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CASE REPORT: A COMPREHENSIVE REVIEW OF AUTOIMMUNE CYTOPENIAS IN EVANS SYNDROME

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ABSTRACT

Background: Evans syndrome is a rare autoimmune hematological disorder characterized by the simultaneous or sequential occurrence of autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia (ITP), with or without autoimmune neutropenia. The condition is often chronic and relapsing, posing significant diagnostic and therapeutic challenges due to its heterogeneous clinical presentation and association with various autoimmune and immunological disorders. **Case Presentation:** A 32-year-old female presented with progressive fatigue, generalized weakness, easy bruising, gum bleeding, and intermittent jaundice. Clinical examination revealed pallor, scleral icterus, petechiae, ecchymoses, and mild splenomegaly. Laboratory investigations demonstrated severe anemia, thrombocytopenia, reticulocytosis, elevated lactate dehydrogenase, indirect hyperbilirubinemia, and a positive direct antiglobulin test, confirming autoimmune hemolytic anemia. Extensive evaluation excluded secondary causes, including autoimmune diseases, infections, and hematological malignancies. Based on the clinical and laboratory findings, a diagnosis of primary Evans syndrome was established. The patient was treated with prednisolone, intravenous immunoglobulin, packed red blood cell transfusions, platelet transfusion, and supportive therapy, resulting in significant clinical and hematological improvement. **Conclusion:** This case emphasizes the importance of early diagnosis, exclusion of secondary causes, and prompt initiation of immunosuppressive therapy in Evans syndrome. Although favorable outcomes can be achieved with appropriate treatment, long-term follow-up is essential because of the disease's chronic relapsing nature. Individualized management and multidisciplinary care remain key to improving patient outcomes and quality of life.

KEYWORDS: Evans syndrome, autoimmune hemolytic anemia, immune thrombocytopenia, autoimmune cytopenias, direct antiglobulin test, case report.

INTRODUCTION

Evans syndrome is a rare autoimmune hematological disorder characterized by the presence of autoimmune hemolytic anemia and immune thrombocytopenia occurring either simultaneously or sequentially in the same patient. In some cases, autoimmune neutropenia may also be present. The condition was first described by Robert Evans in 1951 and continues to be recognized as a challenging disorder because of its variable clinical presentation and unpredictable disease course. Unlike isolated autoimmune cytopenias, Evans syndrome reflects a broader disturbance of immune regulation, resulting in the destruction of multiple blood cell lines and significant morbidity among affected individuals.[1,2]

The exact prevalence of Evans syndrome remains unknown because of its rarity and frequent underdiagnosis. The disorder can occur in both children and adults, although the clinical course often differs between age groups. Patients may present with symptoms related to anemia, thrombocytopenia, or both, including fatigue, weakness, pallor, jaundice, petechiae, ecchymosis, and mucosal bleeding. The severity of symptoms depends on the degree of blood cell destruction and the rate at which cytopenias develop.

Many patients experience recurrent relapses and remissions, making long-term disease management particularly challenging for both clinicians and patients.[3,4]

The underlying pathophysiology of Evans syndrome is complex and not completely understood. Current evidence suggests that abnormalities in immune tolerance lead to the production of autoantibodies directed against red blood cells, platelets, and occasionally neutrophils. Dysregulation of B lymphocytes, T lymphocytes, and regulatory immune pathways contributes to the persistence of autoimmune activity. In recent years, advances in immunology have identified several genetic and molecular defects associated with immune dysregulation syndromes that may predispose individuals to Evans syndrome. These findings have improved understanding of the disease and highlighted its heterogeneous nature.[5,6]

