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A challenging case Report on Evans syndrome with pregnancy

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ABSTRACT

The simultaneous or sequential development of autoimmune hemolytic anemia (AIHA) and idiopathic thrombocytopenic purpura(ITP) is known as Evans syndrome. AIHA is characterized by the development of anti-erythrocyte auto antibodies and the destruction of erythrocytes, which causes moderate or severe anemia. Because Evans syndrome is a rare disease, little is known about the relationship between pregnancy and Evans syndrome.

Keywords: *Idiopathic Thrombocytopenic Purpura, Autoimmune Hemolytic Anemia*

I. INTRODUCTION

In a pregnant women, the hemoglobin concentration and platelet count decrease as a result of marked plasma augmentation. Moderate to severe anemia and thrombocytopenia may however, cause fetal growth restriction or premature birth Thus it is importance to know the cause of anemia and bleeding and PPH. The simultaneous or sequential development of autoimmune hemolytic anemia (AIHA) and idiopathic thrombocytopenic purpura (ITP) is known as Evans syndrome. We experienced a case of Evans syndrome that developed AIHA and ITP during pregnancy.

II. CASE REPORT

A 23years old primi gravida with 23 weeks of gestation age had developed symptoms of generalized weakness, gum bleeding and High grade fever, difficulty in breathing and Hematuria. Up to 22 weeks, her pregnancy was uneventful. Patient got admitted in private hospital for the above said symptoms on 23 rd week of gestation and had treatment for low platelet count and fever. Patient condition worsened and patient was referred to tertiary care Centre. Patient was received in an emergency department in our hospital with High level support of oxygen. Her HR-120b/mt,RR-28b/mt. BP was 137/75 mmHg, spo2-93%. Patient developed Acute Respiratory Distress Syndrome and Diffuse Alveolar Hemorrhage. Patient was shifted to ICU. Immediately Patient got intubated and given lung protective ventilation to her.

III. INVESTIGATION

USG showed mild splenomegaly and ECHO Showed Good LV function with EF -55%. Bilateral infiltrates and consolidation found on the Right middle zone of the lung. Bone Marrow Biopsy revealed Normal megakaryocytic with thrombocytopenia and No Histocytes were found. In Flow cytometry there is no abnormal cells found, negative for DNT and PNH. LDH is 1153, Fe-204, CRP-41.8, B12->2000,

Folate-9.78, D-dimer-732, DCT is 3+, ICT 1+. HRCT chest showed Ground glass opacities in multi focal areas with associated interlobular septal thickening in both lungs with lobular areas of parenchymal sparing found in the lungs. Segmental fibro bronchiectatic changes with calcification and volume loss seen in Left lower lobe. Multiple calcified mediastinal and bilateral hilar nodes were present. CT Angiography in pulmonary system showed chronic hemorrhage in the left upper lobe pulmonary artery, posterior segmental branch shows non opacification with paucity of vessels in this region.

TABLE1: LAB INVESTIGATION CHART

Lab investigation/Date	9/05	11/05	19/05	02/06	6/6	16/6
T.L.C	14.45	9.9	5.65	6.74	7.74	6.05
Hemoglobin	8.6	6.8	6.8	7.1	8.4	8.4
Hematocrit	29.1	22.6	22.2	24.3	29.9	29
Platelets	2000	11000	3000	<10000	<10000	10,000
Total Billirubin	3.8	6.44	3.7	1.63	1.19	1.1
ALT	123	136	406	79	75	51
AST	190	142	357	160	66	50

Patient had higher level of antibiotics and prone ventilation along with platelets blood transfusion and steroids too. She had hematuria and frequent blockage of urinary catheter was present. She had continuous bladder irrigation. Patient got IVIG course along with steroids and antibiotics, patient general condition got improved. Patient extubated successfully and put on oxygen face mask. But platelets counts and Hb level was not improved.

