Case Report

Chronic Thromboembolic Pulmonary Hypertension

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Abstract

The case of a 65 years old lady, diabetic, hypertensive, reduced functional class with history of episodic dyspnoea lasting for few weeks for the last two years. On presentation she was tachypnoiec with reduced oxygen saturation. Jugular venous distension was present with bibasal crackles. Her ECG was normal. Echocardiogram revealed, dilated right sided chambers with right ventricular systolic dysfunction and severe pulmonary hypertension. A CT chest was performed which showed multiple thrombi in the main and branch pulmonary arteries establishing the diagnosis of chronic thromboembolic pulmonary hypertension.

Introduction

Chronic pulmonary hypertension is a relatively rare complication of pulmonary embolism but is associated with considerable morbidity and mortality.[1] It is commonly believed that symptoms manifest only several years after the initial episode of pulmonary embolism. Pulmonary embolism may be recurrent. However, the true frequency is estimated at 0.1 percent among patients who survive a pulmonary embolism. Timing is not well established, although patients with limited activities or bed bound are at higher risk.[2]

Our patient was predisposed to thromboembolism secondary to reduced daily activities. Although her workup for hypercoaguable state was negative and there was no evidence of deep venous thrombosis on ultrasound doppler, computed tomography (CT) was consistent with diagnosis of chronic thromboembolism resulting in severe pulmonary hypertension.[3]

Case Report

A 65 years old female diagnosed diabetic, hypertensive with limited functional activity due to knee osteoarthritis a had history of episodic dyspnoea lasting for 2-3 weeks in last 2 years. She had been treated by general practioners but never had a complete work up.

She presented in the emergency department with history of worsening dyspnoea for the last 2 weeks, which persisted even at rest. On presentation she was tachypnoeic with oxygen saturation of 88%, at room air. Her BP was 190/90 mmHg and heart rate 92/beats per minute. Jugular venous distension was present with bilateral basal crackles. She was admitted with a provisional diagnosis of hypertensive heart failure.

The laboratory work up showed haemoglobin of 13g/dl, total leukocyte count of 13 x 10^9, platelet counts of 283 x 10^9 Renal, liver functions, cardiac enzymes and coagulation were within normal ranges. Blood gases on room air revealed pH 7.46 with Pco_2 34.6, Po_2 59mm of Hg, bicarbonate 24mmol and oxygen saturation of 88%.

Chest radiograph showed prominent bronchovascular markings with upper lobe diversion. ECG was within normal limits. She responded to intravenous nitrates and diuretics with significant symptomatic improvement.

Echocardiogram showed normal left atrium and ventricular dimensions; preserved left ventricular systolic function with ejection fraction of 60%. Right sided chambers were dilated with right ventricular systolic dysfunction, moderate tricuspid regurgitation was present and calculated pulmonary artery systolic pressures was 65 to 70 mm of Hg (Figure 1).

As we were considering preserved left ventricular systolic function and dilated right chambers, she underwent a CT of chest with contrast which showed bilateral filling defects at the branching of major pulmonary arteries extending into segmental arteries with partial canalization indicating pulmonary emboli (Figures 2 and 3).

Work up for hypercoaguable state revealed, normal anti thrombin III, protein C and S levels. D-dimer was raised. Ultrasound Doppler legs was negative for evidence of deep venous thrombosis.

A diagnosis of chronic thrombo-embolic pulmonary hypertension (CTPH) was made and she was started on long term anticoagulation therapy.

Discussion

Pulmonary hypertension results from numerous causes. Thromboembolic obstruction of pulmonary arteries is a potentially correctable cause of pulmonary hypertension. Most patients with chronic thromboembolic pulmonary hypertension (CTPH) presents late in the course of the disease, making it difficult to ascertain the natural history of the disease.[4] Thromboembolic basis of pulmonary hypertension has been questioned, current evidence supports this cause regardless of documented history of acute venous

thromboembolism.[3] This is not surprising because recent studies indicate that pulmonary embolism can occur without symptoms and that symptomatic pulmonary embolism is often overlooked or misdiagnosed.[5] Patients with thromboembolic disease may remain asymptomatic for months or years. The pathophysiological events in the progression of pulmonary hypertension during this period have not been well defined.[6]

As with other forms of pulmonary hypertension, progressive exertional dyspnoea and exercise intolerance are characteristic of thromboembolic pulmonary hypertension. The symptoms are often attributed to other cardiopulmonary disorders, deconditioning, or even psychogenic dyspnoea.[7] As the disease progresses, the signs and symptoms of right ventricular failure became evident.[5]

Once diagnosed the approach would be to determine the severity of pulmonary hypertension, to determine its cause as thromboembolic Pulmonary Hypertension can be amenable to surgical correction. Findings on standard laboratory tests are nonspecific. Duplex scanning of the legs reveals evidence of venous thrombosis in 35 to 45% of patients with CTPH. Transthoracic echocardiography is usually the first study to

