

Evans Syndrome – A Case Report

Dr. Mithil Ghushe¹, Dr. Shweta Bijwe²

¹(Department Of Medicine , MUHS,Nasik, India)

²(Department Of Pathology, MUHS,Nasik,India)

Corresponding auther: Dr. Mithil Ghushe

Abstract-Evans Syndrome Is A Rare Hematological Disease Commonly Defined As Coombs-Positive Hemolytic Anemia And Immune Thrombocytopenia. It Is Associated With Decreased Cluster Of Differentiation (CD)4+ T-Helper Cell Counts, Increased CD8+ T-Suppressor Cell Counts, A Decreased CD4/CD8 Ratio, And Reduced Serum Immunoglobulin G, M And A Levels - Indicating A Complex Immune Dysregulation. Association With Other Autoimmune Diseases Has Been Described With Evans Syndrome. We Describe A Case Of A 23-Years Old Female Affected By Autoimmune Hemolytic Anemia And Thrombocytopenia Successfully Treated With Steroids.

Keywords - Evans Syndrome, Autoimmune Hemolytic Anemia, Autoimmune Thrombocytopenia

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I. INTRODUCTION

Evans Syndrome Is An Uncommon Condition Defined By The Contemporary Or Sequentially Association Of Immune Thrombocytopenia (ITP) And Autoimmune Haemolytic Anaemia (AIHA), With A Positive Direct Coombs Test.) Sometimes Associated With Neutropenia In The Absence Of Known Underlying Etiology. It Has A Chronic Course And There Is No Preferential Distribution By Age, Gender, Or Ethnic Group. It Is Characterized By Recurrent Relapses And Remissions. First-Line Therapy Includes Corticosteroids And Intravenous Immunoglobulin With Good Clinical Response, Although Relapse Is Frequent. Immunosuppressive Drugs And Splenectomy May Be Considered When First Line Treatment Has Failed.[2]

It Is Associated With Decreased Cluster Of Differentiation (CD)4+ T-Helper Cell Counts, Increased CD8+ T-Suppressor Cell Counts, A Decreased CD4/CD8 Ratio, And Reduced Serum Immunoglobulin G, M And A Levels - Indicating A Complex Immune Dysregulation [3].

Association With Different Autoimmune Diseases, Such As Systemic Lupus Erythematosus, Autoimmune Hepatitis, Hashimoto's Thyroiditis, Dermatomyositis And Chronic Inflammatory Demyelinating Polyneuropathy, Has Also Been Described.[4]

II. CASE REPORT

A 23-Years Old Female Was Admitted To ICU With Hematuria, Petechiae And Bruising On The Skin Since 6 Days Alon With History Of Low Grade Fever And Chronic Cough Since 4 Months . On Clinical Examination Pallor And Mild Icterus Was Present. There Was No Organomegaly Or Lymphadenopathy.

A Complete Blood Count Revealed Anemia (6 Gm/Dl) And Severe Thrombocytopenia (Platelet Counts 15,000/Mmc). Raised Total Bilirubin With Increased Indirect Bilirubin, Raised Reticulocyte Count, Decreased Haptoglobulins , Positive Direct Coombs Test And Positive Anti-Neutrophil Antibodies Suggested Diagnosis Of Autoimmune Hemolytic Anemia.

Relevant Investigations Done For Other Autoimmune Diseases Came Negative.

CT Chest Showed Bilateral Upper Lobe, Right Middle Lobe Associated Areas Of Confluent And Discrete Nodular Densities And Gene Expert Study Was Positive , Thus Patient Was Started On ATT.

Bone Marrow Examination Was Done For Suspected ITP, It Showed Dysmorphic Megakaryocytes With Normal Representation Of Erythroid Cells. It Also Excluded Possibility Of Any Malignancy. On The Basis Of Clinical Manifestations And Laboratory Findings That Confirmed Autoimmune Hemolytic Anemia And Thrombocytopenia, The Diagnosis Of Evans Syndrome Was Made. The Patient Was Treated Successfully With Steroids. Blood Transfusion And Platelet Transfusion Was Given. A Complete Blood Count Obtained 1 Weeks Later Showed An Increasing Hemoglobin Levels (8.9g/Dl) And Normalization And Platelet Count (PLT 257.000/ Mmc).

