A CASE OF IDIOPATHIC PULMONARY ARTERIAL HYPERTENSION IN MALE

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ABSTRACT

Primary Pulmonary Hypertension is a rare disease occurring in 1-2 per million population. It is 2-4 times more common in female. Idiopathic or primary pulmonary hypertension is defined as a disorder with no identifiable cause in which resting mean pulmonary artery pressure in adults is above 25 mmHg and 30 mmHg with exercise. Idiopathic or primary pulmonary hypertension is diagnosed after ruling out all the possible secondary causes of pulmonary hypertension. We are presenting a case of middle-aged male who presented with dyspnoea and on further evaluation found to have primary pulmonary hypertension, which is uncommon in male.

KEYWORDS
Primary Pulmonary Hypertension, Dyspnoea.

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CASE HISTORY

A 39-year-old male admitted with history of exertional dyspnoea, NYHA class II since one month and syncope I episode on the day of admission. He is not diabetic or hypertensive. No past history of coronary artery disease, COPD, bronchial asthma, pulmonary tuberculosis or any other illness. He is not a smoker or alcoholic. No familial history of any chronic illness or sudden death. No history of chronic drug intake or sleep disturbance.

On examination, patient was dyspnoeic and tachypnoeic. He was not pale, not icteric; no cyanosis, no clubbing and no bilateral pitting pedal oedema. JVP was elevated and pressure was 12 cm of blood. Pulse 86/min, normal volume, regular rhythm and BP - 100/70 mmHg. On examination of cardiovascular system, P2 was palpable, parasternal heave of grade 2 was present, epigastric pulsations were noted. On auscultation second heart sound was loud in pulmonary area with narrow split, ejection systolic murmur heard in the pulmonary area. Mitral, tricuspid and aortic areas S1, S2 heard normally. No other murmur heard. Respiratory system, Gastrointestinal system and Central Nervous System was clinically normal. Clinical features suggested pulmonary hypertension, but the aetiology could not be determined clinically. Patient was further investigated.

Complete blood count was normal, urea 36 mg, creatinine 0.9 mg, serum electrolytes were within normal limits.

Chest X-ray PA view showed dilated main pulmonary artery with cardiomegaly.

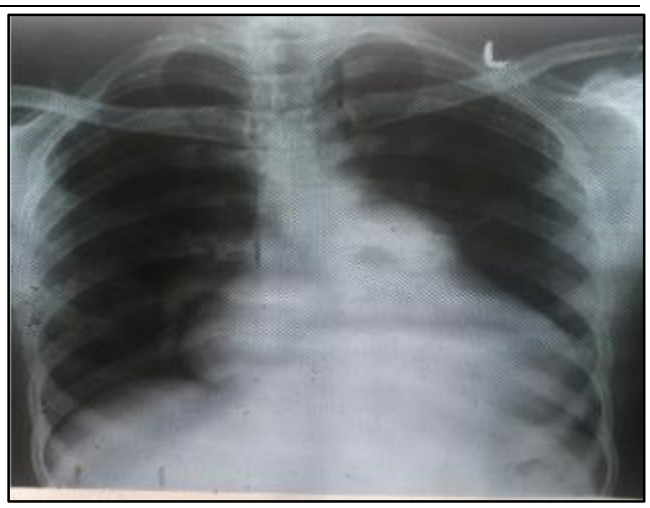

Fig. 1: Chest X-ray PA View showed Dilated Main Pulmonary Artery with Cardiomegaly

ECG showed Right Axis Deviation, Right Atrial Enlargement and Right Ventricular Hypertrophy

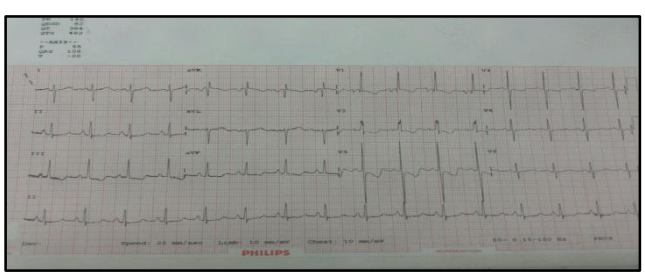

Fig. 2: ECG showing Right Axis Deviation, Right Atrial Enlargement and Right Ventricular Hypertrophy

Echo showed right atrium and right ventricular enlargement, D-shaped left ventricle, severe pulmonary hypertension, main pulmonary artery – 35 mm, right pulmonary artery – 24 mm, left pulmonary artery – 23 mm, pulmonary artery mean pressure of 62 mmHg, TRPG – 84 mmHg, TAPSE 17 mm, PR end diastole 34 mmHg. Mild TR and PR. No evidence of congenital or valvular heart disease with normal left ventricular function and ejection fraction 62%.

