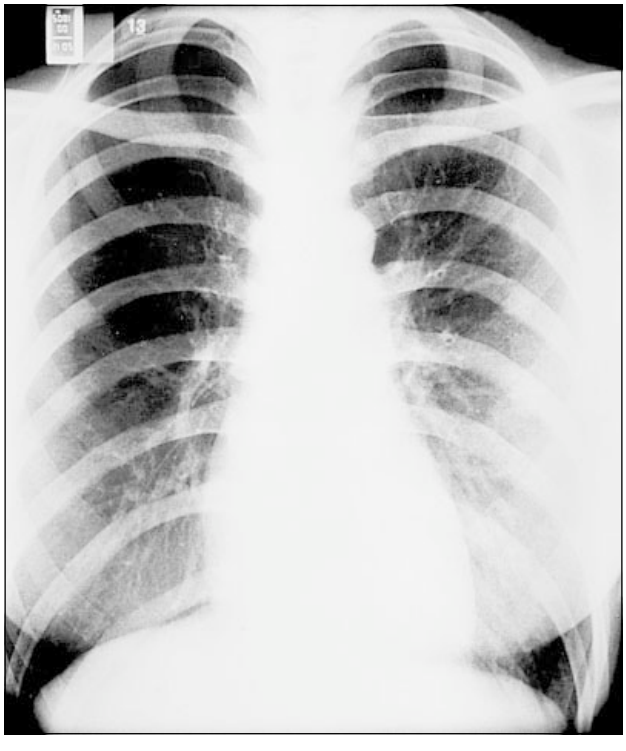

Figure 1. Chest radiography at presentation showed clear lung parenchyma, a diminutive right hilum, and hypoperfusion of the right lung.

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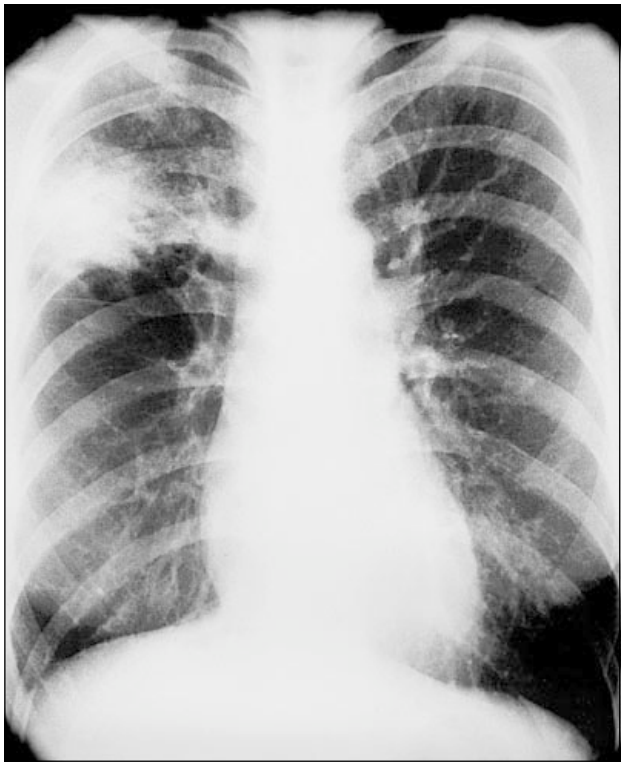

Figure 2. Chest radiography 10 months postpresentation showed a new opacity, with an air bronchogram in the right upper lobe compatible with a pulmonary infarct.

was performed to exclude thromboembolism. This did not detect any thrombus in the main or in the left proximal pulmonary arteries, while the proximal right pulmonary artery could not be clearly seen. Nevertheless, she was started on warfarin in December 2000, since thromboembolism could not be totally excluded. Her complete blood picture, liver and renal function tests, blood gas analysis and coagulation profiles were normal, but proteins C and S, antiphospholipid antibody, and antithrombin III were not checked before warfarin administration. She was later assessed by a cardiologist and a V/Q scan was requested to exclude thromboembolic disease. The chest radiograph taken for the V/Q scan in January 2001 showed a new parenchymal opacity with an air bronchogram in the right upper lobe in addition to the previous findings (Figure 2). Expiratory chest radiography was also performed, but did not show any air trapping such as would occur in Swyer-James/Macleod's syndrome (SJMS).

