Successful outcome of pregnancy following use of Phosphodiestrase- 5 inhibitor in severe pulmonary arterial hypertension

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Introduction

Pulmonary artery hypertension (PAH) is a progressive disease that preferentially affects women of child bearing age and is characterized by an elevated pulmonary vascular resistance often leading to right ventricular failure and death if untreated[1,2]. Historically, patients with pulmonary artery hypertension have been described to experience adverse outcomes as physiologic demands of pregnancy and parturition can precipitate right ventricular failure leading to maternal or fetal complications. Maternal and fetal mortality of up to 11% to 50% have been described in the literature[3,4]. Consequently, expert consensus guidelines strongly recommend that women with PAH, who are of childbearing potential, need to use an effective contraception to avoid pregnancy, and in the event of pregnancy, early pregnancy termination is recommended[5].

Recent advances in diagnosis and medical management of patients with PAH have led to improvement in symptoms, exercise tolerance and survival[6,7]. There are reports in the literature describing the use of PAH-specific medical therapies in pregnant patients with PAH suggest that successful outcomes are possible with careful peripartum administration of calcium channel blockers, prostacyclin analogues, and/or phosphodiesterase inhibitors[8,9,10]. Here, we are reporting an interesting case of severe PAH with pregnancy that had a successful maternal and fetal outcome.

Case report

A woman aged 26 years, in 30th week of her third pregnancy reported to us with complaints of shortness of breath for past 10 days and swelling all over body for past 20 days. She had history of dyspnoea on exertion off and on since childhood but for past 10 days she was dyspnoic at rest also. She had noticed swelling on feet 20 days

back which gradually increased and involved gradually whole of her body. She had congenital torticollis and scoliosis. There was no history of blurring of vision, headache or epigastric pain. There was no personal or family history of thromboembolism. There was no history of smoking. Her first pregnancy was 2 years back and antenatal period was uneventful. She had a full term delivery at home and baby expired after 3 days, cause of death is not known. She had second pregnancy one year back, resulting in spontaneous abortion at 5^{th} month of pregnancy which was uneventful. On physical examination, her pulse rate was 92/min, blood pressure was 140/80 mm Hg, respiratory rate was 36/min, and jugular venous pressure was not raised. On abdominal examination uterus was 24 weeks size, ascites was present, no hepatospleenomegaly. On cardiovascular examination, systolic murmur was present in mitral and pulmonary area; para-sternal heave was present. There were no signs of deep venous thrombosis. Haematological and biochemical indices were normal apart from a mildly raised white cell count consistent with pregnancy (14 X 10^9/L, neutrophils 78%, lymphocytes 15%,eosinophils 2%, monocytes 5%). 12-lead electrocardiogram showed right axis deviation and right ventricular hypertrophy and strain. Chest radiography showed proximal enlargement of pulmonary vasculature and peripheral vascular pruning. Transthoracic echocardiography showed grossly dilated right atrium and right ventricle, RV hypertrophy, Pulmonary artery enlarged, mild to moderate eccentric MR , Right ventricular systolic pressure= 40mm Hg with atrial septal defect. Patient was diagnosed as severe pulmonary artery hypertension.

She was kept in intensive cardiac care unit and put on broad spectrum antibiotics, Inj. Lasix 20mg iv 12hrly, tab Digoxin 0.25mg OD (5 days a week), syp. Potchlor 2tsf BD, nebulisation with ipratropium bromide and tab Sildenafil 12.5mg twice a day. Her condition improved and she went into preterm labour at 32 weeks of gestation and delivered a baby girl of 1.1kg birth weight.

Discussion

Since the widespread commercial availability of PAH-specific therapy (1997-2007) there has been a decline in pregnancy

associated maternal mortality[10]. Still a maternal mortality of 16.7% has been reported in literature[11]. Many a time's pregnancy termination is morally objectionable to both the patient and her family and the clinician may be faced with a clinical situation in which patient's wishes to continue pregnancy like in our case. Several management principles that include development of a multidisciplinary team, early counseling regarding the risks of continued pregnancy, institution of fluid restriction and diuretic use along with salt avoidance , early use of PAH-specific therapies, scheduled Cesarean section, and continued postpartum treatment are key to successful maternal and fetal outcome. During labour and delivery, continuous monitoring of ECG, pulse oximetry, and intraarterial BP should be routine. While pulmonary artery catheterization may be of benefit in selected cases, there is no consensus regarding its routine use[12,13,14]. Regarding the manner of delivery,frequent use of planned cesarean section has been described. While vaginal delivery has been associated with fewer bleeding complications and infection, cesarean section avoids prolonged labour and allows for careful preparation of anaesthesia, optimization of hemodynamics, and development of contingency plans. Moreover, cesarean section does not appear to negatively influence patient outcomes[11].

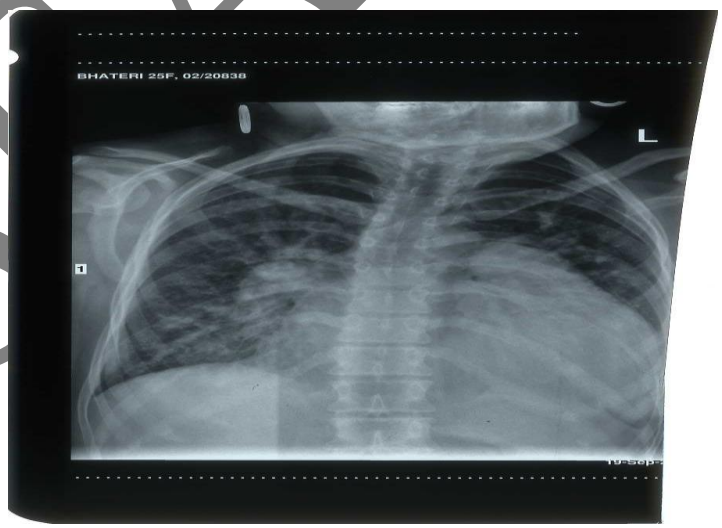

Figure 1: Chest radiograph of the patient showing scoliosis and cardiomegaly

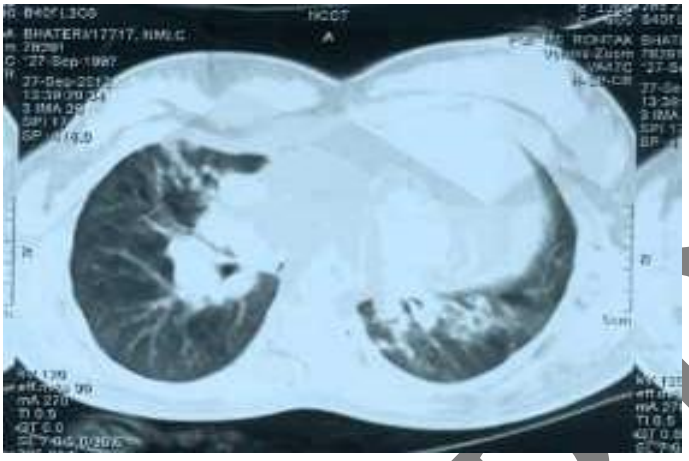

Figure 2: CT chest showing marked cardiomegaly and prominent pulmonary trunk and right and left pulmonary arteries

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