Management of Severe Pulmonary Hypertension in a Parturient with a Recent Diagnosis of Systemic Lupus Erythematosus

Abstract Type: Case Report/Case Series
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High mortality rates are associated with pulmonary arterial hypertension (PAH) in pregnancy (30%). Mortality in pregnant patients with PAH associated with connective tissue diseases is as high as 55% (1). Increased blood volume and cardiac output in pregnancy can result in changes in pulmonary vascular tone due to large volume shifts, hypoxemia, and thromboembolism. A 24 year old G2P1 parturient with history of Raynaud’s disease presented at 28 weeks with new onset dyspnea on exertion. Transthoracic echocardiography revealed systolic pulmonary artery pressures (PAP) of 90 mmHg with right ventricular dilation. Right heart catheterization (RHC) showed mean PAP of 53 mmHg. The patient was started on epoprostenol therapy.

Anticoagulation with low molecular weight heparin was initiated, despite a low platelet count (80,000 cells/mm3) attributed to gestational thrombocytopenia. Subsequent workup for the etiology of PAH revealed positive antineutrophil antibody and antibodies to double stranded DNA consistent with a diagnosis of systemic lupus erythematosus (SLE). Immunosuppression therapy was not initiated acutely and the patient remained hospitalized.

A repeat RHC, one month later, revealed improved mean PAP of 40 mmHg. Suddenly, the patient was noted to become increasingly dyspneic, tachycardic and hypoxic. An urgent repeat cesarean section (CS) and tubal ligation was performed under general anesthesia (GA) at 32 weeks with arterial, central venous, and pulmonary artery catheters and continuous non-invasive stroke volume measurement (Vigileo monitor). A transesophageal echocardiogram probe was used intraoperatively. After delivery, an intravenous milrinone infusion and inhaled nitric oxide were initiated for systolic PAP in excess of 100 mmHg. The patient remained mechanically ventilated and was transferred to the intensive care unit. On postoperative day 1, she developed significant uterine bleeding requiring blood transfusions that eventually resolved with oxytocin and uterine massage. Mechanical ventilation was discontinued on postoperative day 2. Inhaled nitric oxide and intravenous milrinone were stopped despite elevated systolic PAP of 100mmHg. The epoprostenol infusion was increased and sildenafil was initiated with a noted decrease in systolic PAP.

Maternal outcomes with CS compared to vaginal delivery have been worse likely due to the associated hemodynamic instability and neither regional nor GA have proven to be superior. A successful approach includes multimodal vasodilator therapy, anticoagulation, and GA with both invasive and non-invasive cardiovascular monitoring for CS. Despite the fact that vasodilator therapy for parturients with PAH due to SLE may improve outcomes, mortality still remains high.