The V/Q scan showed global hypoperfusion to the right lung, with a triple match defect of absent perfusion in the posterior segment of the right upper lobe (Figure 3). In addition, large segmental mismatch perfusion defects were seen in the superior and inferior lingular segments of left lung. The ventilation study was normal

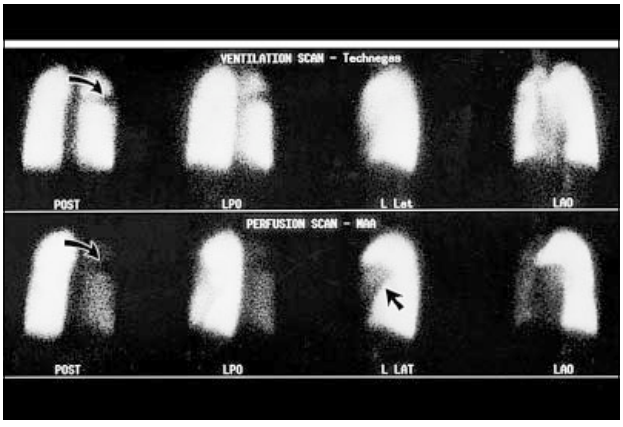

Figure 3. Ventilation/perfusion scan showed hypoperfusion of the entire right lung, in addition to the right upper lobe absent-perfusion triple match defect (curved arrow) corresponding to the pulmonary infarct. The lingular segment of the left lung showed mismatch perfusion defects (arrow). The corresponding ventilation scan was normal except for the right upper lobe triple match defect. The scan was interpreted as a high probability of pulmonary embolism according to PIOPED[6] criteria.

except for the right upper lobe triple match defect. The scan was interpreted as indicating a high probability of pulmonary embolism by revised Prospective Investigation of Pulmonary Embolism Diagnosis (PIOPED) criteria.[6] The V/Q scan clearly demonstrated bilateral perfusion abnormalities.

Enhanced CT of the thorax was performed after the V/Q scan to visualise an obstructive thrombus in the right proximal pulmonary artery. A protocol for imaging of pulmonary embolism was adopted (Iopamiro 370® [iopamidol] 100 mL + 20 mL water at an injection rate of 3 mL/second; 15 seconds delay for helical CT scanning, with 3 mm collimation at hilar level and 10 mm collimation for the rest of the thorax). The CT thorax showed tapering of the proximal right pulmonary artery (Figure 4). Although there was artefact from the hyperdense contrast medium within the superior vena cava, the thrombus could still be discerned in the posterior wall of the proximal right pulmonary artery. The distal branches of the right pulmonary artery were markedly attenuated. The main pulmonary trunk was larger than the adjacent ascending aorta, with pruning of the left pulmonary artery, reflecting changes of pulmonary arterial hypertension. No thrombus was seen in the pulmonary arterial branch of the lingular segment of the left lung. Mosaic perfusion seen in the lung window was noted in the left lung, with hypoperfusion in the lingular segment and relative hyperperfusion of the left lower lobe. Collateral circulation from the hypertrophied right bronchial artery and intercostal arteries was noted. The right upper lobe

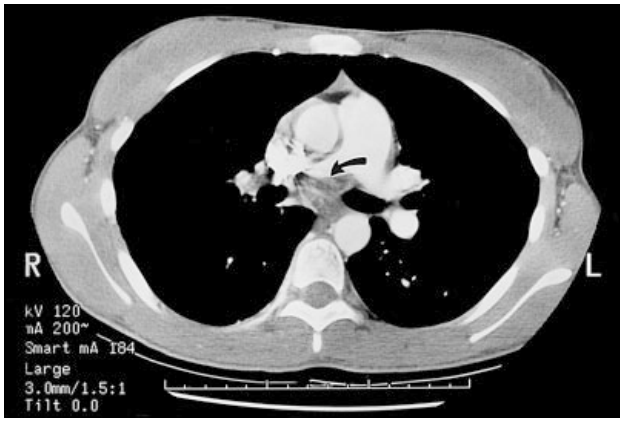

Figure 4. Enhanced CT scan of the thorax showed a thrombus (curved arrow) in the posterior wall of the right pulmonary artery. The main pulmonary trunk is larger than the adjacent ascending thoracic aorta. The prominent subcarinal soft tissue is merely due to the normal oesophagus. No mediastinal pathology causing compression of the pulmonary trunk was noted.

consolidation showed cavitation, compatible with a resolving pulmonary infarct. The airways were normal.

A conventional pulmonary angiogram showed changes which reflected the findings of the perfusion lung scan. There was tapering of the proximal right pulmonary artery, and hypoperfusion of the right lung, with occlusion of the lingular branch of the left pulmonary artery (Figure 5). The measured pulmonary trunk pressure was 66 mm Hg. An aortogram was not performed.

