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Immune System Overdrive: A Case Study of an Obstetric Patient with Hemophagocytic Lymphohistiocytosis

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This offering details the presentation, differential diagnosis and evidence-based interprofessional management of an obstetric patient afflicted with Hemophagocytic Lymphohistiocytosis (HLH).

CASE PRESENTATION

- 33 year old G2P1, 27 0/7 weeks
- Uncomplicated pregnancy; normal fetal growth ultrasound
- **8/21/16 hospital day (HD) #1** – transferred from a community hospital; admitted to a high-risk antepartum unit in a 1,200 bed Magnet® institution
- Symptoms – 6 day history of headache, fever, malaise, intermittent epigastric pain, and decreased appetite
- Sick contact – 2 year old daughter with febrile illness 1 week prior to admission
- Afebrile on admission, stable BP
- Lab Abnormalities – Platelets 94,000 (150,000-400,000), AST 188 (<41), ALT 166 (<56), LD 623 (100-250), total bilirubin 3.1 (0.1-1.0), direct bilirubin 2.5 (0-0.2)

DIFFERENTIAL DIAGNOSIS

- Atypical Hemolysis, Elevated Liver Enzymes, and Low Platelet (HELLP) syndrome
- Viral infection
- Thrombotic thrombocytopenic purpura (TTP)
- Autoimmune disorder

WHAT IS HLH?

Incidence – rare; few cases in pregnancy reported in the literature

Pathophysiology – excessive activity of the immune system; overproduction of inflammatory cytokines

Diagnostic Findings – fever, neurologic abnormalities, cytopenias, hyperferritinemia, hepatosplenomegaly, hypertriglyceridemia, elevated liver enzymes, hemophagocytosis (bone marrow aspirate)

Differential Diagnosis – preeclampsia, systemic inflammatory response (SIRS), sepsis, viral infection, autoimmune disorders, neoplastic disease

Prognosis – high mortality rate; early diagnosis and treatment is essential as initial symptoms are often non-specific

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ANTEPARTUM HOSPITAL COURSE

Hospital Day 2 – 8/22/16

Febrile to 103.0 at 0140, leukopenia with bandemia, maternal (HR 130–140) and fetal (FHT 170s) tachycardia, tachypnea, hypoxemia (pulse ox 88–90% on room air), BP 90/50, complaints of chest pain/pressure and labored breathing

Interventions: Diagnostic testing, IV fluids and antibiotics, additional labs, infectious disease consult, O2 via nonrebreather mask

Rapid decompensation – transferred to ICU 8/22/16 at 0515

Hospital Day 3 – 8/23/16

New onset of encephalopathy at 1950, worsening hypoxia, hypotension, agitation

Interventions: Intubated, placed in a medically induced coma, blood pressure support

Hospital Day 4 – 8/24/16

Further respiratory decompensation, oxygen desaturation to 86–88%; addition of nitric acid therapy and high frequency ventilatory support – not tolerated, maternal hypotension, fetal bradycardia

Interventions: decision made for stat cesarean section at the bedside at 27 3/7 weeks for a 1170 gram living male infant, apgars 3/5 → immediate maternal transport to OR for Extracorporeal membrane oxygenation (ECMO)

POST-OP COURSE

Post-op Course → **DIC** → **ECMO decannulation 9/3/16**

Complicated by: → **Numerous transfusions** → **Stable – extubated 9/11/2016**

Dialysis → **OR visit x 3 for bleeding complications** → **Discharged to home with specialty follow-up 9/20/2016 (HD#31)**

UNCOVERING THE DIAGNOSIS

- **8/25/16**
  - Ferritin level 10,226 (10-291)
  - Triglycerides 1,981 (<150)
- **8/25/16** Bone marrow biopsy – phagocytes suggestive of HLH

TREATMENT

- Goal - to control an overactive immune system
  - Etoposide – chemotherapy medication
  - Steroid therapy – Dexamethasone IV

LESSONS LEARNED

- Seek interdisciplinary support and management in order to provide high quality care to the critically ill obstetric patient.
- Recognize the need for a higher level of care and coordinate timely transport to a tertiary care facility to ensure optimal patient outcomes.
- Think 'outside the box' with pregnant patients – use your interprofessional resources to find answers.

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