Original Research Paper

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Medicine

# A CASE OF EVAN'S SYNDROME

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**KEYWORDS :**

**Introduction:**

**Evans syndrome** is a very rare autoimmune disorder in which the immune system destroys the body's red blood cells, white blood cells and/or platelets. Affected people often experience thrombocytopenia and Coombs' positive hemolytic anemia. Signs and symptoms may include purpura, paleness, fatigue, and light-headedness. The exact cause of this condition is unknown.

**Case report:**

Mrs. Sakina Bano Ansari, 34 years old female, resident of Bapunagar, was admitted on 6/7/17 in Emergency Wards of SCL Hospital, Ahmedabad with chief complaints of fever and yellowish discoloration of sclera since 7 days.

Patient was relatively asymptomatic 7 days ago when she developed fever which was low grade, associated with chills and rigors, without evening rise and night sweats, with easy fatigability and weakness. Patient also noticed yellow discoloration of sclera. There was no history of nausea, vomiting, abdominal pain, itching, constipation, skin changes, haematemesis, malena, hair loss, bruising, prolonged bleeding. She had no history of DM, HTN, TB, COPD, Thyriod dysfunction, Blood Tranfusion, previous surgery. There was no significant family history and she had no addictions.

On Examination, she was vitally stable. She was pale, icteric, her spleen was palpable on abdominal examination. There was no hepatomegaly. No lymph nodes were palpable.

On admission, her findings were suggestive of haemolytic anaemia

Hb- 3.9
TC- 10,200
APC- 1000 (on machine) and 5000 (manually)
HCT- 5.3
MCV-131.8
MCH-88.6]
MCHC-67.2
Reticount- 17%.

Peripheral smear showing macrocytic normochromic RBC,many polychromatic RBS and normoblasts

Total Bilirubin- 8.3
Direct Bilirubin- 3.4
Indirect Bilirubin- 4.9
SGPT-20
Urine Routine/ Microscopy was within normal range (BS/BP absent)
Serum Ferritin- 79.8
Serum Vitamin B 12- 186
Serum Iron- 101
Direct Coomb's Test- +++
Indirect Coombs Test- +
Serum C3 Complement- 0.613 (Normal 0.9-1.8)
Serum C4 Complement- 0.0392 (Normal 0.1-0.4)
LDH-771
ANA by IF- Negative
Anticardiolipin Antibody IgG + (>100)
Fundus- Normal
ECG was Normal Sinus Rhythm, Chest Xray was Normal

Patient was diagnosed as Autoimmune Hemolytic Anemia with Immune Medicated Thrombocytopenia (Evan's Syndrome). We transfused 2 Bags of Whole blood and 8 bags of Platelet Rich Concentrate. She was treated with Inj. Methylprednisolone 1gm/day for 5 days. This was followed by oral Prednisolone 1mg/kg/day initially, and Tablet Hydroxychloroquine 200mg/day BD. Patient was regularly monitored with Complete blood counts and Liver function tests.

On discharge on 19/7/17,
Hb 7.9
TC 10,300
APC- 1,63,000
HCT-25.81
MCV- 109
MCH-33.2
MCHC-30.6
S.bilirubin- 2.7
Direct bilirubin- 1.5
Indirect bilirubin-1.2
SGPT-20

During followup, oral prednisolone was tapered off and patient was maintained on Tablet Azathioprine 50mg OD initially, later on BD, and Tablet Hydrochloroquine 200md/day BD.

**Discussion**

Evans syndrome is the direct Coombs-positive autoimmune hemolytic anemia (AIHA) with immune-mediated thrombocytopenia. Evans et al described this condition in 1951.

The pathophysiology of Evans syndrome is unknown. It is considered an autoimmune disorder. Wang et al showed reduced levels of immunoglobulin G (IgG), immunoglobulin M (IgM), and immunoglobulin A (IgA) in these patients

Savasan et al observed that about 50% of people with Evans syndrome had lymphoid hyperactivity. Teachey et al demonstrated that approximately 58% of them might have autoimmune lymphoproliferative syndrome (ALPS).

Apoptosis of activated lymphocytes is regulated by Fas (CD95) and its ligand, and defective expression of either Fas or Fas ligand results in increased production of mature lymphocytes. Some study results suggest that mutations of the Fas gene can cause severe ALPS in humans.

The diagnosis of ES is a diagnosis of exclusion and thrombotic thrombocytopenic purpura should be ruled out. Systemic lupus erythematosus and other autoimmune diseases are associated with Evan's Syndrome.

They have low serum IgG, IgM, and IgA.

No specific role of immunisation in childhood has been noted.

Corticosteroids are the drug of choice (prednisone at 1 to 2 mg/kg per day tapered over a few weeks). For steroid non responsive patients, or steroid dependent (prednisone ≥ 15 mg required), splenectomy can be considered.

Rituximab and immunosuppressors (such as Azathioprine) may be considered in chronic and corticosteroid-dependent ES, even before planning splenectomy. If patient is resistant to rituximab and/or splenectomy, a immunosuppressant is tried (cyclophosphamide, cyclosporin, or azathioprine) depending on the patients clinical profile and disease severity.

Long-term survival data are limited. The most common causes of death were hemorrhage and sepsis. None of these patients developed any malignancy.

## Conclusion

In conclusion, Evans syndrome (ES) is a rare disease characterized by the simultaneous or sequential development of autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia (ITP) and/or immune neutropenia. No cause has been found yet, but it is considered to be autoimmune in origin. Patient presents with mixed symptoms of jaundice and thrombocytopenia.

It shows good response to steroids and immunosuppressive agents. ES is a potentially life-threatening condition that may be associated with other underlying autoimmune or lymphoproliferative disorders.

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