International Journal of Advances in Medicine
Ramineni HB et al. Int J Adv Med. 2015 May;2(2):183-184
http://www.ijmedicine.com
pISSN 2349-3925 | eISSN 2349-3933
DOI: 10.5455/2349-3933.ijam20150524

Case Report

Evan's syndrome a strange cause of hemolysis: a case report

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Received: 31 March 2015
Accepted: 21 April 2015

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ABSTRACT

Evan's Syndrome is an autoimmune disorder characterized by autoimmune hemolytic anemia and Idiopathic Thrombocytopenia Purpura (ITP) or immune neutropenia in absence of any cause. We report this rare autoimmune disorder in a 37 year old female where she had presented with complaints of bleeding per vagina since 7 days with clots, non painful, bilateral joint pain since 2 days, swelling of foot & puffiness of face since 1 day, shortness of breath and pallor. Based on her past history and laboratory investigations confirmed the diagnosis of Evan' Syndrome. Therapy was initiated with corticosteroids and IV immunoglobulin.

Keywords: Idiopathic thrombocytopenia purpura, Thrombocytopenia, Autoimmune hemolytic anemia

INTRODUCTION

Evan's syndrome is an autoimmune disorder described by Robert Evans in 1951 specifying evidence exists between primary thrombocytopenic purpura and acquired hemolytic anemia. It was characterized by simultaneous destruction of the body's own red blood cells, white blood cells, platelets, neutrophils which causes Autoimmune Hemolytic Anemia (AIHA) and Idiopathic Thrombocytopenia Purpura (ITP) or immune neutropenia in absence of any cause.[1] It is a rare disorder found in only 0.8% to 3.7% of patient population with either ITP or AIHA at onset.[2] It occurs in all age group people. Despite of different therapeutic interventions, the great majority of patients have a chronic and relapsing course, which is associated with significant morbidity and mortality. Etiology of Evan's Syndrome was unknown. Many patients with Evan's syndrome have associated disorders such as systemic lupus erythematosus, chronic lymphadenopathy, or hypogammaglobulinemia, pallor, lethargy, jaundice, heart failure in severe cases, petechiae, bruising, mucocutaneous bleeding. Examination may reveal lymphadenopathy, hepatomegaly and/or splenomegaly.[3] Management of Evan's syndrome is challenging. Almost all patients require therapy during the course of the disease which should be effective, and nontoxic therapy should be continued.[4]

CASE REPORT

A 37 year old female has presented with complaints of bleeding per vagina since 7 days with clots, non painful, bilateral joint pain since 2days, swelling of foot and puffiness of face since 1 day, shortness of breath. On examination her vital signs were blood pressure: 140/80mmHg, pulse rate: 80beats per minute, respiratory rate: 16/min, temperature: afebrile. She was pallor and her systemic examination was found to be normal. On laboratory examination hemoglobin level-7.6gm%, PCV-23%, WBC-4,100 cells/mm^3, ESR-20mm/1st hr, RBC-2.6millions/ mm^3, Platelet count-0.54 lakhs / mm^3, MCV-88fl, MCH- 29pg, MCHC- 33%, DC: N-68%, L-25%, E-4%, M-3%, peripheral blood smear showed RBC: Mild anisopoikilocytosis comprising normocytes and macrocytes, Platelets: Thrombocytopenia was observed,

Prothrombin time-13.11sec, INR-1.0, Activated partial thromboplastin time (APTT)-29.46sec, LDH-837U/L, urine for proteins and creatinine was 0.012gm/dl and 0.0774gm/dl respectively, urine protein loss was found to be 155.8mg/day. Direct comb test and Indirect coombs test was found to be negative. Reticulocytes-0.9%, antinuclear antibodies-16.22. Based on these investigations she was diagnosed with Evans Syndrome. Treatment was initiated with Tab. Prednisolone 50mg, Tab. Shelcal BD, Tab. Tranexamic Acid 500mg, BD, and Cap. Rabeprazole sodium (EC) 20 mg with Domperidone (SR) 30 mg.

DISCUSSION

Evan's syndrome is an autoimmune disorder characterized by autoimmune hemolytic anemia and idiopathic thrombocytopenia purpura (ITP) or immune neutropenia in absence of any cause.[1] The prevalence rate of Evan's syndrome was 0.8-3.7% because it was a rare diagnosis. Etiology of evan's syndrome was idiopathic. In Evan's syndrome autoantibodies are directed against specific antigens which are specific to RBCS, platelets and neutrophils, but these autoantibodies do not cross-react. Most of the patients are presented with decreased igG, igM, and igA levels. It was presented with thrombocytopenia, anemia neutropenia and pancytopenia. Thrombocytopenia may include petechiae, purpura, and ecchymosis and anemia includes fatigue, pallor, shortness of breath and light-headedness along with jaundice. It is associated with some other autoimmune disorders such as systemic lupus erythematosus, chronic lymphadenopathy[5] among all these clinical presentations she was with thrombocytopenia, anemia, pallor, fatigue, shortness of breath. In this case she had acute flare up of SLE also[5] treating of Evan's syndrome is a challenging task. First line agents used for the treatment are corticosteroids like prednisone at a dose of 1-2 mg/kg divided 2 or 3 times daily, iv immunoglobulin for acute episodes. Other immunological agents such as cyclosporine, azathioprine, cyclophosphamide, vincristine, rituximab and alemtuzumab can be used in Evan's syndrome. Rituximab-a monoclonal antibody is considered as second line agent for Evan's syndrome[6] stem cell therapy can also be considered based up on severity. The clinical course of Evan's syndrome may include periods of exacerbation and remission. Patients rarely do well with treatment and often disappointing. Evan's syndrome can be fatal occasionally.[7]

CONCLUSIONS

Evan's syndrome is a rare autoimmune disorder which is a chronic and recurrent condition. It is often refractory to corticosteroids, IV immunoglobulin and splenectomy. Responding to some other agents may be anecdotal and inconclusive. Prognosis may include periods of exacerbation and remission. Patients rarely do well with treatment and often disappointing.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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DOI: 10.5455/2349-3933.ijam20150524
Cite this article as: Ramineni HB, M Chandini, M Sri Vidya, K Narendra Babu, S Vidyadhara. Evan's syndrome a strange cause of hemolysis: a case report. Int J Adv Med 2015;2:183-4.