Diagnosis of Evans syndrome is primarily based on clinical and laboratory evidence of autoimmune hemolytic anemia and immune thrombocytopenia after excluding other potential causes. Laboratory investigations typically reveal anemia, thrombocytopenia, reticulocytosis, elevated lactate dehydrogenase levels, indirect hyperbilirubinemia, and a positive direct antiglobulin test. Because Evans syndrome may occur secondary to autoimmune diseases, infections, primary immunodeficiencies, or lymphoproliferative disorders, a comprehensive diagnostic workup is essential. Accurate diagnosis is important because treatment strategies and long-term outcomes may differ significantly between primary and secondary forms of the disease.[7,8]

Management of Evans syndrome remains challenging because there are no universally accepted treatment guidelines and responses to therapy vary considerably among patients. Corticosteroids are generally considered the first-line treatment and are frequently combined with intravenous immunoglobulin in severe cases. However, many patients experience relapses that require additional therapies such as rituximab, immunosuppressive agents, thrombopoietin receptor agonists, or splenectomy. The present case report describes a patient with primary Evans syndrome who presented with autoimmune hemolytic anemia and severe thrombocytopenia, highlighting the clinical presentation, diagnostic evaluation, therapeutic management, and outcome while reviewing current evidence regarding this uncommon autoimmune disorder.[9,10]

CASE PRESENTATION

Patient Demographics

A 32-year-old female presented to the Department of Hematology of a tertiary care teaching hospital with complaints of progressive fatigue, generalized weakness, easy bruising, and intermittent episodes of jaundice. She was a married homemaker residing in an urban area. The patient belonged to a middle socioeconomic background and had no significant family history of hematological or autoimmune disorders.

Chief Complaints

The patient presented with the following complaints:

- Progressive fatigue for 2 months
- Generalized weakness for 2 months
- Easy bruising over the extremities for 6 weeks
- Gum bleeding while brushing teeth for 3 weeks
- Yellowish discoloration of eyes for 3 weeks
- Exertional shortness of breath for 2 weeks
- Occasional palpitations for 2 weeks
- Dizziness and lightheadedness for 10 days

History of Present Illness

The patient was apparently healthy until two months prior to presentation when she began experiencing progressive fatigue and generalized weakness that gradually interfered with her daily activities. Initially, she attributed the symptoms to work-related stress and inadequate sleep. Over the following weeks, she noticed increasing breathlessness on exertion and episodes of dizziness while performing household activities.

Approximately six weeks before admission, she developed spontaneous bruising over both upper and lower limbs without any history of trauma. Three weeks later, she experienced recurrent gum bleeding while brushing her teeth and noticed yellow discoloration of her eyes. The patient also reported dark-colored urine and occasional palpitations. There was no history of fever, weight loss, night sweats, bone pain, recurrent infections, hematuria, melena, hematemesis, or neurological symptoms. Due to worsening weakness and persistent bleeding manifestations, she sought medical attention and was subsequently referred to the hematology department for further evaluation.

Medical History

The patient had no known history of any disease. She had never been hospitalized previously for any major illness.

Medication History

The patient denied regular use of any prescription medications. No recent vaccinations had been administered before symptom onset.

Family History
There was no known family history of any disease.

Social History
The patient was a non-smoker and denied any history of alcohol consumption or recreational drug use. She followed a mixed diet and reported adequate nutritional intake. There was no history of recent travel, exposure to infectious diseases, or contact with individuals suffering from chronic illnesses. She lived with her husband and children in a hygienic household environment.

Occupational History
The patient was a homemaker with no occupational exposure and there was no history suggestive of occupational hazards contributing to her current illness.

Allergic History
The patient reported no known allergies.

General Examination
On admission, the patient appeared ill-looking, pale, and mildly icteric. She was conscious, alert, and oriented to time, place, and person.

