ORIGINAL RESEARCH PAPER

Obstetrics & Gynaecology

HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS WITH NON- IMMUNE HYDROPS IN PRIMIGRAVIDA AND ITS OUTCOME

KEY WORDS: RH-positive, hemophagocytic lymphohistiocytosis, non-immune hydrops, emergency LSCS, perinatal mortality

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ABSTRACT

Hemophagocytic lymphohistiocytosis is a very rare disease associated with pregnancy with a high mortality rate and with an incidence of in 50,000 with an equal distribution amongst males and females with autosomal recessive inheritance. We are reporting a case of , 27 year old Primigravida with Rh-positive pregnancy with 32 weeks period of gestation who presented to the labor room with scan report showing severe fetal anemia with history intra- uterine blood transfusion done at 20 and 216 weeks and emergency LSCS was done, a female baby was extracted and died on day 7 due to HLH which was confirmed by bone marrow biopsy.

INTRODUCTION

Hemophagocytic lymphohistiocytosis is a life threatening condition. There is an impaired function of natural killer cells and cytotoxic T-cells leading to uncontrolled proliferation of lymphocytes and histiocytes which leads to hyperinflammation. Its of two types: primary which is genetic and secondary which is acquired. It's associated with gene mutations in PRFI gone, MUNC gene, and STXII gene. The clinical presentation of HLH is often non-specific and mimics infections such as cytomegalo virus, rubella, HIV and hence leads to delayed diagnosis and treatment. According to Suzuki et al recommends that neonates with hepatomegaly, thrombocytopenia and elevated LDH levels should be evaluated for HLH

Case Report

A 27 year old Primigravida presented at 32 weeks of gestation with obstetric ultrasound showing fetal anemia and fetal ascites and pleural effusion indicating fetal hydrops. A diagnosis of non-immune hydrops was madebased on the scan findings. On PA examination uterus was relaxed and fetal heart rate was good. On PV os was closed and uneffaced.

Emergency LSCS was done in view of fetal anemia and a single female baby was extracted of weight 1.80 kgs after which the baby was shifted to NICU, the baby was intubated at birth. The fetal hemoglobin level was 5.0gm/dl and two packed red blood cell transfusions were done. The baby had fever spikes and splenomegaly. Bone marrow biopsy was done suspecting hemophagogyctic lymphochistiocytosis. The baby succumbed on day 7 of life.

DISCUSSION

HLH is a reactive process resulting from prolonged and excessive activation of antigen presenting cells and cd8 cells leading to hypersecretion of pro-inflammatory cytokines and this exaggerated inflammatory response is responsible for necrosis and multi-organ failure and leads to uncontrolled transplantation proliferation and phagocytic activity of histiocytes which leads to extensive systemic involvement. The criteria for diagnosis of HLH include 3 of 4 fever, splenomegaly, pancytopenia [atleast 2 cell lineages], hepatitis and atleast 1 of 4: hemophagocytosis, increased ferritin, increased sil2ra and absent or decreased NK cell function. The mainstay of treatment of HLH includes hematopoietic stem cell transplantation.

CONCLUSION

Early diagnosis and prompt treatment is necessary for ensuring a good maternal and fetal outcome.

REFERENCES

1. Malloy, C., Polinski, C., Alkan, S. et al. Hemophagocytic Lymphohistiocytosis Presenting with Nonimmune Hydrops Fetalis. J Perinatol 24, 458-460 (2004).
2. Bellini C. Hennekam RCM. 2012, Non-immune hydrops fetalis: A short review of etiology and pathophysiology. Am J Med Genet Part A 158A:597-605.