SOHO ANNUAL MEETING 2024

HLH (Hemophagocytic Lymphohistiocytosis) in pregnancy: Management of a case with steroid monotherapy and review of the literature

Sara Soliman1, Asad Haider1, Sara Taveras Alam1, Shehzad Merwat2, Adan Rios1
1. Division of Hematology/Oncology, McGovern Medical School, The University of Texas Health Science Center at Houston.
2. Division of Gastroenterology, Hepatology and Nutrition, The University of Texas Health science Center at Houston

UTHealth The University of Texas Health Science Center at Houston

INTRODUCTION

Hemophagocytic lymphohistiocytosis (HLH) in pregnancy is extremely rare with less than 100 reported cases. Symptoms are nonspecific and abnormal laboratory markers can resemble other pregnancy related conditions. Currently, there is no consensus on the diagnosis and treatment of HLH in pregnancy.

OBJECTIVE

Discuss the challenge of pregnancy associated HLH and report one case effectively treated with steroid monotherapy.

INITIAL PRESENTATION

- A 20-week pregnant 38-year-old female presented with abdominal cramps and jaundice.
- Initial workup revealed WBC 6,500/mm3, Hb 6.8 g/dl, PLT 236,000/mm3.
- AST 5,780 IU/L, ALT 2,471 IU/L, ALP 142 IU/L, and total bilirubin 12 mg/dl.
- Ferritin **was >40,000 ng/mL** and IL-2 (CD25) was 950 pg/mL.

DIAGNOSTIC WORKUP

CMV&EBV IgM	Negative
Hepatitis panel	Negative
AMA, antismooth Ab	Negative
Alpha1 antitrypsin and ceruloplasmin	Unremarkable
Abdominal MRI	No thickening or biliary dilation
Trans jugular liver biopsy	Diffuse lobular disarray, lobular inflammation with occasional hemophagocytes

TREATMENT and RESPONSE

She received dexamethasone 40 mg x 10 days

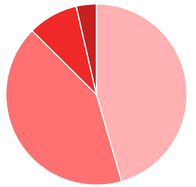
She was discharged home on prednisone taper, prednisone is selected given its completely metabolized by the placenta

Followed up ferritin levels and CD25 remained within normal limits.

LITERATURE REVIEW

1991→2024 literature review of 66 cases of obstetric HLH

Etiology



Idiopathic
Infectious
Lymphoma
Familial HLH

Maternal Mortality-→ 26%

Fetal Mortality-→ 49%

12 cases were treated with steroids only

Maternal Survival: 100%
Fetal Survival: 75%

CONCLUSION

HLH in pregnancy is very rare and can lead to high fetal and maternal mortality. Most secondary cases are due to infections. However, pregnancy itself can trigger HLH. The presentation is usually vague; yet, ferritin levels of >40,000 are almost seen exclusively in HLH. Early clinical suspicion and treatment are critical. Steroids are safe throughout pregnancy and can induce full remission in most cases without the need for chemotherapy.