Case Report

Primary Pulmonary Hypertension: A Case Report

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Abstract

Primary pulmonary hypertension (PPH) is a rare disease which usually presents with shortness of breath. We report a case of a female patient who presented with dyspnoea. We diagnosed her as a case of PPH on the basis of detailed history, careful physical examination and common investigations like electrocardiogram (ECG), chest radiography and Doppler echocardiograhy.

Key words: *Pulmonary hypertension, Primary*

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Introduction

Primary pulmonary hypertension currently known as Idiopathic Pulmonary Arterial Hypertension (IPAH) is a rare disease with an incidence of about 2–3 per million per year and a prevalence of 15 per million. Adult females are almost three times more likely to present with IPAH than adult males.[1]

Pulmonary circulation has an extensive surface area of about 50–70 m^2 at rest. It is normally a high flow, low pressure and low resistance system which can accommodate marked increase in cardiac output without any significant increase in pressure. However, with abnormal pulmonary vasculature, pressure rise can approach up to systemic levels.

Pulmonary hypertension (PH) is defined as mean pulmonary artery pressure (PAP) of >25 mm Hg at rest and >30 mm Hg during exercise[2] and pulmonary arterial hypertension (PAH) is diagnosed when pulmonary capillary wedge pressure (PCWP) or left atrial pressure is <15 mm Hg in presence of PH.[3] We report a patient who had all features of severe PH including exertional dyspnoea, right ventricular heave, loud pulmonary component of second heart sound (P_2) with pulmonary regurgitation (PR), tricuspid regurgitation (TR), with unremarkable chest findings. Although more definitive tests including cardiac catheterisation could not be done due to limited facilities and financial constraints but detailed history, careful examination, chest radiography, ECG and Doppler echocardiography strongly suggest the case as PPH. The objective of reporting the case is to create awareness of this life threatening condition so that PPH is considered when usual causes for common symptoms like shortness of breath are clinically excluded.

Case Report

A 35-year-old, nonsmoker, nondiabetic, normotensive lady presented to us with shortness of breath and palpitation for one year in May 2011. She also complained of compressive chest pain on exertion, but no cough, wheeze, sputum, haemoptysis or leg swelling. She had no remarkable drug history. She had no history of human immunodeficiency virus (HIV) infection, chronic liver disease (CLD), deep vein thrombosis or connective tissue disease. She lives on

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plain land, has normal sleep-pattern and did not suffer from rheumatic fever or bronchial asthma.

She is a housewife of a lower-middle class family having two children. She never worked in or lived near any industry and was not exposed to any noxious substance known to cause interstitial lung disease (ILD). Her family history in this regard was unremarkable.

On physical examination, she was neither cyanosed nor clubbed, but breathless at rest with respiratory rate 24 breaths/min, pulse rate 96 beats/min in regular rhythm, blood pressure (BP) 120/70 mm Hg. Her skin condition and joints were found normal and there were no stigmata of chronic liver disease or any pedal oedema.

Her jugular venous pressure (JVP) was not raised, but there was left parasternal heave. Her first heart sound was normal, but P_2 was very loud in pulmonary area and there was early diastolic murmur along left sternal border and a systolic murmur in tricuspid area. There was no other murmur.

Respiratory examination was unremarkable with normal vesicular breath sound with normal expiratory phase, no rhonchi or wheeze and there was no crepitation over lung bases. Abdominal, musculo-skeletal and other systemic examinations revealed no abnormalities.

Complete blood count (CBC), electrolytes, renal and liver function tests (LFT) were normal, rheumatoid factor and antinuclear antibody (ANA) were negative. Chest radiography (Fig 1) revealed cardiomegaly, prominent pulmonary arteries and oligaemic lung fields. ECG (Fig 2) showed normal sinus rhythm, right axis deviation, right atrial enlargement and right ventricular hypertrophy. Spirometry done with difficulty showed Forced Vital Capacity (FVC) 2.89 L (68% of predicted value), Forced Vital Capacity in 1 second (FEV_1) 2.08 L (69% of predicted value), FEV_1/FVC 71.9% and no bronchodilator reversibility.

Transthoracic Doppler echocardiogram showed dilated right atrium and right ventricle. Interatrial and interventricular septa were intact with no patent ductus arteriosus flow. Mitral and aortic valves were normal. Pulmonary valve motion was suggestive of severe PH. Left ventricular cavity was reduced with normal wall thickness and wall motion with ejection fraction 55%. There was no pericardial effusion, no vegetation or thrombus. Doppler study showed grade II tricuspid regurgitation and grade 1 pulmonary regurgitaion. Pulmonary artery systolic pressure was 85 mm Hg and diastolic pressure 30 mm Hg.

Specialized tests including ventilation perfusion scan (V/Q scan), cardiac catheterisation, sleep study could not be done due to limited resources. After initial assessment she was given furosemide and sildenafil and was advised to avoid hot bath, exertion, extra salt, and pregnancy. She remained stable for about six months before being lost from our follow-up.