Patient was started with second line treatment of intravenous monoclonal antibodies (Rituximab) 600mg od after getting consent from the patient regarding fetal risk and side effects. Despite of second line treatment patient condition was not improved. she has been started for Eltromobhage75mcg od, after three weeks patient platelets and Hemoglobin level got improved. At 34+2 weeks patient felt decreased fetal movements. Immediately she has undergone Emergency Lower segment cesarean section under spinal anesthesia. During Intra operative period blood loss was less than 500ml and managed with fluids and blood products. A live male baby was born with 1650gms and APGAR score were 7/10 and 8/10 at 1st and 5th minutes of period. The baby was shifted to NICU for further management. The mother was shifted to critical care unit for observation. Her post natal period was uneventful. She was shifted to ward.

Postnatal her uterus was contracted well, and her breast was soft and early ambulation was encouraged. She got discharged from the hospital on 20th day of post operative day. During discharge her condition was stable and Hb, platelet level was 11.7 and 4.5 lakhs respectively.

DIFFERENTIAL DIAGNOSIS- ANA-1:100, ANCA was Negative, Anti DS DNA-10IU/ml, Leptospira, scrub typhus, HIV, HCV reports are Negative. Hemolysis, elevated liver enzymes, low platelet count (HELLP) syndrome; hemolytic uremic syndrome (HUS); and thrombotic thrombocytopenic purpura (TTP) were unlikely. Systemic lupus erythematosus was reported as a disease that causes anemia. but was also unlikely because a test for anti-ds DNA was negative. Her C3, C4, and erythrocyte-binding IgG, IgA and IgM levels were also normal. In flow cytometry finding do not meet the diagnostic criteria of autoimmune lympho proliferative syndrome. AntiLKM1 Level is 1.0unit/ml, ASM is Negative. There was no corresponding medical history or symptoms of infection that could have contributed to the observed hemolytic anemia.

IV. DISCUSSION

Evans syndrome (ES) is an uncommon autoimmune disease that was defined by Robert Evans in 1951 when he studied the relationship between autoimmune hemolytic anemia (AIHA) and immune thrombocytopenic purpura (ITP). He described the first diagnosis criteria of ES, including the presence of anemia, reticulocytosis, increased blood bilirubin, no family history of hemolytic diseases, evidence of antibodies against erythrocytes at 37°C, hemolysis of transfused erythrocytes, the presence of purpura, prolonged bleeding time, bone marrow aspiration with normal or increased number of megakaryocytes and the absence of exogenous toxic agents or a baseline disease.

V. ETIOLOGY

An exhaustive clinical history must be taken to determine the risk factors for developing ES, such as infections, malignancies, autoimmune diseases, recent vaccinations, drugs or a family history of immune disorders, complemented with a thorough physical examination focused on signs of anemia or thrombocytopenia.

Table 2: Risk factors /Etiology of Evans syndrome

S.No	BOOK PICTURE	PATIENT PICTURE
1.	Etiology is Unknown	Unknown
2.	Triggering factor • Infection • Underlying disorders	Probable infection
3.	Secondary causes a. Auto immune lympho proliferative syndrome. b. Lupus c. Anti phospholipid syndrome d. Sjogrens syndrome	Nil
4.	IgA deficiency	Nil
5.	Lymphomas	Nil
6	Chronic lymphocytic leukemia	Nil
7	Lack of erythrocyte membrane proteins	Nil

VII. DIAGNOSTIC EVALUATON

The characterization of laboratory features requires a complete blood count and direct examination of peripheral blood; anemia, thrombocytopenia, reticulocytosis, poikilocytosis, mainly due to the presence of spherocytes, increased indirect bilirubin and lactate dehydrogenase and a positive direct anti-human globulin test(DAT) test confirming ongoing immune hemolysis are to be effected.