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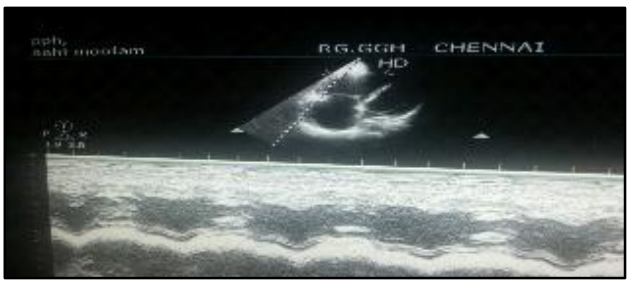

Fig. 3: Echo showing the Relevant Findings

As we could not make out any cardiac causes of pulmonary hypertension by echocardiography, patient was further investigated to rule out other causes of pulmonary hypertension as per diagnostic algorithm. LFT was normal. HBsAg, Anti HCV were negative, HIV negative and ultrasound abdomen was normal. On further evaluation ANA, C-ANCA, Anti-cardiolipin Antibody, Anti-phospholipid Antibody, Anti-topoisomerase Antibody, Rheumatoid factor all were negative. Thyroid function test and pulmonary function test were also normal.

CT Chest showed Normal Study of Lung with Dilated Right Heart and Dilated Main Pulmonary Artery

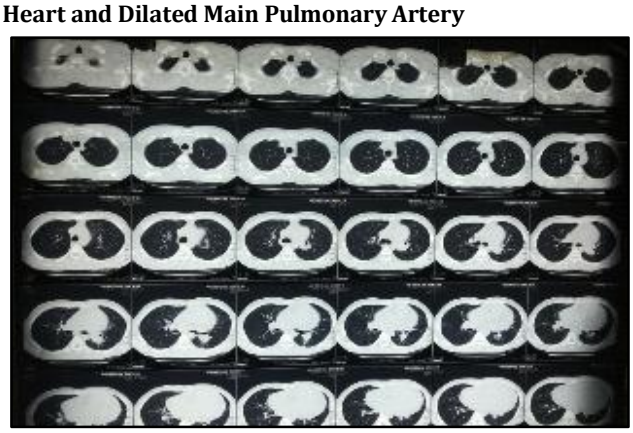

Fig. 4: CT Chest with Relevant Finding

In Doppler study of Lower Limbs, there was no evidence of DVT.

CT Pulmonary Angiogram showed Dilated Pulmonary Artery Measuring 3.9 cm with no Evidence of Thrombosis in the Pulmonary Vasculature

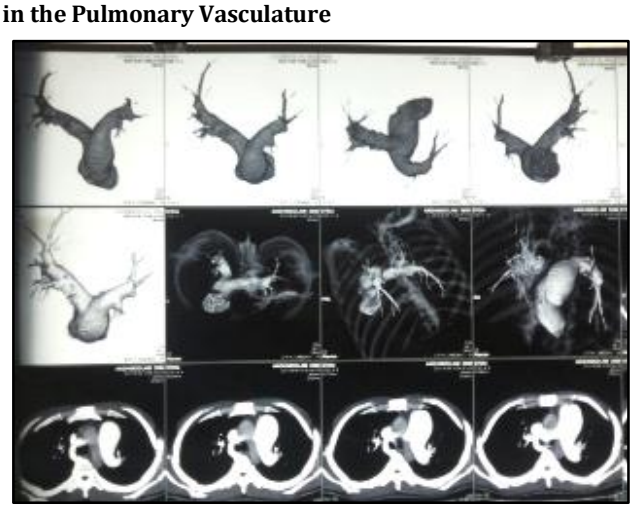

Fig. 5: CT Pulmonary Angiogram showing Relevant Findings

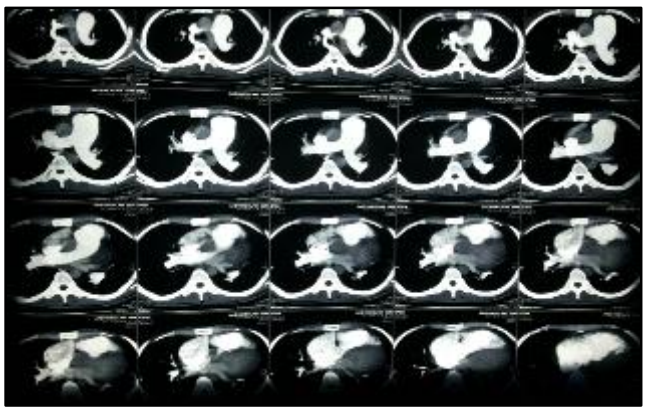

Fig. 6: CT Pulmonary Angiogram showing Relevant Findings

Cardiac catheterization showed severe pulmonary hypertension, no shunt with normal LV function, main pulmonary artery pressure –114/40 mmHg with mean 64 mmHg. There is no vasoreactivity.

