Management and Cesarean Delivery of a Parturient with Recurrent Primary Pulmonary Artery Sarcoma and Severe Pulmonary Hypertension

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Introduction

Pulmonary artery sarcoma (PAS) is an extremely rare tumor from the mesenchymal cells of the pulmonary artery that is frequently asymptomatic until significant pulmonary artery obstruction and pulmonary hypertension occur (1,2). This case report describes the risk stratification, multidisciplinary management and successful cesarean delivery of parturient with recurrent PAS

Case Report

A 38-year-old G2P1 female with recurrent PAS presented at 23 weeks gestation with increasing dyspnea, lower extremity edema (NYHA II) and severe pulmonary artery hypertension (RVSP 66 mm Hg, severe TR; Figure 1). Past medical history was significant for two prior excisions of pulmonary artery sarcoma, left main pulmonary artery dilation, left pulmonary stent placement, postoperative external beam radiation and adjuvant chemotherapy. She was referred to our multidisciplinary team (comprised of obstetrics, anesthesiology, cardiology, neonatology, oncology, cardiac surgery and radiology) based on her history and CARPREG II risk assessment score of 7 (prior cardiac event=3, ventricular dysfunction=2, and pulmonary hypertension=2). Patients with a CARPREG II risk score >4 have a greater than 40% risk of a primary cardiac adverse event in the peripartum period (Figure 2, 3). Her pulmonary hypertension was managed with oral furosemide and sildenafil, and she underwent planned cesarean delivery under general endotracheal anesthesia with intraoperative TEE monitoring at 34 weeks gestation due to worsening functional physical status (NYHA III, Figure 3). A viable male was delivered with Apgars 9/9. Intraoperatively and postoperatively, inhaled nitric oxide and dobutamine were utilized to manage right heart failure. The patient was extubated 3 hours after admission to the ICU and placed on oxygen via high flow nasal cannula for 24 hours. She was weaned from nitric oxide 5 hours following extubation, and oral sildenafil 20 mg PO BID was resumed. Both the parturient and infant were discharged home on postpartum day 3.

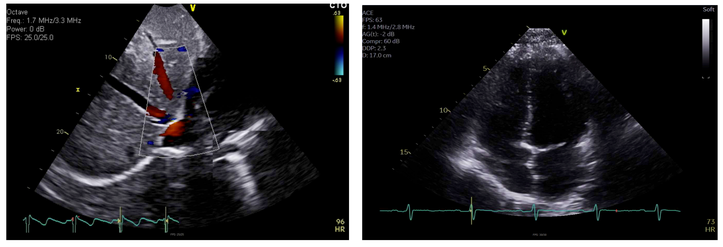

Figure 1. Preoperative TTE images showing increased venous pressure and RV Dilation

Score	Risk of Peripartum Cardiac Event
1	5%
2	10%
3	15%
4	22%
> 4	41%

Figure 2. CARPREG II Risk Assessment Score (adapted from reference 3)

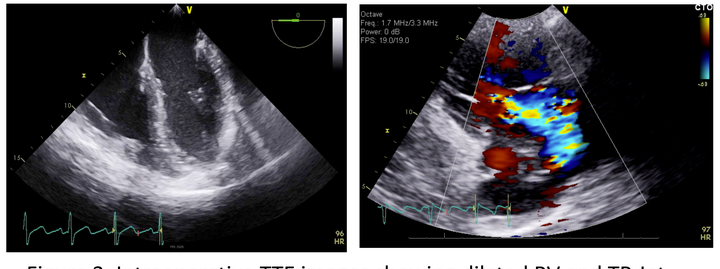

Figure 3. Intraoperative TTE images showing dilated RV and TR Jet

Discussion

Pulmonary artery sarcoma in parturients is extremely rare and is associated with significant morbidity and morality (1,2,5). Our multidisciplinary navigation team facilitated early CARPREG II risk stratification, outpatient management of pulmonary hypertension, and determination of clinical endpoints for delivery. This work enabled the successful cesarean delivery of a healthy male infant with minimal EBL in the setting of high venous pressures as well as the successful postpartum management of maternial cardiopulmonary function.

We performed elective cesarean section under general endotracheal anesthesia because it permitted intraoperative monitoring of cardiac function via TEE (figure 3), administration of inhaled nitric oxide, and if needed, median sternotomy or cannulation for ECMO. In this patient, placement of a PA catheter placement was avoided due to her history of two pulmonary artery resections via median sternotomy. Postoperative ICU care was essential to manage right heart function and postpartum fluid shifts, wean vasopressor and nitric oxide therapy, ensure adequate pain management, and maintain oxygenation and ventilation following extubation (6).

References

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