III. DISCUSSION

Evans Syndrome Was First Described In 1951 By Robert Evans. It Is Rare Disease, A Diagnosis Of Exclusion, And Requires A High Index Of Suspicion. The Etiology Is Unknown And Immune Dysregulation May Be Involved In The Pathogenesis. Constitutive Production Of IL-10 And INF May Lead To Activation Of Autoreactive, Antibody-Producing B Cells [5], Although These Abnormalities Of Immune Response Are Seen In Other Autoimmune Disorders Also. The Appearance Of The Second Cytopenia May Occur Months To Years After The First Immune Cytopenia And May Delay Diagnosis [6]. Neutropenia Occurs In Up To 55% Of Patients At Presentation[6]. Clinically Symptoms Of Hemolysis (Fever, Pallor, Jaundice, Lethargy) And Thrombocytopenia (Petechiae, Bruising And Mucocutaneous Bleeding) May Be Seen. Physical Examination May Reveal Lymphadenopathy, Hepatomegaly And/Or Splenomegaly. These Signs May Be Chronic Or Intermittent And In Same Case May Occur During Acute Exacerbations. The Diagnosis Of Haemolytic Anaemia Requires Direct Coombs Positivity, Although This Investigation May Be Positive Even In The Absence Of Haemolytic Anaemia. The Indirect Coombs Test May Also Be Positive In A Small Percentage Of Patients. Antiplatelet And Antigranulocyte Antibodies Research Is Controversial Because A Negative Result Does Not Exclude The Diagnosis. A Complete Medical History, An Appropriate Physical Examination, A Complete Blood Count And A Peripheral Smear Are Diagnostic. However, Other Causes Of Acquired Immune Cytopenia Should Be Excluded (SLE, Iga Deficiency, CVID, HIV, ALPS, TTP, HUS, Kasabachmerrit Syndrome, Castelman's Disease And Inherited ADAMTS-13 Deficiency).[2]

Bone Marrow Examination Is An Essential Investigation For The Diagnosis Of Evans Syndrome. It Is Necessary To Exclude Infiltrative Process In Patients With Pancytopenia, Especially Before Starting Steroid Therapy. Clinical Presentation, Hemoglobin Levels And Platelet Count Guide The Therapeutic Decisions. First-Line Therapy Is Represented By Corticosteroids And/Or Intravenous Immunoglobulin. Blood And/Or Platelet Transfusions May Also Be Need To Ameliorate Symptoms In Severe Cases, Even Though Their Use Is Not Routinely Recommended. Immunosuppressive Agents May Be Considered After Failure Of First-Line Therapy. Each Treatment Of Second Line Therapy Is Burdened By Short And Long Time Side Effects. For These Reasons, Therapeutic Options Should Be Individualized, Basing On Patient's Age, Natural History And Severity Of The Disease. Third-Line Therapy Provides Oral Administration Of Cyclophosphamide[7] Or Intravenous Administration Of Monoclonal Anti-CD52 Antibodies[8]. 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 [9].

IV. CONCLUSION

Careful Surveillance And High Index Of Suspicion Is Required To Diagnose This Rare Diseases, Which Can Lead To Prompt Treatment , Minimizing The Complications And Improve Patient Outcome.

REFERENCES

- [1] Evans RS, Takahashi K, Duane RT, Payne R, Liu C: Primary Thrombocytopenic Purpura And Acquired Hemolytic Anemia; Evidence For A Common Etiology. *AMA Arch Intern Med* 1951, 87:48–65.
- [2] F. Porcaro Et Al.: Evans Syndrome: A Case Report. *Ped. Med. Chir. (Med. Surg. Ped.)*, 2014, 36: 167-169
- [3] Wang W, Herrod H, Pui CH, Presbury G, Wilimas J: Immunoregulatory Abnormalities In Evans Syndrome. *Am J Hematol* 1983, 15:381–390.
- [4] Simon Et Al.: Evans Syndrome Associated With Sterile Inflammation Of The Central Nervous System: A Case Report. *Journal Of Medical Case Reports* 2013 7:262.
- [5] Karakantza M, Moukaki A, Theodoropoulou M, Bussel JB, Maniatis A. Th 1 And Th2 Cytokines In A Patient With Evans' Syndrome And Profound Lymphopenia. *British Journal Of Haematology* 2000; 110, 968–970.
- [6] Mathew P, Chen G, Wang W. Evans Syndrome: Results Of A National Survey. *Journal Of Pediatric Hematology/Oncology* 1997; 19, 433–437.
- [7] Gombakis N, Trahana M, Athanassiou M, Kanakoudi-Tsakalidou F. Evans Syndrome: Successful Management With Multiagent Treatment Including Intermediate-Dose Intravenous Cyclophosphamide. *Journal Of Pediatric Hematology/Oncology* 1999; 21, 248–249.
- [8] Willis F, Marsh JCW, Bevan DH, Killick SB, Lucas G, Griffiths R Et Al. The Effect Of Treatment With Campath-1H In Patients With Autoimmune Cytopenias. *British Journal Of Haematology* 2001; 114, 891–898.
- [9] Passweg J, Rabusin M, Musso M, Beguin Y, Cesaro S, Ehninger G Et Al. For The Autoimmune Working Party Of The EBMT. Haematopoietic Stem Cell Transplantation For Refractory Autoimmune Cytopenias. *British Journal Of Haematology* 2004; 125, 749–755.

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