Case Report

Evans syndrome and pregnancy: A case report

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ABSTRACT

Evans syndrome is characterised by simultaneous or sequential development of autoimmune thrombocytopenia and autoimmune hemolytic anemia in the presence or absence of a known underlying cause.[1,2] Autoantibodies against erythrocytes, platelets and neutrophils have been shown in Evans syndrome. Both the occurrence of isolated episodes of thrombocytopenia and hemolytic anemia and the results of in vitro studies have suggested the roles of non cross reacting autoantibodies targeted at different antigenic determinants on red cells and platelets. Both cellular and humoral immune response abnormalities have been shown in evans syndrome.

There have been a few case reports of Evans syndrome during pregnancy.[3,4] we present a case of Evans syndrome during pregnancy.

Introduction

A 22 year old primigravid woman was admitted to the hospital at 32 weeks of gestational age with anemia. patient was asymptomatic, on general examination pallor was present .Her height and weight were 164 cm and 62 kg respectively. Blood pressure was 110/70 mm of Hg, pulse rate was 82/min, initial blood tests included Hemoglobin-6.4gm/dl, a white blood cell count of 12,500 cells/cumm with 81.9% neutrophils and a platelet count of 58,000/cumm. The prothrombin time and partial thromboplastin time were normal. HIV, HbsAg were negative. ultrasonography revealed a single viable fetus compatible with 32 weeks of gestation. The peripheral smear on complete hemogram showed a microcytic hypochromic anemia, iron studies were within normal limits. A initial diagnosis of HELLP syndrome was made. But with detailed history patient complained of alopecia and oral ulcers, so a complete work up of the patient was made, and she was positive for direct and indirect coombs test, so ANA and dsDNA were done. anti-nuclear antibody(ANA) and dsDNA were positive. So a final diagnosis of systemic lupus erythematosis (SLE) was made with Evans syndrome.

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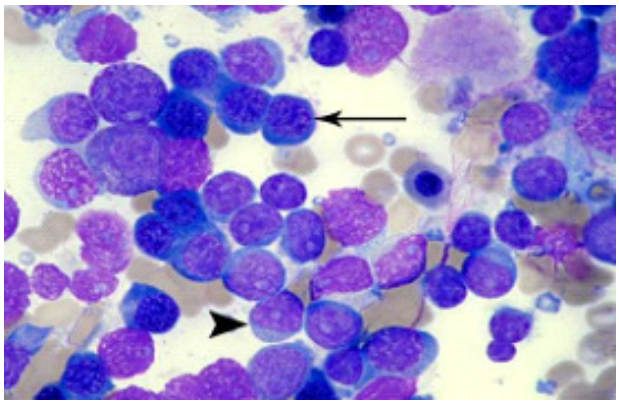

Erythroid precursors are seen in hemolytic anemia

Patient was started on Dexamethasone TID, and the laboratory parameters were regularly monitored.

patient was transfused with 2 units of packed cells. With Dexamethasone patients platelet count and Hb improved and at term (37 weeks of gestation) her Hb was 11gm/dl and platelet count was 1.5 lac/cumm. delivery was uneventful and patient was switched on to prednisolone therapy in post partum period and regularly followed up.

Discussion:

Evan's syndrome is a chronic hematologic condition associated with simultaneous or sequential occurrence of coombs positive autoimmune hemolytic anemia and autoimmune thrombocytopenia.[2] As with autoimmune hemolytic anemia, Evan's syndrome may have an underlying cause. Evan's syndrome is a rare occurrence during pregnancy. Systemic lupus erythematosus (SLE) is an autoimmune disease in which the immune system attacks the body's cells and tissues, resulting in inflammation and tissue damage. it is most commonly seen in females in reproductive age group. Evan's syndrome is a rare manifestation in SLE, occurring in patients with severe multisystemic SLE manifestations.

Complications of SLE in pregnancy include high blood pressure, preeclampsia, hemorrhagic complications[6] such as abruption of placenta and post partum hemorrhage. There is high incidence of miscarriage and fetal loss in women with SLE in pregnancy. Infants born to women with SLE have a higher risk of low birth weight, prematurity, complete heart block. Complications in Evan's syndrome in pregnancy can be divided into the consequences of autoimmune hemolytic anemia and autoimmune thrombocytopenia. In our patient fetal outcome was good.

Treatment of SLE in pregnancy includes prednisolone can be given safely, NSAIDS should be used with caution, and all the other treatment modalities are contraindicated in pregnancy. In our case patient was treated with dexamethasone as per PRIDE study.

Regarding the fetal and neonatal status, no antenatal measures can reliably predict the situation, and maternal response to intervention does not guarantee a favorable fetal or neonatal outcome.[7] To date, only previous neonatal outcome provides a useful predictor of the fetal and neonatal platelet count in a subsequent pregnancy.[8] Nevertheless, the platelet antibody level should be measured in these cases. Platelet antibodies can pass through the placenta and bind to the fetal platelets in fetal thrombocytopenia.

In conclusion, anemia and thrombocytopenia in pregnancy should be thought of Evan's syndrome as differential diagnosis apart from HELLP syndrome. Evans syndrome in pregnancy is a very rare condition. Close surveillance and early management during pregnancy maximize the chance of favorable outcome.

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