Figure 1-platelet counts

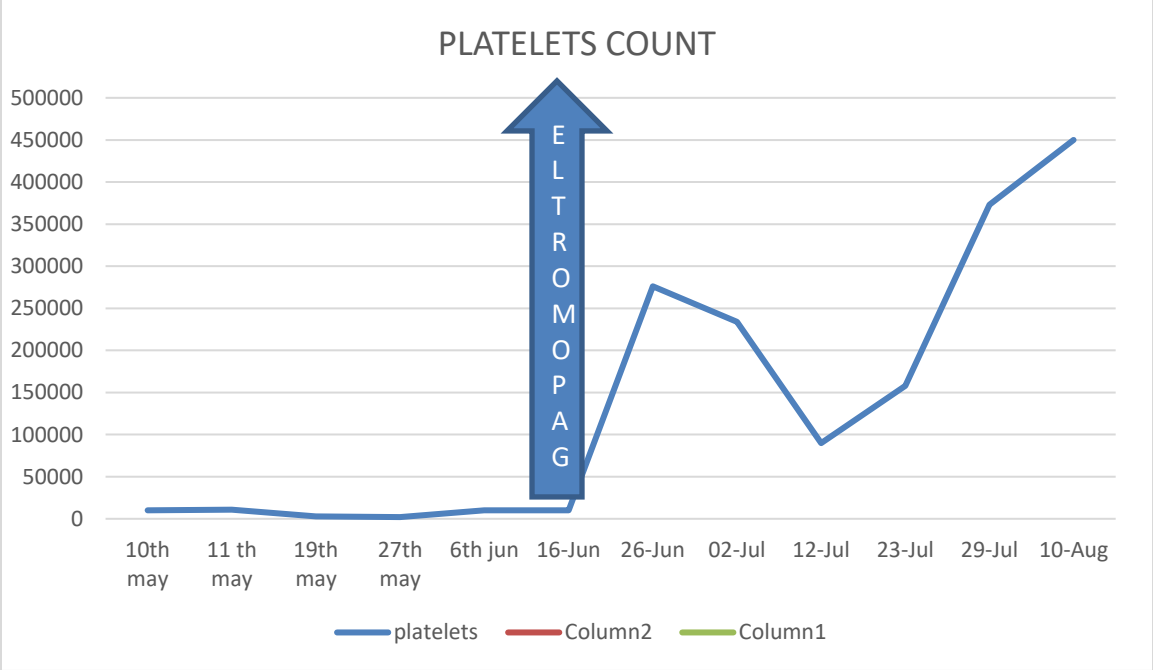

Table 3: Diagnostic Evaluations of Evans syndrome

S.no	Book Picture	Patient Picture
1.	Peripheral smear Retic Count	normal morphology 8.60
2.	Complete blood count	HB-6gms, plts-2000,wbc-20000
3.	Coombs test	DCT 3+, ICT1+

4.	Test for anti neutrophil, antiplatelet anti erythrocyte antibodies	Negative
5.	Serum immunoglobulin levels	IgG levels are normal
6.	cogulation profile	Normal
7.	Liver function test	TB-3.6
8.	Bone Marrow aspirartion	Normal megakaryocytic with thrombocytopenia
9.	Flowcytometry	No abnormal cell, PNH and DNT – ve.

VIII. CLINICAL FEATURES

Clinical features are associated with anemia and thrombocytopenia including pallor, weakness, fatigue, jaundice, petechiae, ecchymosis, Gingivorrhagia and epistaxis. Due to the prolonged immunosuppressive therapy and/or the associated underlying immune deficit, there is a risk of 66.6% of patients developing respiratory tract infections

Table 4: clinical features of Evans syndrome

s.no	Book Picture	Patient Picture
1.	Shortness of breath	Present
2.	Anemia	Hb-6 gms
3.	Bleeding	gingival and lips bleeding
4.	Bruising	NIL
5.	Dizziness	NIL
6.	Feeling cold	Present
7.	Tiny red dots	present on the abdomen and limbs
8.	Low RBC	HB 6 gms
9.	Weakness, fatigue,	Present
10.	Purpura	-
11.	Susceptible to infection	Wbc-20,0000
12.	Thrombocytopenia	2000
13.	HELLP syndrome	-
14.	Increased Heart rate	130b/mt
15.	Jaundice	-
16.	Epistaxis, gum bleeding	Present
17.	Hematuria	Present