DISCUSSION

Idiopathic or primary pulmonary artery hypertension is a condition characterized by sustained elevation of pulmonary artery pressure without an identifiable cause.[1] Diagnostic criteria includes mean pulmonary artery pressure of more than 25 mmHg at rest and more than 30 mmHg on exercise.[2]

Although, the diagnosis of idiopathic pulmonary artery hypertension is one of the exclusion, it can be made with accuracy by excluding all the secondary causes. A detailed history and physical examination as well as appropriate tests should be performed to uncover causative and contributing factors. Idiopathic or primary pulmonary hypertension is 2-4 times more common in middle aged females than males.[2] But our case is the male patient with features of pulmonary hypertension, which is the rare incidence.

The factors which play a role in pathogenesis of idiopathic pulmonary hypertension are vasoconstriction, vascular wall remodelling and thrombosis in situ. The significance of vasoconstriction in pathogenesis was substantiated by vasodilatory response to acetylcholine. Altered pulmonary vascular endothelial function is also important. An imbalance between prostacyclin and thromboxane contribute to pathogenesis. Impaired synthesis of endothelium derived nitric oxide synthesis and enhanced production of endothelium-derived vasoconstrictor response is also associated with pulmonary hypertension.

The most common symptom of idiopathic pulmonary artery hypertension is dyspnoea, which is the initial symptom in 60% of the patients and syncope in 8% of patients.[3] Our patient also presented with dyspnoea and syncope. The physical findings of idiopathic pulmonary artery hypertension are typical of pulmonary hypertension like palpable and loud p2, ejection systolic murmur in pulmonary area which is present in our patient. In severe cases early diastolic murmur in pulmonary area and tricuspid regurgitation murmur can be heard. Patient may have pedal oedema in case of right ventricular dysfunction.

DIAGNOSIS AND MANAGEMENT

In National Institute of Health Registry on PPH, an algorithm was developed listing essential tests to exclude secondary

causes.[3] This starts from echocardiogram to rule out valvular, congenital heart disease and cardiomyopathy. Then it includes pulmonary function testing, HRCT Chest and CT pulmonary angiogram to rule out pulmonary parenchyma and thromboembolic diseases. Lower limb Doppler is needed to rule out deep vein thrombosis. Serological tests are necessary to rule out connective tissue disorders. Thyroid function tests, Retroviral tests are also needed. Ultrasonogram helps to rule out portal hypertension. Finally, cardiac catheterisation is an absolute requirement for confirming the diagnosis and for guiding treatment based on vasoreactivity.

After diagnosis without treatment, idiopathic pulmonary arterial hypertension progress to death in most of the cases due to right heart failure. Many promising newer therapies like prostacyclin analogue, endothelin receptor antagonist, phosphodiesterase inhibitors improve clinical function and haemodynamic measures. These will halt the progression and may prolong survival. Anticoagulation is generally indicated in all the patients with idiopathic pulmonary arterial hypertension.[1]

CONCLUSION

In our patient all the necessary investigations were done to rule out secondary causes of pulmonary hypertension, which were negative. Since all the possible secondary causes are ruled out, the diagnosis of idiopathic pulmonary artery hypertension is made in this middle aged male patient. Then patient was started on sildenafil 25 mg tds, bosentan 62.5 mg bd along with warfarin 2 mg. Patient improved clinically and he was followed up periodically.

REFERENCES

1. Fauci AS, Longo DL. Harrison's principles of internal medicine. 18th edn. Vol 2. McGraw Hill, New York, U.S.A. Chapter no. 250, 2012:2076-82.
2. Perloff, Joseph K. Clinical recognition of congenital heart disease. 6th edn. Philadelphia U.S.A. Elsevier, chapter no. 14, 2012:pp 196.
3. Rubin LJ. Primary pulmonary hypertension. New England Journal Of Medicine 1997;336(2):111-7.