ORIGINAL RESEARCH PAPER

Obstetrics & Gynaecology

A RARE CASE REPORT OF EVANS SYNDROME IN PREGNANCY

KEY WORDS: Evans Syndrome, Autoimmune Hemolytic Anemia, Immune Thrombocytopenia

Dr. Surbhi Pandya Consultant MS Obstetrics and Gynecology.

Dr. Sravanth Gandavarapu Consultant MD General Medicine.

ABSTRACT

Evans Syndrome is a rare autoimmune disorder in which there is a coexistence of Auto Immune Hemolytic Anemia (AIHA) and Immune Thrombocytopenia (ITP) meaning that the individual antibody attacks its own the red blood cells and platelets 1. There can be simultaneous or sequential development of Autoimmune Hemolytic Anemia and Immune Thrombocytopenia. Evans Syndrome can be diagnosed with a full blood count film and Coombs test. Association of Evans Syndrome in pregnancy is very rare and only a few cases have been documented till date. A definite treatment protocol has not been defined and the treatment becomes even more challenging in pregnancy due to concerns of teratogenic effect of the pharmacological agent 2. We describe here a rare case of Evans Syndrome that was diagnosed during pregnancy, its possible treatment options and neonatal outcome.

INTRODUCTION

Dr. Robert Evan first described the syndrome in 1951, and the first case of its occurrence during pregnancy was published in 1966. Evans Syndrome is a rare autoimmune condition characterized by Immune Thrombocytopenia (ITP) and Autoimmune Hemolytic Anemia (AIHA) that can coexist or develop one after another and can be diagnosed with a positive Direct Coombs Test (DCT). Underlying etiology is unknown. Estimated prevalence is 1-9 per million. [3] It is described in both children and adult; it is more common in women (women: men ratio 2:1). [2] It is a chronic diseases with frequent remission and relapse and runs a more benign course during pregnancy (Immunocompromised state). However, the fetal outcome can be less favorable due to transplacental passage of antibodies. In pregnancy it is essential to differentiate between Evans Syndrome and more common disorders such as preeclampsia, disseminated intravascular coagulation (DIC) or syndrome of hemolysis, elevated liver enzymes and low platelet count (HELLP).

Case Report

A 28-year-old primigravida with 38 weeks of gestation presented to casualty with c/o of abdominal pain and decreased fetal movements. She was a diagnosed case of autoimmune hemolytic anemia two years pre-pregnancy and was on oral prednisolone 5mg since then. On general examination, her blood pressure was 120/80 mmhg, pulse rate was 84/min, respiratory rate was 18/min. Her weight and height were 89kgs and 163 cm respectively. Uterus size corresponding to gestational age.

Complete blood count was advised which showed hemoglobin 8.8 g/dl, hematocrit 36.9%, white blood cells 13,000 /microL and platelet count of 60,000/microL . Peripheral smear showed microcytic hypochromic anemia with anisopoikilocytosis with decreased platelets (macro platelet). Direct Coombs test was positive. Anti Platelet Ab positive. LDH was 1014 U/L and Vitamin B12 level was 191 pg./ml. Prothrombin time was within normal limits. ANA and Lupus anticoagulant test was negative. Thyroid profile was deranged with TSH of 7.95, ultra-sonography of neck s/o thyroiditis grade 1 and anti TPO antibodies Negative. Her glucose challenge test, urine examination, liver and renal function tests were within normal limit. Obstetric Ultrasound was done showing a single live fetus of 38 weeks with normal doppler study.

Platelet count dropped to 30,000/UL and decision of stepping up steroids to inj. methylprednisolone 1g/day for three days (pulse therapy) followed by oral prednisolone 1mg/kg/day and SOS IVIg infusion of 0.4mg/kg/day for 5 days.

Patient was taken for Emergency Lower Segment Cesarean Section at 39weeks of Gestation in view of Fetal Distress after transfusing 4 SDP and 1 PRBC as patients HB was 7.3g/dl and Platelet count was 24000/UL. A healthy female newborn of 3.3kg was delivered and shifted to NICU in view of Respiratory Distress and was intubated for the same and managed by neonatologist. Both mother and baby were discharged on Day 11 of delivery.

After discharge patient was continued on Prednisolone 1mg/kg/day and her hb was 9.7g/dl and platelet count was 60,000/UL at the the time of discharge.

DISCUSSION

Evans Syndrome is a rare autoimmune disorder associated with simultaneous or sequential occurrence of Coombs positive hemolytic anemia and thrombocytopenia. It can have an underlying cause and can also be associated with other autoimmune disorders. In the present case Evans Syndrome was diagnosed after sequential development of thrombocytopenia at term.

Our patient was already a diagnosed case of Autoimmune Hemolytic Anemia. Autoimmune Hemolytic Anemia is characterized by development of anti-erythrocyte antibodies (anti-E) and destruction of erythrocytes. It is further classified into warm (65%), cold (30%) and mixed type (5%). The presenting features of AIHA are acute anemia, hemolysis and a positive Direct Coombs Test (DCT). Common conditions associate with hemolysis and thrombocytopenia in pregnancy like HELLP, preeclampsia, DIC, acute fatty liver of pregnancy and drug induced conditions were ruled out with detailed history, examination and blood investigations. Direct Coombs test was positive indicating further likelihood of immune thrombocytopenia. Sequential development of immune thrombocytopenia in a known case of Coombs positive autoimmune hemolytic anemia was noted and diagnosis of Evans Syndrome was made.

Treatment modalities available are intravenous gamma globulins, danazol, cyclophosphamide, vinca alkaloids, azathioprine, plasmapheresis and even splenectomy in refractory cases. In our case treatment was given with corticosteroids, patient responded well to the steroid therapy and an increase in platelet count was noted.

CONCLUSION

Evans Syndrome in pregnancy is a very rare condition and requires multidisciplinary approach involving specialist in

Obstetrics and Gynecology, Medicine, Hematology and Fetal medicine expert/ Neonatologist. Regular follow up is required during pregnancy to ensure a favorable maternal and fetal outcome.

REFERENCES

1. Evans Syndrome - StatPearls - NCBI Bookshelf (nih.gov)
2. Evans' Syndrome: From Diagnosis to Treatment - PubMed (nih.gov)
3. Evans' Syndrome in Adults: An Observational Multicentre Study | Blood | American Society of Hematology (ashpublications.org)