Vital Signs

Parameter	Finding
Temperature	36.8°C
Pulse Rate	104 beats/min
Blood Pressure	108/68 mmHg
Respiratory Rate	20 breaths/min
Oxygen Saturation	98% on room air

Systemic Examination
Skin and Mucous Membranes
- Marked pallor
- Multiple petechial spots over both lower limbs
- Ecchymotic patches over forearms and thighs
- Mild scleral icterus
- Bleeding gums

Lymphatic System
- No palpable lymphadenopathy

Abdominal Examination
- Mild splenomegaly (2 cm below left costal margin)
- No hepatomegaly
- No ascites

Cardiovascular System
- Tachycardia
- Normal heart sounds
- No murmurs

Respiratory System
- Bilateral air entry present
- No adventitious sounds

Central Nervous System
- No focal neurological deficits
- Higher mental functions intact

Laboratory Investigations
Complete Blood Count

Parameter	Result	Reference Range
Hemoglobin	6.8 g/dL	12–16 g/dL
RBC Count	2.4×10^{12}/L	$4.0–5.2 \times 10^{12}$/L
Hematocrit	21%	36–46%
MCV	88 fL	80–100 fL
MCH	29 pg	27–33 pg
MCHC	33 g/dL	32–36 g/dL
Total Leukocyte Count	5.6×10^9/L	$4–11 \times 10^9$/L
Platelet Count	18×10^9/L	$150–450 \times 10^9$/L

Reticulocyte Studies

Parameter	Result	Reference Range
Reticulocyte Count	8.5%	0.5–2.5%
Corrected Reticulocyte Count	6.2%	0.5–2.0%

Hemolytic Analysis

Parameter	Result	Reference Range
Lactate Dehydrogenase (LDH)	720 U/L	120–250 U/L
Total Bilirubin	3.2 mg/dL	0.3–1.2 mg/dL
Direct Bilirubin	0.7 mg/dL	<0.3 mg/dL
Indirect Bilirubin	2.5 mg/dL	<1.0 mg/dL
Haptoglobin	Undetectable	Normal
Direct Coombs Test (DAT)	Positive	Negative
Indirect Coombs Test	Positive	Negative

Peripheral Blood Smear
Microscopic examination revealed:
- Normocytic normochromic anemia
- Polychromasia
- Numerous spherocytes
- Anisopoikilocytosis
- Marked thrombocytopenia
- Adequate leukocyte morphology
- No blast cells or abnormal infiltrates

Coagulation Profile

Parameter	Result	Reference Range
Prothrombin Time (PT)	13.2 sec	11–15 sec
INR	1.1	0.8–1.2
Activated Partial Thromboplastin Time (aPTT)	31 sec	25–35 sec

Biochemical Investigations

Parameter	Result	Reference Range
Serum Creatinine	0.8 mg/dL	0.6–1.2 mg/dL
Blood Urea Nitrogen	18 mg/dL	7–20 mg/dL
AST	32 U/L	<40 U/L
ALT	28 U/L	<40 U/L
Alkaline Phosphatase	96 U/L	44–147 U/L
Serum Albumin	4.0 g/dL	3.5–5.0 g/dL

Autoimmune Profile

Investigation	Result
ANA	Negative
Anti-dsDNA	Negative
Complement C3	Normal
Complement C4	Normal
Rheumatoid Factor	Negative

Infectious Disease Screening

Test	Result
HIV 1 & 2	Negative
HBsAg	Negative
Anti-HCV	Negative
EBV Serology	Negative
CMV Serology	Negative

Bone Marrow Aspiration and Biopsy

Bone marrow examination showed:

- Hypercellular marrow
- Erythroid hyperplasia
- Increased megakaryocytes
- Adequate myeloid maturation
- No dysplastic changes
- No evidence of leukemia, lymphoma, or marrow infiltration

FINAL DIAGNOSIS

Based on the patient's clinical presentation, hematological findings, and comprehensive diagnostic evaluation, a final diagnosis of Primary Evans Syndrome (ES) was established. The diagnosis was supported by the coexistence of warm autoimmune hemolytic anemia (AIHA), evidenced by severe anemia, reticulocytosis, elevated lactate dehydrogenase, indirect hyperbilirubinemia, reduced haptoglobin levels, spherocytosis on peripheral smear, and a positive direct antiglobulin (Coombs) test, along with immune thrombocytopenia (ITP) manifested by severe thrombocytopenia and mucocutaneous bleeding. Extensive investigations excluded secondary causes, including autoimmune disorders, infections, lymphoproliferative malignancies, and immunodeficiency syndromes, confirming the diagnosis of primary Evans syndrome.

