It's Not a Teratoma: Neonatal Intraoperative Death Associated with Surgical Excision of a Misdiagnosed Kaposiform Hemangioendothelioma

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BACKGROUND
Fetal tumors are diagnosed clinically, have varying presentations, and require treatment associated with high morbidity and mortality.

This case presents a tumor with two valid differentials and describes how management of a neonatal tumor with conflicting treatment strategies led to intraoperative death.

Kaposiform hemangioendothelioma (KH)
- Rare invasive, vascular tumor that affects infants
- Disease incidence is unknown due to rarity
- Associated with a consumptive coagulopathy called Kasabach-Merritt Syndrome (KMS)
- KMS is characterized by profound thrombocytopenia and hypofibrinogenemia
- KMS has a mortality rate of 20-30% even with aggressive treatment such as chemotherapy

Neonatal Teratomas
- Most common fetal tumors occurring 1:30,000 births
- First line treatment includes excision and AFP monitoring

CASE PRESENTATION
8 day-old term neonate without airway compromise presents for resection of a right 6x5x5cm neck mass.

Preoperative period was complicated by anemia, thrombocytopenia, and coagulopathy requiring transfusions of PRBCs, FFP, and platelets. After medical optimization, surgical resection of the tumor was thought to definitively treat the ongoing coagulopathy.

Clinical Course
- Electively intubated DOS by the neonatologist in the ICU, and brought to OR by the anesthesiology team where GA was induced with sevoflurane uneventfully
- The initial en bloc tumor resection surgery proceeded successfully with EBL of 75-100mL requiring a 10 mL/kg bolus of PRBCs intraoperatively
- Patient was transported to NICU intubated in stable condition
- The immediate postoperative period was complicated by severe acidosis, DIC, and hemodynamic instability
- The patient was emergently brought back to the OR on vasopressors and massive transfusion protocol was initiated
- Surgical and medical interventions to control bleeding were futile and the patient died after two hours of resuscitation

DISCUSSION
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CONCLUSION
This case represents the difficulty in diagnosis and management of rare diseases where clinical evidence can be misleading. It highlights the importance of multidisciplinary preoperative discussions regarding perioperative risks in the context of uncertain diagnoses and high risk medical-surgical management.