The patient was initially scheduled for pulmonary thromboendarterectomy. However, her pulmonary arterial pressure decreased from 70 mm Hg initially to 50 mm Hg after 5 months' warfarin treatment, and her symptoms improved significantly. Therefore, surgery was withheld and the patient was observed on anticoagulation therapy.

DISCUSSION

Less than 0.5% of all patients with thromboembolic disease have an unresolved thrombus, leading to CTEPH. Patients of all ages may be affected, except for the first decade of life. The symptoms of patients suffering from CTEPH are usually non-specific, and patients may only present with progressive exertional dyspnoea. Chest radiographs of patients with CTEPH usually show enlarged hila and pruning of pulmonary vasculature. Occasionally, a unilateral diminished hilum is seen.[2]

There are several differential diagnoses, of which the most important is primary pulmonary hypertension

Figure 5. A pulmonary digital subtraction angiogram showed tapering of the right pulmonary artery (arrow), and occlusion of the lingular branch of the left pulmonary artery (curved arrow).

(PPH). Although thrombus in situ, as seen in PPH, may be difficult to differentiate from thromboembolism in CTEPH,[7] the pulmonary arteries in patients with PPH are not absent or asymmetrical in size, as in patients with CTEPH.[2] A normal or low probability of pulmonary embolism on V/Q scan is typical for patients with PPH, while multiple mismatched perfusion defects characterise CTEPH.[2] Interruption of the proximal right pulmonary artery[8] or congenital agenesis of the right pulmonary artery[9] are usually associated with unilateral small lung volume, and cannot explain the bilateral perfusion abnormalities seen in the V/Q scan of our patient. Furthermore, the absence of air trapping on the expiratory chest radiograph or bronchiectasis on CT, and the clinical presence of pulmonary hypertension in our patient already excluded SJMS. She had no clinical or biochemical evidence of vasculitis. Coagulatory disorders such as the presence of lupus anticoagulants, deficiencies in antithrombin III, and proteins C and S occur in only a minority of patients with CTEPH.[2]

Ventilation/perfusion scan is regarded as the most sensitive and non-invasive test to provide evidence that pulmonary hypertension is related to chronic thromboembolism.[3] V/Q scan typically shows a high

probability of pulmonary embolism in CTEPH but a low probability in PPH.[10] However, there is some overlap between patients with CTEPH and those with PPH, as thrombosis in situ in severe pulmonary hypertension of any cause cannot usually be distinguished from pulmonary embolism.[7] It may be difficult to distinguish CTEPH from a vasculitis such as Takayasu's arteritis, as the V/Q scan may be of high probability in both conditions.[11] However, Takayasu's arteritis can be excluded on clinical grounds, and from its characteristic angiographic findings.[12]

Helical CT is a useful alternative to conventional pulmonary angiography for the diagnosis of chronic thromboembolism, and segmental vessel disease is also more accurately determined with a CT scan than with MRI.[2,4] The two features that most reliably indicate CTEPH are mosaic attenuation and asymmetric size of the segmental vessels.[13] In patients with CTEPH, variation in segmental pulmonary arterial diameter is due to chronic multifocal vascular obstruction, in which thromboembolic material becomes organised or fibrotic, with subsequent attenuation of some vessels but compensatory hypertrophy of others.[14] Other CT features in CTEPH include bronchial and intercostal arterial collaterals, and pulmonary infarction.[2,4,14]

MR angiography has depicted the absence and truncation of segmental arteries in CTEPH, as well as abnormally large peripheral vessels, presumably due to bronchial collaterals supplying parenchyma distal to pulmonary emboli.[2]

CT and V/Q scans are complementary diagnostic tests for CTEPH. Pulmonary angiography is necessary to select the best candidate for pulmonary thromboendarterectomy.[3] The central thrombus may sometimes be difficult to discern in angiography, as the thromboembolic material is incorporated concentrically in the vessel walls, with new epithelium smoothing the inner contour.[4] Fortunately, CT can depict central thromboembolic material clearly.[4]

Chronic thromboembolic pulmonary hypertension is ideally treated with pulmonary thromboendarterectomy.[5] The thromboembolic material, the intima, and a variable portion of the media are removed with the endarterectomy specimen; thus, the procedure is not simply embolectomy. Most patients with CTEPH who undergo thromboendarterectomy improve clinically in terms of gaseous exchange and pulmonary haemodynamics.[3] When all surgical procedures are contraindicated, medical therapies such as long term anticoagulants, diuretics, vasodilators, inotropes, and oxygen therapy remain other options, but medical treatment is not considered curative.[5]

In conclusion, CTEPH is a rare but treatable disorder, in which early diagnosis can be achieved with a high index of clinical suspicion and the use of helical CT and V/Q scans.

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