DIFFERENTIAL DIAGNOSIS

Several conditions were considered before establishing the diagnosis of Evans syndrome. Systemic lupus erythematosus (SLE) was excluded due to negative antinuclear antibody and anti-double-stranded DNA tests. Autoimmune lymphoproliferative syndrome (ALPS) and common variable immunodeficiency (CVID) were considered but were unsupported by the clinical and laboratory findings. Thrombotic thrombocytopenic purpura (TTP) was ruled out due to the absence of schistocytes, neurological manifestations, and renal dysfunction. Hematological malignancies, including lymphoma and leukemia, were excluded through bone marrow examination. Infectious causes such as HIV, hepatitis B, hepatitis C, Epstein–Barr virus, and cytomegalovirus infections were also ruled out through serological testing.

TREATMENT AND MANAGEMENT

Upon admission, the patient was closely monitored in the hematology unit because of severe anemia (hemoglobin 6.8 g/dL) and profound thrombocytopenia (platelet count 18×10^9/L) associated with active mucosal bleeding.

Initial management focused on stabilizing the patient and preventing life-threatening complications. Vital signs, bleeding manifestations, complete blood counts, and hemolytic markers were monitored regularly. The patient was advised strict bed rest during the acute phase, avoidance of intramuscular injections, and implementation of bleeding precautions. Adequate hydration was maintained with intravenous normal saline as required. Nutritional support and counseling were provided to optimize recovery and improve overall health status during treatment.

Due to symptomatic severe anemia accompanied by fatigue, dyspnea on exertion, dizziness, and tachycardia, transfusion support was initiated. The patient received 2 units of leukocyte-reduced packed red blood cells (PRBCs) administered slowly under close supervision.

Since patients with autoimmune hemolytic anemia often have pan-reactive antibodies that complicate cross-matching, the least incompatible blood units were selected after consultation with the transfusion medicine department. The patient tolerated the transfusions well without any transfusion-related adverse reactions. Following transfusion, hemoglobin levels improved gradually, resulting in significant relief of symptoms and enhanced functional capacity.

Because the patient presented with severe thrombocytopenia and active gum bleeding, platelet transfusion was administered as an emergency supportive measure. She received one therapeutic adult dose of single-donor platelet concentrate (approximately 3×10^{11} platelets). Although platelet transfusions in immune thrombocytopenia are generally reserved for significant bleeding episodes due to rapid immune-mediated destruction, they can be lifesaving when clinically indicated. The transfusion successfully controlled active bleeding and provided temporary hemostatic support while definitive immunosuppressive therapy was initiated. No transfusion reactions or

complications occurred during or after platelet administration.

First-Line Corticosteroid Therapy

Systemic corticosteroids were initiated as first-line treatment for Evans syndrome. The patient received Prednisolone 1 mg/kg/day orally (60 mg once daily).

Corticosteroids suppress autoantibody production, reduce macrophage-mediated destruction of antibody-coated blood cells, and help restore hematological parameters. Clinical improvement was noted within one week of therapy, with a gradual rise in hemoglobin and platelet counts. The treatment was continued at full dose for four weeks, followed by a slow taper over several months based on clinical response and laboratory monitoring. Blood pressure, blood glucose levels, body weight, and potential corticosteroid-related adverse effects were regularly assessed throughout treatment.