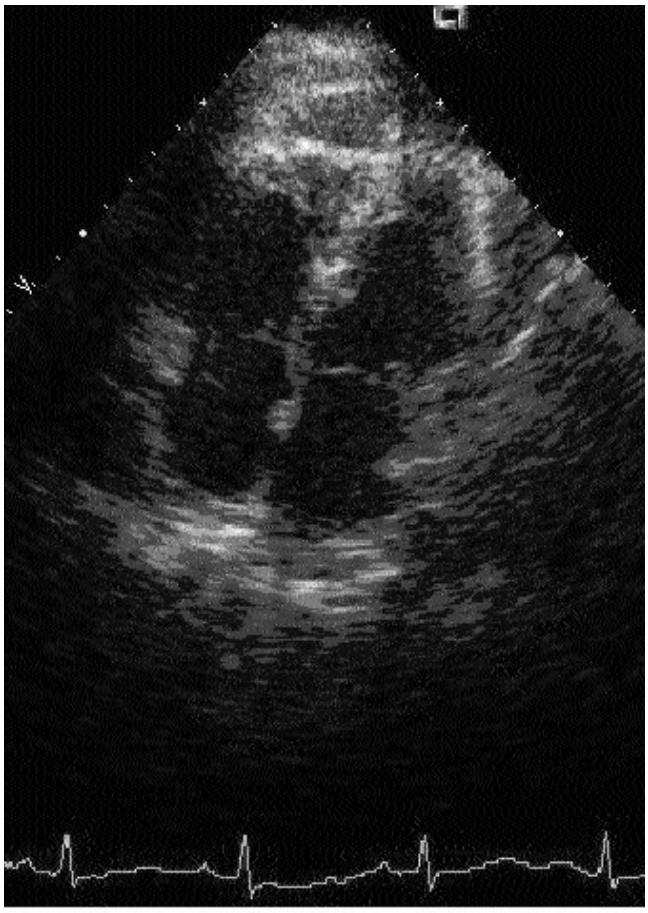

Figure 1: Transthoracic echocardiogram, apical four chamber view showing dilated right atrium and right ventricle.

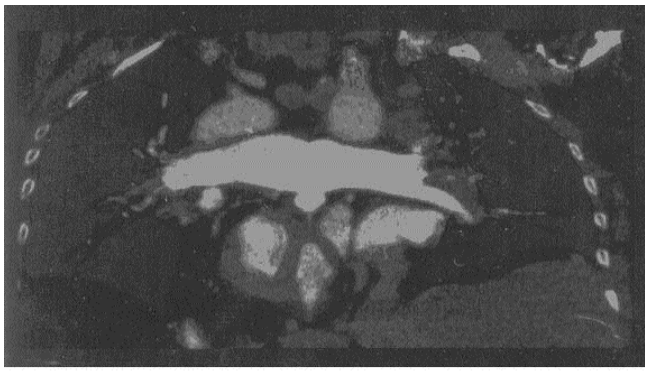

Fig 2: Contrast computed tomogram of chest showing presence of filling defect in left main pulmonary artery.

suggest an abnormality of right heart due to pulmonary hypertension.[8] Computed tomography studies do not provide essential haemodynamic data but they are particularly useful in the evaluation of the main pulmonary arteries. Computed tomography provides extent of disease and in cases of chronic thromboembolism shows recanalization due to partial embolus resolution. High-resolution CT (HRCT) of the lung shows a mosaic pattern in CTPH that is virtually diagnostic.[9]

Pulmonary thromboendarterectomy is considered in symptomatic patients who have haemodynamic or ventilatory impairment at rest or with exercise. Distal thrombi are not amenable to thromboendarterectomy with current techniques.[10] Those patients who are at high risk or in whom the disease is beyond central arteries medical treatment options for pulmonary hypertension including treating the source of embolism if evident, anticoagulation, vasodilators like calcium channel antagonists, prostaglandin analogue, bosantin, sildenifil and supplemental oxygen therapy can be tried.

Our case highlights that chronic thromboembolic

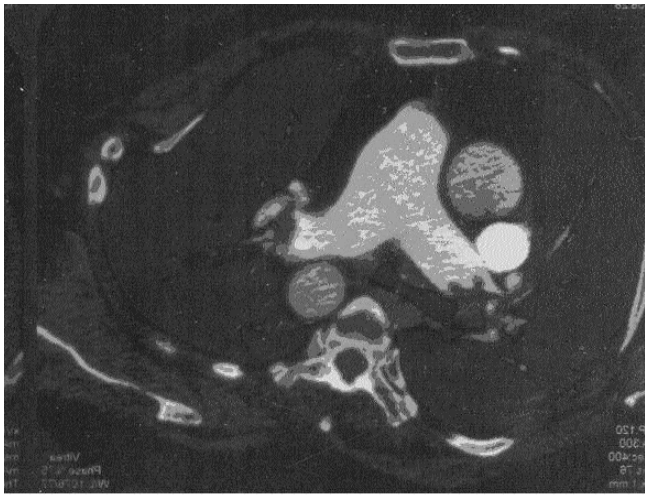

Fig 3: Contrast computed tomogram of chest showing presence of filling defect in major pulmonary arteries extending into segment branches.

pulmonary hypertension which is considered as a disease of west, could be found in our population as well. Our local data is very sparse about this etiology. This case also demonstrates that symptoms are often vague and ultrasound doppler for deep venous thrombosis may be negative in these patients so a great deal of clinical suspicion is needed for precise diagnosis.

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