Chronic Thromboembolic Pulmonary Hypertension

K. Gowrinath¹ and K. Padmanabha Kamath²

Departments of Tuberculosis and Respiratory Diseases¹ and Cardiology,² Kasturba Medical College, Manipal, India

ABSTRACT

Chronic thromboembolic pulmonary hypertension (CTEPH) is an uncommon consequence of acute pulmonary embolism. We report CTEPH in a 58-year-old male who had pleurisy with a small haemorrhagic pleural effusion three months ago. The six-month course of anti-coagulation therapy failed to resolve thromboemboli completely or improve pulmonary hypertension. Computed tomographic pulmonary angiography (CTPA) is useful for the diagnosis as well as for the follow-up of chronic pulmonary thromboembolism. [Indian J Chest Dis Allied Sci 2009;51:45-47]

Key words: Thromboembolic pulmonary hypertension, Pulmonary embolism, Computed tomographic pulmonary angiography, Pulmonary thromboembolism.

INTRODUCTION

Chronic thromboembolic pulmonary hypertension (CTEPH) is an infrequent complication of incomplete resolution of large pulmonary thromboemboli.¹ If the embolic material within major pulmonary arteries obstructs the blood flow significant enough to increase the right ventricular afterload, pulmonary hypertension may develop after a symptom free period ranging from months to years.² Early diagnosis of CTEPH is usually not possible as a history of symptomatic acute pulmonary embolism may not be evident in two-thirds of patients with CTEPH.³ We report a case of CTEPH following a mild illness of pleurisy with effusion.

CASE REPORT

A 58-year-old male was admitted with exertional breathlessness, chest pain and apprehension of six weeks duration. Chest pain was retrosternal, aggravated by minimal activity and he was unable to walk for a few yards because of breathlessness. There was no history of orthopnoea or wheezing. Three months ago, he had developed pleurisy, a small right-sided haemorrhagic pleural effusion and peripheral blood eosinophilia which resolved within a week after receiving diethylcarbamazine citrate. His cardiac function was normal and he was feeling well. He was an ex-smoker. Physical examination showed elevated jugular venous pressure. He had varicose veins in both legs. His radial pulse rate was 84 beats per minute, respiratory rate was 22 breaths per minute, blood pressure was 130/80 mmHg and oral temperature was 98.5 °F. Respiratory system examination revealed continuous flow murmur over lower lobes. On cardiovascular system examination, left parasternal heave and loud second heart sound at pulmonary area were evident. Rest of the physical examination. He had platelet count of 268,000/mm³ with a normal blood chemistry. Arterial blood gases were within normal limits. Chest radiograph (Figure 1) showed a prominent right hilum.

Electrocardiogram (ECG) showed normal sinus rhythm, right atrial enlargement, right ventricular hypertrophy and non-specific T-wave changes in the

Figure 1. Chest radiograph (PA view) showing prominent right hilum (white arrow).

[Received: January 8, 2007; accepted after revision: April 24, 2007]

Correspondence and reprint requests: Dr K. Gowrinath, Department of Tuberculosis and Respiratory Diseases, Kasturba Medical College, Manipal-576 104 (Karnataka), India; Phone: 91-0820-2922294; Fax: 91-0820-2571934; E-mail: drkgowrinath@gmail.com
interior leads. Computed tomography (CT) of the chest (Figure 2) showed mosaic attenuation of lung parenchyma; CT pulmonary angiography (CTPA) (Figure 3) showed eccentric filling defect in the right main pulmonary artery and pouch like filling defect in the left main pulmonary artery (Figure 4). Doppler ultrasound study of lower limbs was normal.

Anticardiolipin antibodies and lupus anticoagulant were not detected. Antithrombin III, protein S and protein C levels were normal. Transthoracic echocardiogram showed moderate to severe pulmonary arterial hypertension (gradient 78 mmHg), severe tricuspid regurgitation and normal left ventricular systolic function. There was no significant clinical improvement despite six months of warfarin along with initial seven days course of enoxaparin. At the end of six months of anti-coagulation therapy, transthoracic echocardiogram still showed pulmonary arterial hypertension (gradient 73 mmHg) and features of cor-pulmonale. The CTPA revealed residual filling defect in the right main pulmonary artery. He was referred for pulmonary thromboendarterectomy but he did not undergo surgery and has been taking oral warfarin. He still had pulmonary hypertension (gradient 77/mmHg) at the end of one year follow-up.

DISCUSSION

Following the first episode of angiographically proven symptomatic pulmonary embolism, the incidence of CTEPH was about one percent after a minimum follow-up period of three years. History suggestive of an embolic event like pleurisy, peripheral oedema, haemoptysis, etc, can be elicited in most patients with CTEPH. In our case, CTEPH followed pleurisy with a small haemorrhagic pleural effusion which was probably the acute embolic event. The pathophysiologic events leading to CTEPH are not known completely and the only factor that has been linked to CTEPH is presence of anticardiolipin antibodies which were detected in 10% to 24% patients. In our case, anticardiolipin antibodies were negative.

Patients with CTEPH manifest symptoms similar to those seen in pulmonary hypertension, the most common being exertional breathlessness; physical signs of right heart failure may also be evident. Occasionally a continuous flow murmur due to turbulent blood flow through larger pulmonary arteries partially occluded by the thrombi can be heard over the lower lobes. This finding is usually absent in primary pulmonary arterial hypertension. We used CT of the chest as this imaging technique has been found to be a useful alternative to angiogram. Central vessel emboli are more accurately detected with helical CT than with angiogram. The CT features of pulmonary hypertension secondary to chronic pulmonary thromboembolism include mosaic attenuation of parenchyma due to irregular perfusion, eccentric filling defects representing organised thrombi adjacent to the vessel wall, enlarged central pulmonary arteries (often greater in diameter than aorta) or asymmetry in the size of central pulmonary artery. In our case, mosaic

![Figure 2. CT of the chest showing mosaic attenuation of lung parenchyma.](image)

![Figure 3. CTPA showing eccentric filling defect in right pulmonary artery (white arrow).](image)

![Figure 4. CTPA showing pouch like filling defect in left main pulmonary artery (white arrow).](image)
attenuation of lung parenchyma, eccentric filling defect within the right main pulmonary artery and enlarged pulmonary trunk were indicative of CTEPH. For early detection of pulmonary hypertension, echocardiography has been suggested routinely at six weeks after acute pulmonary embolism. Early detection of CTEPH was not possible in our case, as acute pulmonary embolism was not suspected in view of mild illness and normal cardiac function. In CTEPH, life-long anticoagulation is required for prevention of recurrent thromboembolic events and adequate anticoagulation therapy may still fail to stop its progression. Prognosis without treatment is poor in cases of CTEPH; the two-year survival was only 30% in cases where pulmonary arterial hypertension was above 30 mmHg. The treatment of choice for CTEPH is surgical pulmonary endarterectomy which leads to permanent improvement in pulmonary hypertension.

REFERENCES