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Chapter 10

Evans Syndrome: A Rare Case of Concurrent Autoimmune Haemolytic Anaemia and Immune Thrombocytopenia

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Abstract

Evans syndrome (ES) is a rare condition characterized by the concurrent or sequential development of autoimmune haemolytic anaemia (AIHA), immune thrombocytopenia (ITP), and/or immune neutropenia. It is marked by frequent relapses, a significant treatment burden, and increased risks of infection and thrombosis, substantially impacting survival. ES can be primary or secondary to conditions such as lymphoproliferative disorders, systemic autoimmune diseases, or primary immunodeficiencies. The syndrome severely affects survival and quality of life and is often severe and potentially fatal. Effective management requires prompt therapy, including anti-infectious and anti-thrombotic prophylaxis.

Keywords: Evans Syndrome, Autoimmune Haemolytic Anaemia, Idiopathic Thrombocytopenic Purpura, NSAIDs, Autoimmune Disorder, Hematologic Disease

1. Introduction

Evans syndrome (ES), first described in 1951, is a rare autoimmune condition characterized by the coexistence of immune thrombocytopenic purpura (ITP) and Coombs-positive autoimmune haemolytic anaemia (AIHA) [1–3]. In this disorder, the body produces autoantibodies targeting red blood cells (RBCs), white blood cells (WBCs), and platelets, leading to symptoms related to anemia, leukopenia, or thrombocytopenia. Diagnosis requires excluding alternative conditions such as thrombotic microangiopathies, vitamin deficiencies, myelodysplastic syndromes, paroxysmal nocturnal haemoglobinuria, bleeding-related anaemia complicating ITP, or haemolysis, elevated liver enzymes, and low platelets (HELLP) syndrome in pregnancy [4]. AIHA in ES is typically driven by warm antibodies, predominantly IgG (rarely IgA), ruling out cold agglutinins [5]. Autoimmune neutropenia (AIN) affects approximately 15% of adults and 20% of children with ES [6]. The condition is more prevalent in boys than girls (1.4:1 ratio) in children, while in adults, women are more commonly affected [7, 8].

A retrospective study in Denmark (1977–2017, n=242) reported an annual prevalence of 21.3 per million and an incidence of 1.8 per million person-years [9]. The widespread use of non-steroidal anti-inflammatory drugs (NSAIDs) may contribute to the increased incidence of ES [10].

2. Case Presentation

A 42-year-old male presented to the outpatient department (OPD) of a tertiary medical college and research centre with complaints of fever with chills, general weakness, anxiety (”Ghabrahat”), and epigastric discomfort. He had a history of head trauma 15–20 days prior and was a known case of type 2 diabetes mellitus (T2DM). He denied cough, burning micturition, sore throat, loose stools, or melena. The patient was normotensive, euthyroid, an occasional cigarette smoker, and denied alcohol or drug use. Physical examination revealed pale conjunctivae.

Ultrasound of the whole abdomen showed an enlarged liver (16 cm, mild hepatomegaly with parenchymal changes), splenomegaly, and minimal ascites.

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3. Lab Investigation

The following table summarizes the laboratory findings:

Test Name	Result	Normal Range
Hemoglobin	3.8 g/dL	13.0–17.0 g/dL
Neutrophils	81%	40–80%
Lymphocytes	11%	20–40%
Eosinophils	0%	1–6%
Platelets	60,000/mcL	150,000–450,000/mcL
PCV	11%	42–52%
RDW	25%	4–16%
Bilirubin Total	1.5 mg/dL	0.2–1.0 mg/dL
Bilirubin Indirect	1.2 mg/dL	0.0–0.7 mg/dL
Total Protein	6.30 g/dL	6.6–8.3 g/dL
Globulin	2.7 g/dL	2.8–4.5 g/dL
Sodium	128.9 mEq/L	135–155 mEq/L
Reticulocyte Count	4.7%	0.5–2.0%
Direct Coombs Test	Positive	Negative

Based on the chief complaints, chest X-ray, ECG, and laboratory investigations strongly suggestive of AIHA, the patient was diagnosed with Evans syndrome.

4. Treatment

The patient received an initial transfusion of one pint of packed red blood cells (PRBC) and was closely monitored with antibiotics, iron tablets, and vitamin B12 supplementation. Blood parameters showed some improvement. On days 4 and 7, he received additional PRBC transfusions (one pint each). A second antibody therapy was initiated, and the steroid dosage was tapered. The treatment yielded positive results, with the patient reporting improvement and eventual discharge from the hospital.

5. Discussion

Evans syndrome, first described by Robert Evans in 1951, is a rare autoimmune disorder characterized by the simultaneous or sequential occurrence of autoimmune haemolytic anaemia (AIHA) and immune thrombocytopenia (ITP) [11–13]. Diagnosis requires a high index of suspicion and exclusion of other causes of cytopenias [14]. The condition results from autoantibodies targeting RBCs and platelets, driven by T-cell activation and autoantibody production against specific antigens [15].

Diagnosis involves identifying typical symptoms, obtaining a detailed patient history, clinical examination, and specialized tests, as there is no definitive test for ES. It is a diagnosis of exclusion, confirmed when AIHA (with a positive direct Coombs test) and ITP coexist, even if not simultaneously. Common diagnostic tools include a complete blood count (CBC) to detect anemia and thrombocytopenia, a bone marrow examination to rule out underlying marrow disorders, and a direct antiglobulin test (DAT) to confirm autoantibodies on RBCs.

Treatment typically involves immunosuppressive therapies, such as corticosteroids and rituximab, and, in some cases, splenectomy. The choice of therapy depends on disease severity and patient response. ES is often chronic and relapsing, with significant morbidity and mortality risks due to underlying autoimmune processes. However, appropriate treatment can lead to long-term remission and improved quality of life.

In summary, Evans syndrome is a rare autoimmune disorder characterized by the coexistence of AIHA and ITP. Diagnosis relies on laboratory and clinical findings, with treatment involving immunosuppressive therapies and, occasionally, splenectomy. While the prognosis is generally poor, effective management can achieve long-term remission.

6. Conclusion

Evans syndrome is a complex autoimmune disorder with an unclear etiology and variable pathophysiology. Its prognosis is poor, and treatment strategies are not well-defined. Early diagnosis and prompt treatment are critical for improving patient outcomes.

Conflicts of interest

The authors declare no conflicts of interest.

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