Evans syndrome During Pregnancy

Evans syndrome during pregnancy is not frequent and usually the diagnosis is established previously; main differential diagnoses in this circumstance are HELLP syndrome, thrombotic thrombocytopenic purpura and uremic hemolytic syndrome. There is a passive immune transfer of maternal IgG antibodies via placenta to the fetal circulation, which explains the transient thrombocytopenia or hemolytic anemia previously reported in newborns or fetus of women with autoimmune cytopenia. In general, pregnant women with ES have a good outcome if appropriate treatment is administered opportunely; however, there is an increased risk of developing abruptio placentae and postpartum hemorrhage. Fetuses usually have a bad prognosis associated with high-risk complications occurring such as severe hemolytic anemia or hemorrhages secondary to significant thrombocytopenia, mainly intracranial bleeding with intra-extra uterine death or neurological impairments. Hematological therapy is challenging because of drugs' teratogenic effects; treatment of choice is steroids plus IVIG. Its action mechanisms include a decreased level of antibodies crossing the placenta and of maternal IgG antibodies by down regulation, with a reduction of these in fetal circulation. Last treatment options are azathioprine or splenectomy, exclusively in refractory third trimester cases.

Table 5: Effects of Evans syndrome in pregnancy

S.NO	Book Picture	Patient Picture
1.	Still birth	Nil
2.	Postpartum Hemorrhage	Nil
3.	Abruption placenta	Nil
4.	Sepsis	stenotrophomonas maltophilia in blood
5.	Maternal cardiovascular problems	Nil
6.	Fetal thrombocytopenia	
7.	Fetal subdural Hematoma	Nil

X. TREATMENT

The main objective consists of achieving a long-term complete response. There is no therapeutic regimen established. Steroids with and without IVIG are recommended as front-line therapy. Red blood cell/platelet transfusion is indicated only in severe symptomatic patients due to the risk of exacerbations. It is postulated that steroids inhibit the ability of macrophages for clearing platelets and erythrocytes. Prednisolone or prednisone at 1–2 mg/kg/day must be administrated in all cases, and in patients with severe clinical manifestations, an initial dose of 4–6 mg/kg/day within the first 72 hours is recommended. This is an important life-saving resource in uncontrolled patients; the most common dose administered is 0.4 g/kg/day for 4 days and 5 g/kg in the case of predominance of AIHA. Rituximab is a chimeric anti-CD20 targeted drug that has been increasingly used as the second-line treatment in steroid-refractory or relapsing ES. It has an immunosuppressive effect with B-cell depletion in peripheral blood after one infusion. Splenectomy, HSCT and cyclophosphamide are third line treatments in patients who doesn't respond to other treatments.

Table6: Treatment option for Evans syndrome

S.NO	Book picture	Patient Picture
1.	Steroids	Inj. Dexa 6mg OD
2.	IVIG	Inj.IvIg 60 ms
3.	Immunosuppressant	Inj. Rituximab 600mg once in a week
4.	supplemental therapy	Tab. Folvit od, Tab calcium 500mg bd
5.	Thromboplastin Receptor agonist	T. Eltromobhage 75 mcg od
6.	Stem cell transplantation	-

XI. CONCLUSION

In this case report a 23 year old primi-gravida mother diagnosed with Evans syndrome, it is an uncommon hematological condition which is difficult to diagnose. Evans syndrome is a chronic disease with relapse and remission. It was a very challenging for clinician, obstetrician, hematologists and nurses to diagnose and manage the patient during pregnancy. Meticulous fetal and maternal surveillance is necessary throughout the peripartum period. Early diagnosis and timely management may help the patient to recover from the disease and have a un eventful intrapartum and lead a normal post partum life.

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