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Case report *Medical research*

Primary Evans syndrome - case report

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ABSTRACT

Evans syndrome is a rare hematological disorder in which autoimmune hemolytic anemia and immune thrombocytopenia occur in the same patient. Patients usually have relapses and remissions. We report a female aged 32 years who presented with menorrhagia and purpuric rashes over her extremities. Investigations revealed thrombocytopenia and Coombs positive Autoimmune Hemolytic Anemia (AIHA) which was suggestive of Evans syndrome.

Keywords: Evans, Autoimmune hemolytic anemia, Thrombocytopenia

INTRODUCTION

Evans syndrome is an uncommon autoimmune disorder characterized by simultaneous or sequential development of Autoimmune Hemolytic Anemia (AIHA) and Idiopathic Thrombocytopenia Purpura (ITP) with or without immune neutropenia. [1] Evan's syndrome is diagnosed in only 0.8% to 3.7% of all patients with either ITP or AIHA at onset. [1] There is no preferential distribution of Evans syndrome by age, gender, or ethnic group. It has a chronic course and is characterized by recurrent relapses and remissions.

CASE REPORT

A 32 year old female was admitted with complaints of mennorhagia for 15 days. She had history of easy fatigability, bleeding gums on and off for the past two months. She also had menstrual disturbances over the past 3 years having 15/30 days cycles. She was also treated for subdural hematoma 2 years back. On examination, she was anemic and she had few petechial rashes over the left forearm. Systemic examination revealed splenomegaly.

Her labaratory values showed hemoglobin was 7.1 g/dL, platelet count of 67,000 /µL. Her liver function tests revealed indirect hyperbilirubemia. Her renal parameters, prothrombin time, APTT and INR were normal. Her peripheral smear showed microcytic hypochromic anemia with thrombocytopenia. Direct Coombs test was positive and serum LDH was elevated (520 IU/L, 110 – 220 IU/L). She underwent bone marrow aspiration which showed a cellular marrow with mild to moderate hyperplasia, increased reticuloendothelial cells, giant megakaryocytes and reduced marrow iron stores. Features were suggestive of autoimmune hemolytic anemia with immune thrombocytopenia and a diagnosis of Evans syndrome was made. She was investigated to rule out secondary causes of Evans syndrome. ANA, APLA, HBsAg, Anti HCV was found to be negative, suggesting our patient to be a case of primary Evans syndrome. She was started on intravenous Dexamethasone 8mg BD for first three days following which she was switched over to oral Prednisolone 30mg and Danazole 200mg daily. Subsequently she is currently on follow up and remains asymptomatic with normal platelets and menstrual cycles.

DISCUSSION

Evans syndrome was first described in 1951 by Evans. [2] It has been considered as a coincidental combination of ITP and AIHA and or immune neutropenia. There is increasing evidence to suggest that Evans reflects the state of profound immune dysregulation as opposed to coincidental combination of immune cytopenias. Evans Syndrome can be classified as primary (idiopathic) or secondary. In adults, an underlying cause can be expected in about 70% cases. [2]

Evans syndrome is a chronic and recurrent disease. Acute presentation and rapid deterioration as seen in our patient is not very common. Patients can present with fatigue, petechial rashes, bleeding gums, menorrhagia, subdural hematoma and other bleeding manifestations. Bone marrow examination is an essential investigation for the diagnosis of Evans syndrome.

Marrow examination reveals increased reticulocytosis and megakaryocytes. It is necessary to exclude infiltrative process in patients with pancytopenia. Significance of Coomb's test in patients with thrombocytopenia and anemia needs to be reemphasized. Other causes of acquired immune cytopenias like SLE, IgA deficiency, CVID, HIV, APLAS, TTP – HUS, Kasabach – Merrit syndrome, Castelman's disease and inherited ADAMTS-13 deficiency [1-3] should be excluded.

The management of Evan's Syndrome is challenging. Blood and platelets transfusion is the treatment given to alleviate symptoms and gain time but its use should be minimized. First – line therapy is represented by corticosteroids and/or intravenous immunoglobulin. The dose of prednisolone is 1 – 4 mg/kg. [3] Good clinical response may also be obtained with intravenous methylprednisolone. Norton et al proposed intravenous immunoglobulin (2g/kg in divided doses) for patients for whom steroids are ineffective or who require unacceptably high doses to remain in remission. [4] Successful use of rituximab (monoclonal anti – CD 20 antibody) has also been shown for Evans syndrome resistant to first-line therapy and other second-line treatments. [5]

Immunosuppressive agents such as ciclosporin, mycophenolate mofetil, vincristine, danazol may be considered after failure of first – line therapy. Third – line therapy maybe oral administration of cyclophosphamide [6] or intravenous administration of Alemtuzumab (monoclonal anti-CD52 antibodies). [7] Autologous and allogenic transplant stem cells may be performed only in a limited number of patients affected by Evans syndrome, after failure of pharmacological treatment. [8-9]

CONCLUSION

It is necessary to exclude all secondary causes to name a patient as primary Evans syndrome. It is vital to start steroids early in this autoimmune disease to prevent complications and mortality. In our case the response to treatment was good and the cytopenia reversed to normal. We have reported this case to highlight the need for awareness of this rare entity. This requires a high index of suspicion.

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