Intravenous Immunoglobulin Therapy

Given the severe thrombocytopenia and active mucosal bleeding, Intravenous Immunoglobulin (IVIG) was administered at a dose of 1 g/kg/day intravenously for two consecutive days. IVIG acts by saturating Fc receptors on macrophages and reducing immune-mediated platelet destruction. The patient demonstrated a favorable response with a rapid increase in platelet count within several days. IVIG was particularly useful in achieving early hemostatic control while awaiting the full therapeutic effect of corticosteroids. The infusion was administered slowly under close monitoring for adverse events, including headache, fever, chills, hypertension, and infusion-related reactions, none of which occurred.

Folic Acid Supplementation

Because chronic hemolysis results in increased erythropoietic activity and higher folate consumption, the patient was prescribed Folic Acid 5 mg orally once daily. Folate supplementation supports effective erythropoiesis and prevents the development of folate deficiency secondary to ongoing red blood cell destruction and bone marrow compensation. The medication was continued throughout the treatment period and follow-up phase.

Adequate dietary intake of folate-rich foods was also encouraged to support hematologic recovery and maintain normal red blood cell production.

Gastrointestinal and Bone Protection

Considering the prolonged use of high-dose corticosteroids, prophylactic measures were implemented to minimize treatment-related complications. The patient received Pantoprazole 40 mg orally once daily to reduce the risk of gastritis, peptic ulcer disease, and gastrointestinal bleeding associated with corticosteroid therapy. To prevent steroid-induced osteoporosis, she was prescribed Calcium Carbonate 500 mg orally twice daily and Vitamin D3 (Cholecalciferol) 60,000 IU orally once weekly for eight weeks, followed by monthly maintenance supplementation. Regular assessment of bone health and counseling regarding physical activity were also provided.

Monitoring and Follow-Up Management

The patient underwent regular monitoring with complete blood counts, reticulocyte counts, liver function tests, serum lactate dehydrogenase, bilirubin levels, and coagulation parameters. Weekly assessments were performed during the first month, followed by monthly follow-up visits. Hemoglobin improved from 6.8 g/dL to 11.2 g/dL, while platelet counts increased from 18 × 10^9/L to 168 × 10^9/L after one month of therapy. The patient achieved hematological remission with resolution of bleeding manifestations and jaundice. Prednisolone tapering was initiated gradually, and long-term follow-up was planned to monitor for relapse, a common feature of Evans syndrome.

S. No.	Drug/Therapy	Dose	Route	Frequency	Duration	Indication
1	Prednisolone	1 mg/kg/day (60 mg/day)	Oral	Once daily	4 weeks followed by gradual taper over 3–6 months	Autoimmune hemolytic anemia and immune thrombocytopenia
2	Intravenous Immunoglobulin (IVIG)	1 g/kg/day	Intravenous infusion	Once daily	2 consecutive days	Severe thrombocytopenia with active mucosal bleeding
3	Packed Red Blood Cells (PRBCs)	2 units (least incompatible, leukocyte reduced)	Intravenous transfusion	As required	Single transfusion episode	Severe symptomatic anemia (Hb 6.8 g/dL)
4	Single Donor Platelet Concentrate (SDP)	1 therapeutic adult dose (~3 × 10^{11} platelets)	Intravenous transfusion	As required	Single transfusion episode	Severe thrombocytopenia with active bleeding
5	Folic Acid	5 mg	Oral	Once daily	Continued throughout	Chronic hemolysis

					treatment and follow-up	
6	Pantoprazole	40 mg	Oral	Once daily	Until corticosteroid taper completion	Gastroprotection during corticosteroid therapy
7	Calcium Carbonate	500 mg	Oral	Twice daily	Throughout corticosteroid treatment	Prevention of steroid-induced bone loss
8	Vitamin D3 (Cholecalciferol)	60,000 IU	Oral	Once weekly	8 weeks, followed by monthly maintenance	Prevention of steroid-induced osteoporosis
9	Normal Saline (0.9% Sodium Chloride)	75–100 mL/hour (as clinically indicated)	Intravenous	Continuous	During hospitalization	Supportive care

