EUROPEAN JOURNAL OF BIOMEDICAL AND PHARMACEUTICAL SCIENCES
http://www.ejbps.com

ISSN 2349-8870
Volume: 7
Issue: 9
344-347
Year: 2020

CASE REPORT ON EVANS SYNDROME ASSOCIATED WITH SJOGRENS SYNDROME

Judy James[1]*, Gopika KS[2], Tonya Clara Thomas[2], Dhivya Jose[2], Smitha Mariyam Thomas[2] and Fathimath Dilsha[2]

[1]Sri Ramakrishna Institute of Paramedical Sciences, College of Pharmacy, Siddhapudhur, Coimbatore, 641044.
[2]Pharm D Intern, National College of Pharmacy, Calicut, India.

***Corresponding Author: Judy James**
Sri Ramakrishna Institute of Paramedical Sciences, College of Pharmacy, Siddhapudhur, Coimbatore, 641044.

Article Received on 13/07/2020 **Article Revised on 03/08/2020** **Article Accepted on 23/08/2020**

ABSTRACT
Evans syndrome is a rare and chronic autoimmunedisorder which is characterized by autoimmune hemolytic anemia and immune thrombocytopenic purpura with a positive direct anti-human globulin test. It predominates in children, mainly due to primary immunodeficiencies or autoimmune lymphoproliferative syndrome. It is mainly classified as primary and secondary, with the frequency in patients with autoimmune hemolytic anemia being 37-73%. People affected with evans syndrome often experience thrombocytopenia and Coomb's positive hemolytic anemia. The pathophysiology of this disease include deficiencies of CTLA-4, LRBA, and a decreased CD4/CD8 ratio. It is usually treated with corticosteroids and intraveneous IgG. In refractory cases, cyclosporine, rituximab, azathioprine, cyclophosphamide and mycofenolate mofetil have been used. Here we report a case report of Secondary Evans syndrome associated Sjogrens syndrome presented with fever, yellowish discolouration of eyes and shortness of breath. Hence the patient is treated with steroids, calcium supplements, antibiotics. Blood and platelets transfusion is the treatment given to improve symptoms and gain time but its use should be minimized.

KEYWORDS: Evans syndrome, Sjogren syndrome, Thrombocytopenic purpura, Coomb's test, IVIG(Intraveneous immunoglobulin).

INTRODUCTION
Evans syndrome is a rare autoimmune hematological disease which is first explained by Evans in 1951 when he studied the relationship between the autoimmune hemolytic anemia and immune thrombocytopenic purpura.[1] It is a rare, chronic, refractory disease but sometimes it may presentely actively. The typical course of evans syndrome is characterized by an heterogeneous chronic disease with clinical variability at onset, spontaneous remissions and exacerbations.[1,2,3] The first diagnosis of ES includes the presence of anemia, reticulocytosis, increased blood bilirubin and fecal urobilinogen, no family history of hemolytic diseases, evidence of antibodies against erythrocytes at 37˚C , the presence of purpura, prolonged bleeding time.[1] The pathophysiology of ES have different disease pathways which is not known yet, but it is summarised by the presence of immune dysregulation with antibodies against erythrocytes, platelets and/or granulocytes and decreased CD4:CD8 ratio.[4]

The classification of ES includes primary and secondary. The primary classification being an exclusion diagnosis with no underlying conditions, and secondary in the presence of baseline disease. Evans syndrome may occur in combination with another disorder as a secondary condition.[5] Secondary Evans syndrome can be associated with other disorders including lymphoproliferative syndrome, Sjogren's syndrome, antiphospholid syndrome, IgA deficiency, certain lymphomas.

It is usually treated with corticosteroids, IV immunoglobulin. In refractory cases, steroid sparing agents such as rituximab, cyclosporine, azathioprine, cyclophosphamide, thrombopoietin-receptor agonists are used.[6]

CASE REPORT
A 35 year old female patient was admitted in the medicine department with complaints of fever, yellowish discoloration of eyes and shortness of breath for past five days. Fever was associated with chills and rigors with generalized body pain, but not associated with any bleeding manifestations. Discoloration of eyes was not associated with pruritis. Shortness of breath was increased on exertion, insidious onset and relieved with rest. There were no evident history of chest pain, abdominal pain, palpitation, vomiting and cough. The patient received one unit blood transfusion from outside hospital which was uneventful.

On general examination, patient was conscious and alert. The patient's vitals were seem to be normal except temperature (101 F). Her Hemoglobin (Hb) level was 3.6 g/dl, WBC 10,000 mg/dl, Platelet 60,000 cells/cumm. Peripheral smear showed severe degree of microcytic hypochromic anaemia with thrombocytopenia. Direct Coomb's test was positive. Total bilirubin was 14.7 mg/dl , SGPT 102 IU/L, SGOT 90 IU/L. Serum Lactate Dehydrogenase was 1048 IU/L and ESR was found to be 60mm/hr. Chest X-ray, USG Abdomen and Echo investigations were normal. Anti nuclear Antibody test was undergone in this patient and found to be143 IU/L . Bone marrow examination showed increased myelopoiesis with micro and megaloblastic erythropoiesis. The patient was given 2 units packed cell transfusion. Injection Artesunate 2.4 mg IV, injection ceftriaxone 1g iv, clindamycin 600 mg iv bd and tablet paracetamol 500 mg tid.

