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Case report on Evans syndrome

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***Abstract*: Evans syndrome is a rare disorder in which the body's immune system produces antibodies that mistakenly destroy red blood cells, platelets and sometimes certain white blood cell known as neutrophils. This leads to abnormally low levels of these blood cells in the body (cytopenia). The premature destruction of red blood cells (hemolysis) is known as autoimmune hemolytic anemia or AIHA. Thrombocytopenia refers to low levels of platelets (idiopathic thrombocytopenia purpura or ITP in this instance). Neutropenia refers to low levels of certain white blood cells known as neutrophils. Evans syndrome is defined as the association of AIHA along with ITP; neutropenia occurs less often. In some cases, autoimmune destruction of these blood cells occurs at the same time (simultaneously); in most cases, one condition develops first before another condition develops later on (sequentially). The symptoms and severity of Evans syndrome can vary greatly from one person to another. Evans syndrome can potentially cause severe, life-threatening complications. Evans syndrome may occur by itself as a primary (idiopathic) disorder or in association with other autoimmune disorders or lymphoproliferative disorders as a secondary disorder. (Lymphoproliferative disorders are characterized by the overproduction of white blood cells.) The distinction between primary and secondary Evans syndrome is important as it can influence treatment. Treatment for Evans syndrome depends on many factors, including the severity of the condition; the signs and symptoms present; and each person's response to certain therapies. For example, people who need to be hospitalized due to severe anemia or thrombocytopenia are often treated with blood transfusions followed by therapy with corticosteroids or intravenous (IV) immune globulin. Other treatment options include immunosuppressive drugs. Most affected individuals respond to these treatments; however, relapse is frequent.**

***Keywords:* Evans syndrome, Autoimmune Hemolytic Anemia (AIHA), Idiopathic thrombocytopenia purpura (ITP), Corticosteroid.**

1. INTRODUCTION

Evans syndrome was first described in the medical literature in 1951 by Dr. Robert Evans and associates. For years, the disorder was considered a coincidental occurrence of AIHA with thrombocytopenia and/or neutropenia. However, researchers now believe that the disorder represents a distinct condition characterized by a chronic, profound (more than in ITP or AIHA alone) state of immune system malfunction (dysregulation). The symptoms and severity of Evans syndrome can vary greatly from one person to another as can the onset, course and duration of the disorder. Most individuals exhibit a chronic course with periods of worsening symptoms (exacerbation) and remissions usually induced transiently by treatment. Most symptoms are caused by low levels of specific blood cells in the body. Some individuals with Evans syndrome may first present with accelerated destruction of red blood cells faster than the body can replace them. Low levels of circulating red blood cells, known as anemia, can cause a variety of symptoms including fatigue, pale skin color (pallor), lightheadedness, shortness of breath, dark colored urine, and a rapid heartbeat. Some individuals may develop yellowing of the skin and especially the whites of the eyes (jaundice).

Other individuals may first present with low levels of platelets, known as thrombocytopenia. Thrombocytopenia may cause tiny reddish or purple spots on the skin (petechiae), larger purplish discoloration on the skin caused by bleeding from ruptured blood vessels into subcutaneous tissue (ecchymosis), and purpura, a rash consisting of purple spots cause by internal bleeding from small blood vessels. Affected individuals may be more susceptible to bruising following minimal injury and spontaneous bleeding from the mucous membranes.

2. CASE REPORT

A 47 years old female patient presented with generalized weakness, intermittent chills and rigors, easy fatiguability and burning sensation on hip in the last 1 month.

On physical examination she has shown the sign of pallor and bilateral pitting edema, reduced appetite and sever weight lost, further examinations found were splenomegaly and hepatomegaly, autoimmune hemolytic anemia (AIHA) and thrombocytopenia

Patient is a known case of type 2 diabetic mellitus (T2DM) and had a history of several blood transfusion in other hospitals but the blood count test was still abnormal and remarkably low especially neutrophil count (20%)

Hematologic test (complete blood count test) (CBC), revealed remarkable reduction in number of Red blood cells (RBC)=6.8g/dl (AIHA), white blood cells (WBC)=1180/mm3 (neutropenia/pancytopenia) and platelet count =5000/mm3

Liver function test (LFT) shows a very high LDH=759U/L and very low AST=9U/L

From the subjective and objective data patient was diagnosed with Evans syndrome and the treatment has been continued with another multiple blood transfusion section and injection piperacillin 4.5g/100mlNS, Tablet paracetamol 500mg 3 times a day, vitamin B complex capsule, and insulin injection to control DM.

3. DISCUSSION

According to the general guidelines and standard practical's treatment of Evans syndrome, first line therapy is corticosteroids such as prednisolone which is used to reduce the recurrence of autoimmune Hemolytic anemia (AIHA) and thrombocytopenia. In case of inadequate response in refractory cases, the physician may prescribe cyclosporine and in very rare cases Azathioprine, in this case report corticosteroid was not prescribed and the treatment used was blood transfusion and even after final diagnosis (Evans syndrome) ongoing blood transfusion was administered with no additional corticosteroid or immunosuppressant therapy. Studies elicit high remission rate of AHIA among the patient with Evans syndrome who receive corticosteroids and cyclosporine. Blood transfusion is among the last line therapy for very severe or refractory cases in contrast with this case whereby the treatment introduced were multiple blood transfusion.

4. CONCLUSION

Evans syndrome is a very rare autoimmune disorder in which the immune system destroys the body's RBC, WBC and/or platelets. The exact cause of this condition is unknown. The best treatment options for Evans syndrome depend on many factors, including the severity of the condition; the sign and symptoms present; and each person's response to certain therapies.

However, as with many other rare disorders, progress may depend upon the acquisition of detailed information through national/international databases and international multi centre randomized trials to accrue sufficient numbers of patients; long-term follow-up is also essential for the chronic relapsing nature of this condition.

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