DISCUSSON

The present case highlights the classical presentation of Evans syndrome, a rare autoimmune disorder characterized by the coexistence of autoimmune hemolytic anemia and immune thrombocytopenia. The patient presented with fatigue, jaundice, easy bruising, gum bleeding, and severe anemia, all of which are commonly reported manifestations of this condition. Laboratory findings, including a positive direct antiglobulin test, elevated lactate dehydrogenase, indirect hyperbilirubinemia, and severe thrombocytopenia, strongly supported the diagnosis. Similar clinical features were described by Jaime-Pérez et al., who reported fatigue, pallor, jaundice, and mucosal bleeding as the most frequent presenting symptoms in Evans syndrome patients. Likewise, Maqsood et al. emphasized that simultaneous anemia and thrombocytopenia often lead to significant diagnostic challenges due to the rarity and heterogeneity of the disease.[11,12]

The diagnostic process in this case required careful exclusion of secondary causes of cytopenias. The patient underwent extensive investigations to rule out systemic lupus erythematosus, viral infections, lymphoproliferative disorders, and bone marrow pathology before confirming primary Evans syndrome.

This approach is consistent with current recommendations because Evans syndrome remains a diagnosis of exclusion. Audia et al. noted that nearly half of adult cases may be associated with autoimmune diseases, malignancies, or immunodeficiency disorders, making comprehensive evaluation essential. Similarly, the recent case review by Palvia et al. emphasized that disorders such as thrombotic thrombocytopenic purpura, myelodysplastic syndrome, systemic lupus erythematosus, and viral infections should always be excluded before establishing the diagnosis of Evans syndrome.[13,14]

The favorable response observed in this patient following treatment with prednisolone, intravenous immunoglobulin, blood transfusion, and supportive therapy reflects the effectiveness of first-line management strategies in Evans syndrome.

Corticosteroids remain the cornerstone of treatment because they suppress the abnormal immune response responsible for blood cell destruction. Intravenous immunoglobulin is particularly useful in patients presenting with severe thrombocytopenia and active bleeding. In a multicenter study involving adult Evans syndrome patients, Fattizzo et al. found that most patients initially responded to corticosteroids, although many required additional therapies due to disease relapse. Similarly, Jaime-Pérez et al. reported that corticosteroids and IVIG remain the most widely accepted initial treatment modalities for achieving rapid hematological stabilization and symptom control.[15]

Despite achieving remission, long-term follow-up remains essential because Evans syndrome is characterized by frequent relapses and an unpredictable clinical course. Patients often require prolonged monitoring for recurrent cytopenias, treatment-related adverse effects, infections, and thrombotic complications. The chronic nature of the disease can significantly affect quality of life and may necessitate second-line therapies such as rituximab, mycophenolate mofetil, cyclosporine, or splenectomy. Norton and colleagues described Evans syndrome as a chronic disorder marked by repeated exacerbations and remissions, emphasizing the need for individualized long-term treatment strategies. Similarly, Erolmez and colleagues highlighted that adult Evans syndrome patients remain at risk for significant thrombotic complications, particularly in the presence of persistent hemolysis and additional risk factors.[16,17]

CONCLUSION

Evans syndrome is a rare but potentially serious autoimmune disorder in which the body's immune system mistakenly attacks its own red blood cells and platelets, leading to anemia, bleeding tendencies, and

other complications. This case highlights the importance of early recognition of symptoms such as fatigue, jaundice, easy bruising, and mucosal bleeding, followed by a thorough diagnostic evaluation to exclude secondary causes. Prompt initiation of appropriate therapy, including corticosteroids, intravenous immunoglobulin, transfusion support, and close monitoring, can result in significant clinical and hematological improvement. However, because Evans syndrome often follows a chronic relapsing course, long-term follow-up is essential to detect recurrences and manage complications. A multidisciplinary approach and individualized treatment strategy remain crucial for achieving favorable patient outcomes and improving quality of life.

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