The "High-Risk Hemorrhage" Section that Wasn't: Advanced Planning Meets Great Luck

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Introduction

Lymphatic malformations are rare, non-malignant growths of abnormal lymphatic tissue. They are typically present at birth, may progress with age, and most often affect the head or neck. We present an unusual case of diffuse lymphangiomatosis affecting pregnancy and perinatal anesthetic management.

Case Presentation

A 33-year-old G0 with extensive lymphangiomatosis was referred to our high-risk obstetric anesthesiology clinic by maternal-fetal medicine as part of multidisciplinary preconception counseling.

Background:

- LAM originally a small birthmark on neck, progressed to large multicompartmental mass
- Over 30 sclerotherapy treatments to slow growth and manage related pain and superficial bleeding
- MRI/MRA (Figures 1-3) demonstrated diffuse disease involving the chest and abdomen
- No other major medical comorbidities

Imaging

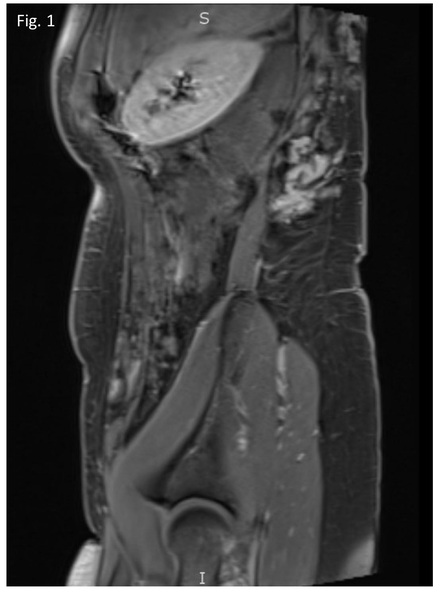
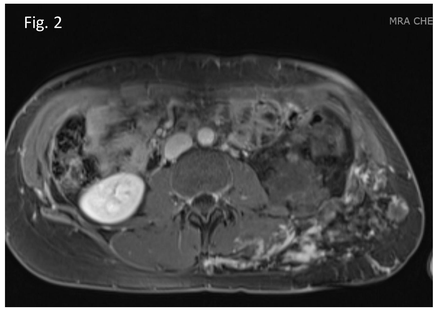
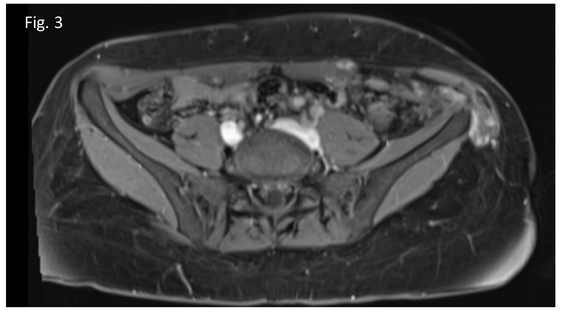


Figure 1: MRA - Sagittal view
Demonstrates thoracic involvement with mass abutting the epidural space. Also noted to be involving left pleural space.

Figure 2: MRA Abdomen
Large lymphovenous malformation of left flank with significant vascularity, displacing left kidney.

Figure 3: MRA Pelvis
Extension of mass into left adnexa, pelvic sidewall, and lateral abdominal wall, disrupting musculature. Normal right-sided abdominal wall structures appreciated.





Preconception Anesthesiology Consult

Although pregnancy was not absolutely contraindicated in this patient, we discussed increased risk involved and potential anesthetic management strategies.

We discussed the paucity of data on the behavior of LAM in pregnancy. However, physiologic changes of pregnancy (decreased systemic vascular resistance and hormonal changes) could result in vascular growth, raising concern for potential compromise of nearby aortoiliac vessels, progression to pulmonary involvement, or high-output heart failure.

We counseled against neuraxial anesthesia given concern for both spinal involvement and risk of significant bleeding should the mass be violated during placement. Alternatives such as opioid PCA for labor and general anesthesia for cesarean delivery were offered.

In the event of cesarean delivery, adnexal and abdominal wall involvement would pose significant challenges for appropriate surgical access as well as incur additional bleeding risk.

There was additional multidisciplinary discussion with maternal-fetal medicine and interventional radiology regarding mode of delivery, and concerns were raised about the ability to target potentially diffuse venous bleeding with IR embolization if the LAM was impacted by Valsalva or fetal descent during a potential vaginal delivery.

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Case Presentation (continued)

Upon becoming pregnant about 1 year later, she maintained regular follow-up with maternal-fetal medicine and underwent repeat MRI in her third trimester, which again demonstrated diffuse LAM.

Delivery planning was in progress when the patient unexpectedly presented in preterm labor at 36 weeks and 0 days.

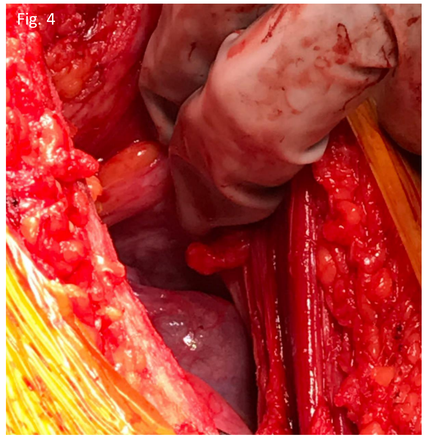
Based on prior consultations and discussions between maternal-fetal medicine, radiology, and anesthesiology, the decision was collectively made to proceed urgently with cesarean delivery.

Perioperative Management

- A rapid infusion catheter and an arterial line were placed in the triage/preop holding area prior to proceeding with delivery
- Crossmatched blood was ordered and immediately available in the operating room
- We proceeded with general anesthesia – intubation via video laryngoscopy and TIVA for maintenance with BIS monitoring
- A midline vertical skin incision allowed for avoidance of the pelvic sidewall with vacuum-assisted fetal delivery utilized to minimize pressure on the adnexa
- Blood loss was within normal limits for a routine cesarean section and our patient was extubated uneventfully
- Truncal blocks were not performed due to lymphangiomatous invasion of the abdominal wall, so post-operative pain was managed with IV PCA
- She recovered well and was discharged on post-operative day 4, doing well several months postpartum

Intraoperative Findings

Figure 4. Left Adnexa
Visible vascularity and lymphangiomatous growth seen within the pelvis and left adnexa, extending toward the abdominal wall.



Conclusions

This case highlights the importance of multidisciplinary consultation and communication in the care of complex obstetric patients, especially in scenarios where existing literature is scant or absent. Experience with large lymphovenous malformations remains extremely limited without accepted guidelines for management. As a result, the careful coordination of multidisciplinary expertise and shared decision-making proved critical to crafting and executing a safe delivery plan.

SOAP 2022 ANNUAL MEETING