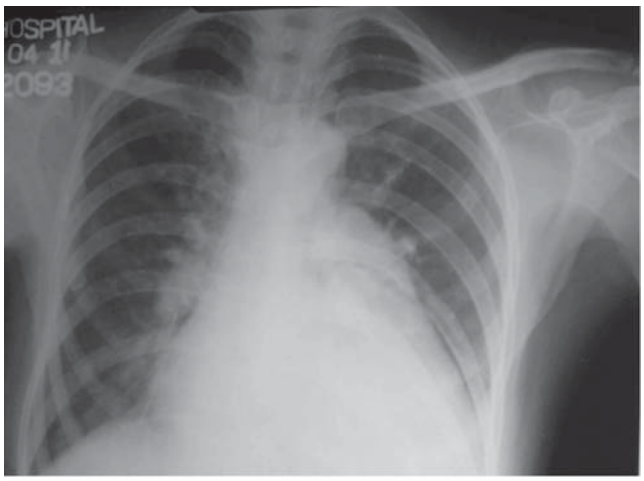

Fig 1. Chest radiograph showing cardiomegaly

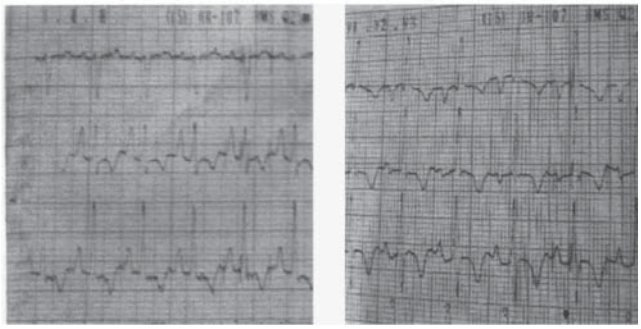

Fig 2. Electrocardoigram

Discussion

PAH is diagnosed when PAP is >25 mm Hg at rest and >30mm Hg during exercise with PCWP < 15 mm Hg. The term 'Primary pulmonary hypertension' was first used in 1951 to describe the clinical feature and haemodynamic abnormality of 39 patients.[4]

Although traditionally, PH was divided into primary and secondary types, the third world conference on PH in 2003 classified PH into five major categories with further subdivisions depending on pathogenesis.[5]

Patients with PPH can present with varied cardiopulmonary symptoms similar to PH from other causes. Sometimes PH is asymptomatic and may be diagnosed by incidental echocardiographic finding in early stage.

Clinical examination reveals features suggestive of RVH and/or RVF. Careful examination helps to exclude congenital or acquired heart disease as well as pulmonary pathology like interstitial lung disease, chronic obstructive lung disease etc.

ECG and chest radiography reveal features of RVH and PH respectively. Echocardiography is helpful in confirming the diagnosis of PH as well as excluding the cardiac lesions as the etiology of PH. Both systolic and diastolic pulmonary artery pressures can be estimated precisely by Doppler study. Pulmonary function test is done to evaluate possible obstructive or restrictive lung disease. V/Q scan is recommended to evaluate chronic pulmonary thromboembolisn though pulmonary angiography is the definitive test in this regard. Full sleep study is helpful to exclude patients having sleep apnoea. Cardiac catheterisation is required in most patients with PAH to confirm the diagnosis, assess its severity, guide medical therapy and provide prognostic information. Diagnosis of PAH requires the presence of PH with two other conditions – pulmonary capillary wedge pressure must be less than 15 mmHg and pulmonary vascular resistance must be greater than 3 wood units and these measurements can only be done by right sided Swan-Ganz catheter. All haemodynamic data are obtained at baseline as well as after giving a short acting pulmonary vasodilator, eg, nitric oxide. A positive vasodilator response is defined as a decrease of at least 10 mmHg mean PAP with no change in cardiac output and no fall in BP. This group of patients are candidates for calcium channel blocker.[3,6,7]

Management

Treatment of PH secondary to other causes focuses on the treatment of underlying disease. Management of PPH can be discussed under the following headings:

Life style change

Hypoxia is a potent pulmonary vasoconstrictor leading to increase in afterload worsening PH, so all activities, eg, heavy exertion, hot bath should be avoided. Smoking and excess salt should be forbidden.

Recommendation for women of child bearing age

Although successful pregnancy outcomes have been reported in patients with PH, early termination of pregnancy is recommended in view of high mortality of up to 50% .[6,8]

Immunization

Influenza and pneumococcal vaccination is strongly recommended to prevent respiratory infections.

Medical therapy

Traditional therapy includes anticoagulation based on improved survival data from two retrospective studies as well as evidence of microscopic in situ thrombosis.[9] Diuretics are used for RVF, but their effects on mortality in PAH are not known.[10,11] Digoxin is used in RVF with atrial fibrillation but has not been studied extensively in PH patients.[10,12] Oxygen supplementation is recommended in patient who is hypoxic (PaO_2<55 or SaO_2 <80% at rest).[10,13]

Pulmonary vasodilator therapies include long term use of calcium channel blocker which has shown good result in only 5% patients with positive vasodilator response.[14] Prostanoids, eg, epoprostenol have short half lives and are used only for severely ill patients.[3,6,15]

Endothelin receptor antagonists, eg, bosentan prevent endothelial proliferation and vasoconstriction and have been approved by FDA.[16] Phosphodiesterase 5 inhibitors, eg, sildenafil which prevent degradation of cyclic guanosine monophosphate (GMP), have recently been approved for PAH by US FDA.[17]

Interventional and surgical therapy

Atrial septostomy involves creating shunt between right and left atria to decompress the failing right heart and is largely seen as a bridge to lung transplant.[18] Lung transplan-

tation gives patients median survival of five years[19] and remains an option for patients who deteriorate despite best medical therapy.

Prognosis

Overall untreated median survival of PPH patients is 2–3 years from time of diagnosis with usual cause of death being RVF, but outcomes have changed over the last two decades because of newer drug therapy.[20]

Conclusion

Although PPH is a rare disease but its grievous nature demands active consideration and investigation of the condition when common causes are excluded by physical examination and simple investigations in a patient presenting with symptoms like shortness of breath. Further trials and research regarding treatment of this condition is imperative to save patients from early death.

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