

## Evans Syndrome and pregnancy: A case report with literature review

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### Abstract

Evans syndrome is a rare autoimmune disease in which an individual's antibodies attacks the body's own red blood cells and platelets.<sup>1</sup> There is a coexistence of Immune thrombocytopenia (ITP) with Autoimmune haemolytic anaemia (AIHA) and both of these events may occur simultaneously or one follows the other.<sup>2</sup> Association of Evans syndrome with pregnancy is very rare, and only a few cases have been published in medical literature. No definite treatment protocols are defined. Treatment options during pregnancy are further limited due to concerns of teratogenic effect of pharmacological agents.<sup>3</sup> Evans syndrome can be diagnosed with a full blood count film and a Coombs test. We describe here a rare case that was diagnosed as Evans syndrome during pregnancy that resulted in birth of stillborn twins after the case became complicated with the onset of Eclampsia. We have also briefly discussed the pathophysiology, the possible treatment options during pregnancy and prenatal outcome of Evans syndrome. This case was presented in June 2015 at the outpatient department of Obstetrics & Gynecology in Nishtar Medical College and Hospital, Multan.

**Keywords:** Evans syndrome, Evans syndrome during pregnancy, Thrombocytopenia, Autoimmune haemolytic anemia

### Introduction

Evans syndrome is an uncommon condition characterised by Immune thrombocytopenia (ITP) and Autoimmune haemolytic anaemia (AIHA) that can coexist or one follows the other, and it is diagnosed with a positive Direct antiglobulin test (DAT). The underlying etiology is unknown. It has a more benign state during pregnancy than in non-pregnant state. The foetal outcome may become less favorable, as it is affected by transplacental passage of antibody. It is suggested that Evans syndrome may be a stage of a broader spectrum, generalised

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immune deregulation due to high incidence of quantitative serum immunoglobulin abnormalities, lymphoid hyperplasia and associated systemic manifestations.<sup>3</sup> Dr. Robert Evans was the first one to describe the syndrome in 1951, and the first case of its occurrence during pregnancy was published in 1966. Since then, very few cases of Evans syndrome during pregnancy have been reported.<sup>4</sup> This is why a case diagnosed at the Nishtar Hospital Multan, Pakistan is presented in this case report.

### Case Report

In June 2015, a 20-year-old primigravida married for 11 months was presented in the outpatient department of Obstetrics & Gynecology in Nishtar Medical College and Hospital, Multan. She had gestational amenorrhea of 6 months, complaints of general body weakness for the last 3 months, rash for the last 2 months and blood in stools for four days. She was admitted in the medical ward and diagnosed with Evans syndrome after the following investigations: 1) Haemoglobin=3.3gm/dl 2) Platelets=20,000/mm<sup>3</sup> 3) Direct Coombs test = +ve. Rest of the investigations such as the antinuclear antibodies, antismooth muscle antibody, antimitochondrial antibody, serum hepatoglobin level and R-A factor were all normal. She was vaccinated against H-influenza and pneumonia. She was given 4 units of platelets and 3 units of blood after which her haemoglobin improved from 3.3 gm/dl to 8.8 gm/dl and platelets from 20,000/mm<sup>3</sup> to 69,000/mm<sup>3</sup>. Foetuses were alive and healthy at the time, with normal anomaly scan. Unfortunately the scan images were not saved. Patient was put on a high dose of 40mg prednisolone per day and discharged from hospital.

One month later she was presented in the labour ward with generalised tonic clonic fits, bleeding from gums and an elevated blood pressure. She was diagnosed with eclampsia associated with Evans syndrome. Foetal cardiac activity was normal but termination of the pregnancy was planned considering the mother's health. As there was no contraindication, and a poor foetal outcome was expected, a vaginal delivery was planned. The patient expelled stillborn diamniotic twin male

foetuses, 6-7 hours after induction of labour. No immediate primary postpartum haemorrhaging was found. Patient became conscious within 20 hours after delivery.

Post-delivery, she was kept on the same dose of prednisolone 40mg/d but her platelet count continued to decline to 20,000/mm<sup>3</sup> on the 5th day of postpartum, although her liver enzymes and blood pressure had been normal on the 4th day after delivery. A splenectomy was planned and she was referred to the surgical unit. However after a successful splenectomy, the patient was lost to follow-up. Therefore, her consent could not be sought to submit a report on her rare case. Instead permission was taken from the head of the concerned department.

### Discussion

Evans syndrome is a rare disease that is associated with Coombs positive autoimmune haemolytic anaemia and autoimmune thrombocytopenia. Both these conditions may present simultaneously or sequentially.<sup>5</sup> Most reported cases were diagnosed during childhood. As per our knowledge based on review of medical literature, only a few cases of Evans syndrome during pregnancy have been reported.<sup>6</sup> We treated our case with high dose corticosteroids, same as reported in previous cases. The initial results of treatment were acceptable. Her platelet count increased after treatment and remained stable throughout her pregnancy. Unfortunately after delivery, her platelet count showed a continuous decline so a splenectomy was decided upon.

The main feature of Evans syndrome is ITP, with or without a Coombs positive autoimmune haemolytic anaemia (AIHA).<sup>3</sup> For diagnosis, other causes of thrombocytopenia are ruled out. In our case there was thrombocytopenia with positive Coombs test. Steroid such as prednisone is the initial treatment of choice for Evans syndrome. Chemotherapeutic agents, Splenectomy and plasmaphoresis are other therapies for refractory cases.<sup>3</sup> Glucocorticoids decrease the destruction of platelets and red blood cells (RBCs) by reducing sequestration, and Splenectomy is useful as it entails removing a primary site of antibody production and sequestrations. With Plasmaphoresis, antibody bound platelets and RBCs are replaced with unbound cells without affecting the IgG concentration. On the other hand chemotherapeutic agents inhibit the immune

system thus affecting antibody production.<sup>7</sup> During pregnancy there are no reliable parameters that can predict foetal platelet status or foetal outcome, and even maternal response to treatment that may not end in a desired outcome.<sup>8</sup> The only outcome prediction parameter is the patient's previous history of neonatal outcome.<sup>8,9</sup> The platelet antibody level should be measured in these cases as the platelet antibodies can pass through the placenta and bind with the foetal platelets, resulting in foetal thrombocytopenia. The diagnosis of Evans syndrome in pregnant women does not affect the mode of delivery, which depends on obstetric indications. In our case, normal vaginal delivery was conducted after induction of labour, followed by a referral to general surgery for Splenectomy that was performed successfully.

### Conclusion

Association of Evans syndrome with pregnancy is a very rare disorder and it should be kept in mind for differential diagnosis in patients presenting with unexplained thrombocytopenia during pregnancy. Close follow up, early management, careful planning and preparation for delivery in such women would enhance the chances of a favorable outcome.

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**Conflict of Interest:** None

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