INTRODUCTION

Primary pulmonary hypertension (PPH) is a rare autosomal dominant disorder,1-2 and accounts for about 10% of total cases while secondary pulmonary hypertension is more common.3 Pulmonary circulation is a low resistance vascular bed. In pulmonary hypertension, pulmonary arterial bed resistance increases due to vascular constriction and leads to decreased oxygenation of blood in lungs resulting in generalized hypoxemia.

Hypoxia is major stimulus to release angiogenic factors like hypoxia induced factors -1a (HIF – 1a) and vascular endothelial growth factor (VEGF) in ischemic tissue.4 These are the transcription factors when released in response to hypoxia stimulate angiogenesis (new vessel formation) in ischemic tissues, including the heart, brain and retina.5

This case report describes a lady with retinal neovascularization induced by PPA.

CASE REPORT

A female patient 45 years of age, presented with complaints of gradually worsening shortness of breath on exertion and dry cough for last three years. She also complained of swelling on feet and gradual decrease in vision in both eyes for last one year.

Examination revealed fine crackles at lung bases, signs of right ventricular failure (right parasternal heave, peripheral edema and enlarged liver) and pansystolic murmur at tricuspid area. Examination of eye revealed vision 20/200 bilaterally. On slit lamp examination cornea anterior chamber, lens and iris were all normal. Fundoscopic examination revealed neovascularization of retina and this was further confirmed by fluorescence angiography. Fundus florescence angiography did not show engorgement or sheathing of the blood vessels but showed abnormal vessels in the temporal part of the posterior fundus involving the macular area (Figure 1).

DISCUSSION

Pulmonary hypertension is defined as pulmonary artery pressure > 25 mmHg at rest and > 30 mmHg with
Primary pulmonary hypertension has familial form which is autosomal dominant disease, which occurs in mid 30s with female predominance. It results from idiopathic medial hypertrophy of vessel, intimal proliferation and thrombosis in pulmonary vascular bed. When it occurs it leads to decreased oxygenation of blood in lungs and ultimately causes generalized hypoxemia and right sided heart failure. Hypoxia is major stimulus to release angiogenic factors like hypoxia induced factors -1a (HIF – 1a) and vascular endothelial growth factor (VEGF) in ischemic tissue. These are the transcription factors when released in response to hypoxia, stimulate angiogenesis (new vessel formation) in ischemic tissues, like heart, brain and retina. In the eye, vascular development is stringently regulated by co-ordinated expression of ‘vascular endothelial growth factor’ (VEGF) which is a potent angiogenic factor and ‘pigment epithelium derived factor’ (PEDF) a potent angiostatic factor. These angiogenic regulators are expressed in reciprocal fashion, so that in hypoxia VEGF expression is enhanced while PEDF expression is suppressed, leading to retinal neovascularization resulting in loss of vision.

The pharmacological treatment available to improve the symptoms of primary pulmonary hypertension are, Sildenafil, Prostacyclins analogue and Bosentan (Endothelin receptor blocker).

This neovascularization process can be prevented by agents that block vascular endothelial growth factor (VEGF) and hypoxia inducible factor 1a (HIF – 1a), which is still under trial, and so far there is no therapy available to prevent this neovascularization process.

In this particular case however, the control of primary pathology including hypoxia led to regression of the symptoms. So measures controlling the hypoxia primarily, may be employed to arrest the neovascularization process.

The ultimate treatment for primary pulmonary hypertension is heart lung transplantation otherwise prognosis is poor in these patients.

REFERENCES


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