On second visit,the patient's Hb level was 6.4 g/dl, WBC 9,400 cells/cumm, platelet count 45,000 cells/cumm. The patient was prescribed Prednisolone 1mg/kg/day , tab Pantoprazole and tab calcium 500mg OD. One unit packed cell transfusion was given.

After 10 days, the patient's Hb level was increased to 9.7g/dl, platelet count was raised to 90,000 cells/cumm and total bilirubin was 3.6mg/dl. Then after 4 weeks the same drugs were prescribed and the haemoglobin level was found to be normal ie., 11.6g/dl. Platelet count 1,65,000cells/cumm. Then total (1.1mg/dl) and indirect bilirubin (0.7mg/dl) was found to be normal.

DISCUSSION

Evans syndrome is a hematologic disorder characterized by the sequential or simultaneous development of DAT positive autoimmune hemolytic anaemia (AIHA), immune thrombocytopenia (ITP) and /or immune neutropenia in the absence of a known etiology. This rare disease is diagnosed in only 0.8% to 3.7% of all patients with either ITP or AIHA at onset.[7] More recent data suggest the spectrum of the disease has broadened specially in children and there is increasing evidence of Evans syndrome reflecting the state of profound immune dysregulation as opposed to coincidental combination of immune cytopenias.[8] This syndrome is classified as primary (idiopathic) or secondary (associated with some other disease) and it has been demonstrated that secondary disease responds better than primary variable.[9] The diagnosis of ES still implies the exclusion of other disease conditions and especially thrombocytopenic purpura which requires prompt management. In a study of Michael et al. sixty eight patients of Evans syndrome mortality was seen in 23.4% cases. The possible causes being septic shock, associated cancers, stroke, refractory anemia with lymphomas. However, in clinical practices, true cases of ES may well show or precede avariety of underlying diseases or conditions which may influence both the management and outcome. Significance of Coomb's test (direct or indirect) in patients with thrombocytopenia and anaemia needs to be reemphasized.

The management of Evan's syndrome is difficult and challenging. Blood transfusions and platelets transfusion is the treatment given to improve symptoms and gain time but its use should be minimized. The first line of treatment is Prednisolone and intravenous immunoglobulins (IVIG). Second line of treatment consists of immunosuppressants, Monoclonal antibody and chemotherapy. More recently small number of patients have been treated by stemcell transplantation.[7,8,9]

CONCLUSION

Evan's syndrome is a rare disease with a heterogeneous course characterized by multiple relapses during lifetime, despite multimodal treatment. A combination of genetic and epigenetic factors appears to be involved in its pathogenesis, and the knowledge of its precise etiology will help to design specific targeted therapies with less-severe adverse effects, significantly improving patients quality of life. In patient's presenting with immune thrombocytopenia and anemia with hemolytic factor, DAT is mandatory test. Instead of monotherapy with corticosteroids, combination of steroids with newer modalities is like immunosupressants and rituximab should be instituted as early as possible inorder to prevent or delay life threatening complications.

ACKNOWLEDGEMENT

The authors thank the Department of General Medicine, KMCT Medical college hospital, for their valuable support throughout the study.

Conflict of interest

The authors have declared no conflicts of interest.

REFERENCES

1. Evans R, Takahashi K, Duane RT, Payne R, Liu C. Primary thrombocytopenic purpura and acquired haemolytic anemia: evidence for a common etiology. *AMA Arch Intern Med*, 1951; 87(1): 48-65.
2. Dhingra KK, Jain D, Mandal S, Khurana N, Singh T, Gupta N. Evans syndrome: a study of six cases with review of literature. *Hematology*, 2008; 13(6): 356-360.
3. Pui CH, Wilimas J, Wang W. Evans syndrome in childood. *J Pediatr*, 1980; 97(5): 754-758.
4. Wang W, Herrod H,Pui CH, Presbury G,Wilimas J. Immunoregulatory abnormalities in Evans syndrome. *Am J Hematol*, 1983; 15(4): 381-390.
5. Costallat GL, Appenzeller S, Costallat LT. Evans syndrome and systemic lupus erythematosus: clinical presentation and outcome. *Joint Bone Spine*, 2012; 79(4): 362-364.
6. Mathew M. Evans syndrome: practice essentials, background, pathophysiology, https://emedicine.medscape.com/article/955266-overview#showall, 2017.

7. Benesch M, Urban C, Platzbecker U, Passweg J. Stem cell transplantation for patients with Evans syndrome. Expert Review of Clinical Immunology, 2009; 5(3): 341-348.
8. Savasan S, Warrier I, Ravindranath Y. The spectrum of Evan's syndrome. Archives of Disease in Childhood, 1997; 77(3): 245-248.
9. Michel M, Chanet V, Dechartres A, Morin AS, Piette JC, Cirasino L, et al. The spectrum of Evans syndrome in adults: new insight into the disease based on the analysis of 68 cases. Blood, 2009; 114(15): 3167-3172.