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# Evans with Primary Tuberculosis- A Perilous Amalgam; A Case Report

Dr. Harsha Mudireddy, Dr. Muppidi Shruthi, Dr. K.Ravinder Reddy, Dr. John Israel

*MD, Junior Resident; Department of Internal Medicine , Prathima institute of medical sciences,Karimnagar, Telangana, India*
*MD, Junior Resident ; Department of Internal Medicine, Prathima institute of medical sciences,Karimnagar, Telangana, India*
*MD DNB, Consultant cardiologist,Professor and HOD,(corresponding author)Department of Internal medicine,Prathima institute of medical sciences,karimnagar, Telangana, India*
*MD, Professor, Department of Internal medicine, Prathima institute of medical sciences, karimnagar, Telangana, India*

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**ABSTRACT**
Evans syndrome is an autoimmune condition that refers to the co-occurrence of two or more immune cytopenias, most often, the autoimmune hemolytic anemia (AIHA), immune thrombocytopenia(ITP), and/or autoimmune neutropenia. AIHA is caused by autoantibodies that react with self red blood cells and cause them to be destroyed. Warm AIHA , due to antibodies that are active at body temperature, is the most common type of AIHA. Evans syndrome is considered more difficult to treat than isolated warm AIHA. The co occurrence of Tuberculosis and Evans is an extremely rare and dreadly condition because its makes therapy more challenging as glucocorticoids to treat Evans can increase the risk of tuberculosis infection.

**KEY WORDS:** Evans syndrome, Primary Tuberculosis, Autoimmune hemolytic anemia, Immune thrombocytopenia,Glucocorticoids, Anti tuberculosis treatment(ATT)

## I. CASE REPORT

A 50 year old female presented to the ER with shortness of breath since a week before presentation. Shortness of breath was aggravated on exertion. She also gives a history of fever since a month which was insidious in onset, on and off; cough associated with sputum . This was also associated with easy fatiguabiity, loss of weight since 2 months. She gives history of chestpain only on coughing. No history of hematemesis/malena or any bleeding manifestations was given

On examination patient was conscious, coherent,dyspnoeic. She appeared pale and icteric . Her PR was 120/min, BP 100/70 mm of Hg, RR 30/min, SpO2 91 @ room air, temperature was 101*F. On auscultation , both heart sounds were heard, decreased breath sounds on left suprascapular region. Per abdomen revealed palpable spleen. There was no edema, palpable lymphnodes, or any ecchymosis.

Immediately oxygen support was given and treated symptomatically. A provisional chest xray was done which showed right upper lobe fibrocavitatory lesion. The USG abdomen showed splenomegaly of 16 cm size with hyperechoic lesions in liver –likely hemangioma and in the upper pole of right kidney likely angiomyolipoma. HRCT chest showed cavity lesion in right upper lobe of lung, Tree in bud appearance of both lobes.

Routine blood investigations were sent which showed Hb 4g/dl, WBC 5400/mm3 with neutrophils 79% and lymphocytes 18% and platelets 89000/microL. Liver function test showed total bilirubin 3.3mg/dL, direct bilirubin 1.1mg/dL ,ALT 24U/L, AST 38U/L, ALP 72/L. Renal function test showed blood urea 18 mg/dL, serum creatinine 1.00mg/dL. Lactate dehydrogenase (LDH) was elevated 1470 U/L.

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PRBC transfusion was done immediately. Sputum sample was sent foe gene xpert which turned positive with rifampicin sensitive. Out of suspicion, a Direct coombs test was done which was positive for antibodies against RBC. Viral serologies were negative. ANA, Anti ds DNA, ASMA were negative.

Bone marrow aspiration showed normoblastic picture with normal myelopoiesis, with raised megakaryocytes and suggestive of erythroid hyperplasia.

Bone marrow biopsy revealed 80% cellularity, normal distribution of erythroid and myeloid cells with megaloblastic change. And a reactive marrow.

A presumptive diagnosis of EVANS syndrome concurrent with Primary tuberculosis was made and treatment strategies were discussed.Patient was weighing 50kgs and started with wysolone (prednisolone)40mg , ATT was initiated With Tab. ISONIAZID 300mg/PYRIZINAMIDE 1500 mg/ETHAMBUTOL 1000mg, and iron and folic acid capsules along with B complex tablets, pyridoxine 40mg(1/2 tab), calcium supplements...RIFAMPICIN was not started as it is known to cause immunologically mediated intravascular hemolysis and autoimmune hemolytic anemia.

