

Massive Transfusion During Emergency Ex-Utero Intrapartum Treatment Procedure For Delivery Of Infant With Congenital Infantile Fibrosarcoma

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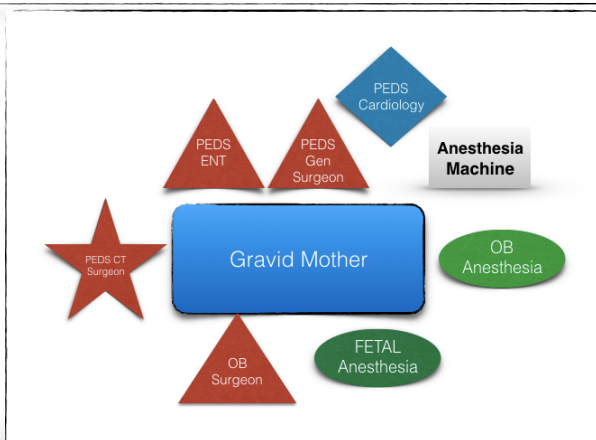
INTRODUCTION:

- Infantile Fibrosarcoma (IFS): a rare tumor accounting for 1% of all childhood cancers. It involves limbs, head, neck and trunk.
- We present our case of a giant anterior cervical IFS with an intrathoracic extension that progressed to hydrops and mandated an emergent EXIT (ex-utero intrapartum treatment procedure) with intense resuscitation and massive transfusion initiated on placental circulation.

CASE DESCRIPTION - BACKGROUND:

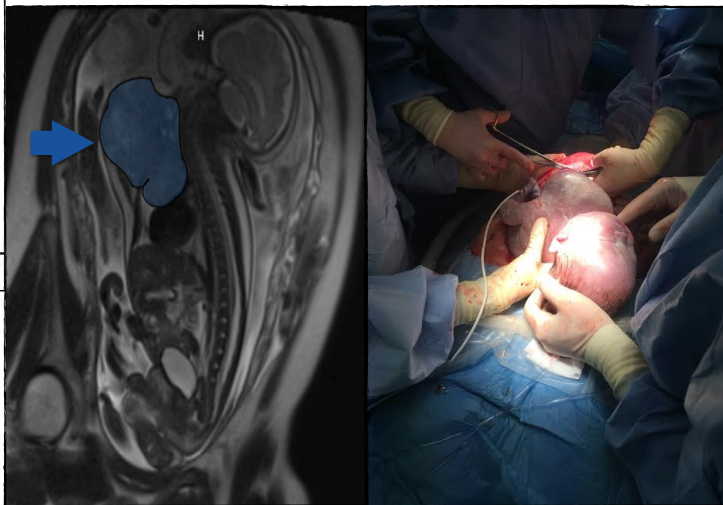
- The fetus of an 18 year old, at 34 6/7 weeks was diagnosed with a large anterior cervical neck mass on ultrasound (US).
- Fetal MRI: an anterior neck mass, 8.1 x 5.8 x 8.2 cm in size with a significant intrathoracic component and compression of the trachea, right heart border and superior vena cava.
- At 36 6/7 EGA, she presented with decreased fetal movement.
- Fetal ultrasound: scalp edema, ascites, and bilateral pleural effusions concerning for hydrops.
- Fetal echocardiography: dilated right ventricle, flattened intraventricular septum with qualitatively normal biventricular function.

MULTI-DISCIPLINARY TEAM:



INTRAOPERATIVE COURSE:

- The mass was a highly vascular tumor which bled profusely during debulking prompting the initiation of massive transfusion while on placental circulation.
- Real-time fetal echo was crucial in directing resuscitation and assessing left ventricular filling.
- The fetal airway was secured emergently by ENT under direct visualization due to premature placental separation.



RESUSCITATION OVERVIEW:

- PRBC, FFP, Cryoprecipitate, Platelets, Bicarbonate, Calcium Gluconate administered: 600cc/kg over a 4 hr period.
- 500 mcg of Epinephrine administered both IV and Intracardiac
- EBL ~ 1.5 liters.
- ABG at the end of the procedure: 7.28/39/68 BE -7.8 Hb: 8.9g/dL

POST-OPERATIVE COURSE:

- The infant was weaned off inotropic support in 48 hours. Chest closure performed on POD#3.
- MRI: residual tumor with no metastasis.
- The patient was extubated to supplemental oxygen on DOL#16 and was discharged home after 41 days in the neonatal ICU.
- Follow up: 6 months, 6 kg, on Entrectinib



DISCUSSION:

- This is the first case reported in literature of a massive transfusion initiated during an EXIT procedure.
- The novelty and extreme nature of this case emphasizes the importance for
 - ❖ optimizing team work & planning,
 - ❖ anticipation of potential emergent crises,
 - ❖ preparedness for intense resuscitation
 - ❖ massive transfusion
 during management of an EXIT procedure with a giant fetal neck mass.

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2. Pandey A, Kureel SN, Bappavad RP. Chest Wall Infantile Fibrosarcomas - A Rare Presentation. Ind J Surg Onc. 2016. 7:127-129.