



Interdisciplinary management of a medically complex parturient with sarcoidosis and splenomegaly

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Patient presentation

History of Present Illness

- 30yo G4P1102 at 25wks EGA with hepatic sarcoidosis and cHTN presented with fevers and abdominal fullness. She reported splenomegaly diagnosed on obstetric US earlier in pregnancy and several months of non-adherence with low dose prednisone therapy prescribed for her sarcoidosis.

Physical Exam

- HR 116, BP 142/79, RR 22, SpO2 100%, T 37.1
- LUQ tenderness, hepatomegaly, diaphoretic

Laboratory Data

- Hgb 7.9, WBC 2.48, Plt 239, LDH 341, Alk phos 511, all other LFTs wnl

Imaging/Tests

- Abdominal US: Progression of splenomegaly from 21 cm to 27 cm in craniocaudal direction over period of 2 wks; patent hepatic and splenic veins
- PFTs: mild restrictive defect
- CT chest: scattered micronodules, lymphadenopathy

Multidisciplinary considerations

Patient considerations: patient wished to proceed with pregnancy. Multidisciplinary team convened including hematology, rheumatology, pulmonology, general surgery, vascular surgery, MFM and OB anesthesia to counsel patient on further management.

Medical management of sarcoidosis: hematology and rheumatology consultants concluded etiology of hepatosplenomegaly was consistent with splenic sequestration from sarcoidosis. Initiated on infliximab and high dose steroid therapy. Supervision of spleen with serial abdominal US. The teams felt the patient's splenomegaly would likely stabilize with appropriate treatment of sarcoid.

Surgical considerations: general surgery consultant did not feel level of risk of spontaneous rupture warranted urgent splenectomy and agreed with medical management to achieve greater gestational latency. In the event of spontaneous splenic rupture, vascular surgery would perform endovascular aortic cross clamping given gravid uterus, necessitating emergent cesarean delivery.



CT abdomen and pelvis performed at 25w2d EGA demonstrates significant hepatosplenomegaly. Spleen measures 22.6 cm in craniocaudal direction. No evidence of vein thrombosis or portal hypertension. Gravid uterus.

Clinical course

Splenomegaly: spleen decreased in size from 22 to 17 cm after infliximab and steroids over 2 wks; blood counts also improved; inpatient monitoring throughout pregnancy for daily fetal monitoring and weekly abdominal US

Cardiac and neurologic complications: the patient developed paroxysmal AVNRT, treated with beta blockers. At 35wks EGA, TTE revealed mildly reduced LVEF; concern for cardiac sarcoid; One episode of transient unilateral upper extremity weakness, suggesting possible neurological involvement

Delivery, obstetric and anesthetic management: scheduled primary cesarean section at 35w0d with preoperative stress-dose steroids. Underwent CSE anesthesia with delivery of a healthy infant; No arrhythmias or hemodynamic instability were observed during in the perioperative period; QBL 420mL.

Recovery: uneventful; the patient was discharged postoperative day four

Discussion

Sarcoidosis is a multisystem granulomatous disease of unknown etiology. Massive splenomegaly is seen in only about 3% of patients with sarcoidosis.

No formal guidelines exist for the medical or surgical management of splenomegaly during pregnancy. The literature includes case series and reports.

Pregnancy increases the risk for spontaneous rupture of an enlarged spleen, with the highest risk in the third trimester. Mechanical factors including reduced peritoneal cavity volume and uterine contractions can cause diaphragmatic compression and a predisposition to splenic trauma.

Conversely, splenectomy during the second and third trimester carries a substantial risk of maternal morbidity and risk for abortion.

As splenomegaly improved with medical management, the risk of rupture and need for splenectomy decreased. In our assessment, the benefits of additional gestational latency and improved management of sarcoidosis outweighed the risk of spontaneous rupture under careful monitoring and continuous reassessment. The presence of competing risks and benefits requires multidisciplinary communication and involvement of the patient in shared decision-making.