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

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Case Report

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Abstract

Immunological thrombocytopenia (ITP), immune neutropenia, and/or autoimmune haemolytic anaemia (AIHA) can develop concurrently or sequentially in Evans syndrome (ES), an uncommon illness. Frequent relapses, a heavy therapeutic load, and an elevated risk of infection and thrombosis are its hallmarks, all of which have a substantial impact on survival. Several illnesses, including as lymphoproliferative disorders, other systemic autoimmune diseases, and primary immunodeficiencies, can be categorized as primary or secondary to ES. The syndrome has a major influence on survival and quality of life and is frequently severe and sometimes fatal. Effective management of ES requires prompt therapy, including anti-infectious and anti-thrombotic prophylaxis.

Keywords: Evans Syndrome; Autoimmune Haemolytic Anaemia; Idiopathic Thrombocytopenic Purpura; Nsaids; Autoimmune Disorder; Hematologic Disease

Abbreviations: ES: Evans Syndrome, ITP: Immunological Thrombocytopenia, AIHA: Autoimmune Haemolytic Anaemia, IAHA: Idiopathic Autoimmune Haemolytic Anemia.

Introduction

A relatively rare autoimmune condition called Evans syndrome (ES) was originally identified in 1951. It is the result of combining immune thrombocytopenic purpura (ITP) with Coomb positive idiopathic autoimmune haemolytic anemia (IAHA) [1-3]. The body produces antibodies against its own red blood cells (RBCs), white blood cells (WBCs), and platelets in this uncommon illness. People may exhibit indications and symptoms as a result of insufficient platelets, leukocytes, or anemia. When AIHA and ITP occur simultaneously, the diagnosis has to rule out other possibilities like thrombotic microangiopathies, vitamin deficiencies, myelodysplastic syndromes, paroxysmal nocturnal haemoglobinuria, anaemia from bleeding that complicates ITP, or certain conditions like haemolysis, elevated liver enzymes, and low platelets (HELLP) when they occur during pregnancy [4]. The cause of ES-anemia, an AIHA, is warm antibodies, typically of the IgG isotype (rarely of the IgA isotype), ruling out cold agglutinins [5]. Another component of ES is autoimmune neutropenia (AIN), which affects 15% of adults and 20% of children [6]. At a ratio of 1.4 to 1, it is more common in boys than in girls, and in adults, women appear to suffer more than men [7,8]. According to a statewide retrospective study carried out in Denmark, which included 242 patients treated between 1977 and 2017, the annual prevalence of Evans syndrome was 21.3/million people, and the annual incidence was 1.8/million person-years [9]. Non-steroidal anti-inflammatory medicines (NSAIDs) are widely available and used, which may be a factor in the elevated incidence of Evans syndrome [10].

Case Presentation

A 42 years old male came to the OPD with the chief complaints of Fever with chills, general weakness, Ghabrahat, Epigastric discomfort to the tertiary medical college and research Centre. He had a history of head trauma -15-20 days ago and was a known case of T2DM.He had a no history of cough, burning micturition, sore throaty, loose stool and melena. HE was normotensive and euthyroid. He occasionally smoked cigarettes, he denies consuming alcohol or drugs. On physical examination with pale conjunctivitis is found.

On the ultrasound of whole abdomen liver is found enlarged in size measuring approx. 16 cm with altered echotexture (mild hepatomegaly with liver parenchymal changes), spleen is enlarged in sized and minimal ascites is found.

Lab Investigation

Test Name	Result	Normal Range
Hemoglobin	3.8	13.0-17.0 mg/dl
Neutrophils	81	40-80 %
Lymphocytes	11	20-40%
Eosinophils	0	1-6 %
Platelets	60	150000-450000/mcl
PCV	11	42-52%
RDW	25	4-16%
Bilibiruin total	1.5	0.2-1.0 mg/dl
Bilirubin indirect	1.2	0.0-0.7 mg/dl
Total Protein	6.3	6.6-8.3 g/dl
Globulin	2.7	2.8-4.5g/dl
Sodium	128.9	135-155 mEg/L
Reticulocyte count	4.7	0.5-2.0 %
Direct COOM	positive	Negative

Table 1: Chest X-ray, ECG and lab investigations strongly suggestive for AIHA.

Treatment

Following the investigation, he received a pint of packed blood cells for his initial symptoms, and he was closely monitored with the administration of antibiotics, iron tablets, and vitamin B12. His blood parameters showed some improvement. On days 4 and 7, he received another transfusion of 1 pint of packed cell volume. A second antibody was started, and the dosage of steroids was reduced. There was some improvement in his blood parameters. The above treatment produced a positive result. He began to feel better over time, and he was allowed to leave the hospitals.

Discussion

Evans Syndrome was first described in 1951 by Robert Evans. The diagnosis is infrequent and requires a high index of suspicion with exclusion of other disorders characterized by autoimmune hemolytic anemia and thrombocytopenia [11-13].

Evans syndrome is a rare autoimmune illness defined by the simultaneous or sequential onset of at least two autoimmune cytopenias, usually autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia. Evans syndrome is diagnosed when both AIHA and ITP are present, with autoimmune hemolytic anemia being the most common presentation [14].

Evans syndrome is caused by the formation of autoantibodies against red blood cells and platelets, which destroy them and cause anemia and thrombocytopenia. T-cells are activated and autoantibodies are produced to target particular antigens on red blood cells and platelets [15].

Evans syndrome should be considered in situations when test results indicate a severely low platelet count and low haemoglobin level. Evans syndrome diagnosis requires identifying typical symptoms, a full patient history, clinical examination, and specialized testing. There is no clear test for Evans syndrome, and the diagnosis is determined by ruling out alternative possibilities. Evans syndrome is a diagnosis of exclusion. Evans syndrome can be diagnosed when both autoimmune haemolytic anaemia (with a positive direct Coombs test) and ITP occur in the same patient, even if they do not develop simultaneously [16].

A complete blood count (CBC) to check for anaemia and thrombocytopenia, a bone marrow examination to assess for any underlying bone marrow disorders, and a direct antiglobulin test (DAT) to detect the presence of autoantibodies on red blood cells are commonly used in the diagnosis of Evans syndrome. Evans syndrome is usually treated with a mix of immunosuppressive medications, including rituximab and corticosteroids, and, in certain situations, a splenectomy. The severity of the illness and the patient's reaction to treatment determine the therapeutic option. Evans syndrome has a poor prognosis since it is usually a chronic, recurrent condition with a high risk of morbidity and death because of the underlying autoimmune processes. However, many individuals can achieve long-term remission and a better quality of life with the right therapy.

In summary, Evans syndrome is a rare autoimmune disorder characterized by the simultaneous occurrence of

AIHA and ITP. The diagnosis is based on laboratory tests and clinical features, and treatment typically involves a combination of immunosuppressive therapies and splenectomy. The prognosis is generally poor, but with appropriate treatment, many patients can achieve long-term remission and improved quality of life.

To summarize, Evans syndrome is an uncommon autoimmune disease characterized by the co-occurrence of ITP and AIHA. Laboratory testing and clinical symptoms are used to make the diagnosis, and immunosuppressive medications and splenectomy are usually used in conjunction for therapy. Although the prognosis is usually dismal, many individuals can experience a better quality of life and a long-term remission with the right care.

Conclusion

Evans syndrome is a mysterious illness with a vague etiology and a varied pathophysiology. Its prognosis is bad and its course of therapy is unclear. For improved patient outcomes, early identification and treatment are necessary.

Conflicts of Interest: The authors declare no conflict of interest.

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