After specialist opinion,as an alternative to rifampicin treatment, a modified ATT regimen was started with Tab ISONIAZID 300mg/OD, Tab ethambutol 1000mg/OD on inj streptomycin 750mg IM/OD for 2 months followed by Tab ISONIAZID 300 mg/OD and Tab Ethambutol 1000mg/OD for 10 months.Renal function test was monitored after one week of initiation of streptomycin.A total of 4 PRBC transfusions were done. Patient improved and got discharged on day 10 of admission.

she was followed up with complete hemogram and Liver function test every month .After 2 months tab Wysolone (prednisolone) was tapered to 30 mg/OD for one week to 20mg/OD for another week to 10mg/OD for a week followed by 5mg/OD to continue. Her base line hemoglobin remained around 10 g/dl with average platelet count around 1.2L/dL , serum bilirubin around 2mg/dl with direct birlirubin around 0.9mg/dL….for 10 months till now. She has no bleeding manifestations. She has black stools with the effect of iron folic acid tablets. Patient is doing well with no need for blood transfusions till now.

## II. DISCUSSION

Evans syndrome is an autoimmune condition that refers to the co-occurrence of two or more immune cytopenias, most often, the autoimmune hemolytic anemia (AIHA), immune thrombocytopenia(ITP), and/or autoimmune neutropenia (in about 15% patients). AIHA is caused by autoantibodies that react with self red blood cells and cause them to be destroyed. Warm AIHA , due to antibodies that are active at body temperature, is the most common type of AIHA. In ITP, the immune system is directed against GPIIb/IIIa on the platelets.Evans syndrome is a rare condition, diagnosed in 5% and the mean age of diagnosis is 52 years.It could be primary(idiopathic) or secondary to Systemic lupus erthematosus(SLE)/ common variable immunodeficiency(CVID)/ autoimmune lymphoproliferative syndrome(ALPS)/ non hogdkins lymphoma(NHL)/ chronic lymphocytic leukemia(CLL), viral infections like HIV/Hepatitis C and following allogenic transplantation.

Evans syndrome can sometimes be associated with benign liver hemangiomas and kidney angiomyolipomas. There is no proper evidence about this association yet.

TB still remains to be one of the top 10 causes of death worldwide .Primary tuberculosis and Evans syndrome can occur together. They could be predisposing factors for one another. The occurrence of Evans in a TB patient may be due to the production of antibodies against the blood cells by lymphocytes in response to the tubercular pathogen. Molecular mimicry involving unknown antigens of tubercular bacilli, and the platelet surface antigens could be responsible for thrombocytopenia ..TB infection can seriously affect hematopoietic system during its course, with involvement of both myeloid and lymphoid cell lines and plasma components .Patients with tuberculosis have increased risk of infection due to the immunocompromised status and also glucocorticoids given for Evans can increase risk of tuberculosis infection. Patients should receive both ATT and glucocorticoids when tuberculosis and Evans occur together. Other treatment options could be immunosuppresants,intravenous immunoglobulin, blood transfusion, splenectomy, and hematopoietic stem cell transplant. Median survival for Evans syndrome is 7.2 years( primary evans-10.9years, secondary evans 1.7 years). Secondary evans syndrome is associated with higher mortality, with a 5 year survival rate of 38%.

Our patient here is a 50 year female who is a home maker with no comorbidities and addictions and with no family history of autoimmune diseases

gets diagnosed with primary tuberculosis with incidental Evans syndrome. She was initially managed with blood transfusions. Later started on with modified ATT without Rifampicin, as it itself causes hemolysis . Modified ATT for one year with 2 months of inj.streptomycin,tab isoniazide and tab.ethambutol and 10 months of isoniazide and ethambutol along with Prednisolone 40 mg which was tapered to 4mg after 2 months. Proper follow up of the patient with complete hemogram and Liver function tests is mandatory

## III. CONCLUSION

We report a very rare case of a 50 year female with primary tuberculosis with concurrent Evans syndrome , who was cautiously treated with Glucocorticoids and modified ATT. This treatment for this condition is a double- edged sword as glucocorticoids still remains as the first –line treatment for Evans and on the other side it can increase susceptibility for infection to tuberculosis ;and immunosuppression can worsen tuberculosis. Both ATT and glucocorticoids should be given to the patients. Proper evaluation reduces morbidity and regular follow up is mandatory in the treatment protocol for any worsening of the condition or drug adherence or any drug reactions.

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