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Case Report

Evans Syndrome as Rare Presentation in Systemic Lupus Erythematosus

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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. It is one of the rare presenting features of autoimmune disorders, especially systemic lupus erythematosus (SLE), and sometimes may even precede the onset of disease. Primary Evans syndrome with no cause is very rare and is seen in children. Here, we describe a case of secondary Evans syndrome with severe autoimmune hemolytic anemia leading to acute kidney injury. This is one of the rare presentations of SLE and there are only few case reports.

Key word: Evans syndrome, systemic lupus erythematosus, autoimmune haemolytic anaemia.

INTRODUCTION

Evans syndrome (ES), which was first described in 1951, is an autoimmune disorder characterized by the simultaneous or sequential development of autoimmune hemolytic anemia (AIHA) and immune (ITP) and/or immune neutropenia in the absence of any underlying cause. [1,2] ES has been since its first description considered or defined as an "idiopathic" condition and thus mainly as a diagnosis of exclusion, ES may be associated with or show other diseases or conditions such as systemic lupus erythematosus (SLE), [3] lymphoproliferative disorders, [4,5] or primary immuno deficiencies. [6] In childhood, ES may also show an autoimmune lymphoproliferative syndrome (ALPS), a disorder of disrupted lymphocyte homeostasis related to some mutations in the Fas apoptotic pathway. [7] ES is a rare condition because it is diagnosed in only 0.8% to 3.7% of all patients with either ITP or AIHA at onset. [8] Few and mainly pediatric data on ES are available in the literature; [9-11] therefore, the characteristics and outcome of adult's ES are poorly known.

CASE REPORT

A 28 aged female came to emergency department with the complain of breathlessness for past 5 days. On examination, patient was pale and tachypneic, systemic examination was normal. Patient was further investigated. [Table-1]

ANA by immunofluorescence method is positive for Nucleosome. Then based on the positive coomb's test and presence of spherocytes on the smear, both autoimmune hemolytic anemia and immune thrombocytopenic purpura was identified and diagnosis of Evans syndrome was made. The absence of schistocytes on the peripheral smear, normal FDP, excludes the diagnosis of thrombotic thrombocytopenic purpura, hemolytic uremic syndrome and DIC.

The patient was transfused with 3 units of least incompatible RCC and 6 units PC. Hb improved to 5g/dL. The patient was treated with IV Methyl prednisolone for 5 days and followed by oral prednisolone 1mg/kg. Both Hb and platelets started increasing and patient condition improved.

Table no.1 Investigations

Haemoglobin	3.2gm%	Peripheral Smear
WBC count	14,000/cmm	Anisocytosis 3+
Platelet count	50,000/cmm	Poikilocytosis 2+
Serum creatinine	2.6 mg/dl	Microcytes +
LDH	1690	Hypochromic +
Total bilirubin	3.2 mg/dl	RBC-Microspherocytes seen;many RBC seen in aggregates
Direct bilirubin	1 mg/dl	WBC-Polymorphic leucocytosis and toxic granules seen
Indirect bilirubin	2.2 mg/dl	Platelets-reduced
Uric acid	10.9	Picture suggesting of Hemolytic anemia
Reticulocyte count	3.2	Direct Coomb's-Positive
Urine Albumin	++	Indirect Coomb's-Positive
USG ABDOMEN	No Organomegaly	

DISCUSSION

This presentation is most consistent with Evans syndrome, which is defined by a combination of Coombs positive autoimmune hemolytic anemia, immune thrombocytopenic purpura, and, less commonly, autoimmune neutropenia. The pathophysiology of Evans syndrome is not clearly understood, but likely involves autoantibodies directed against a base protein of Rh blood group, thus destroying red blood cells, and a separate group of autoantibodies directed against platelet GPIIb/IIIa, thus destroying platelets. [12] Interestingly, nearly 50% of cases of Evans syndrome are associated with autoimmune conditions such as systemic lupus erythematous, lymphoproliferative disorders, and common variable immunodeficiency. [13] First line treatment is steroids or steroids in combination with IVIG. [14] Second-line treatment options include rituximab (which induces remission in the majority of cases, but responses are often sustained for <12months) and other immunosuppressive agents such as cyclophosphamide, mycophenolate, cyclosporine, vincristine, danazol and azathioprine. [15] Third line treatment includes splenectomy. For severe and refractory cases, hematopoietic cell transplantation is the only chance for cure, with limited data showing that allogeneic hematopoietic stem cell transplantation may be superior to autologous hematopoietic stem cell transplantation. [16] [Figure-1]

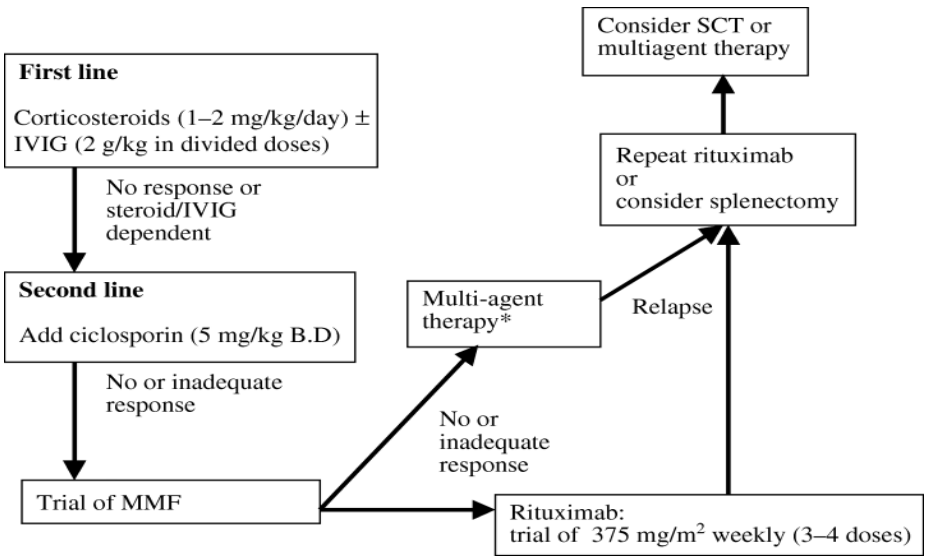

Fig 1. Management of Evans syndrome: a sequential approach. *Multiagent therapy: steroids/IVIG/vincristine/danazol/ciclosporin (Scaradavou & Bussel, 1995); vincristine/methylprednisolone/cyclosporine (Williams & Boxer, 2003).

We have reported this case to highlight the need for awareness of this rare entity. This requires a high index of suspicion among primary care physicians as well as other specialities like gynaecology. Evan's syndrome is a chronic and recurrent disease. Acute presentation and rapid deterioration is not very common. Significance of Coomb's test in patients with thrombocytopenia and anaemia needs to be reemphasized. Newer modalities of treatment Rituximab along with steroids should be instituted early for more favourable outcome.

CONCLUSION

Evan's Syndrome is a rare chronic, relapsing and refractory disease but sometimes may present acutely. In patients presenting as immune thrombocytopenia and anaemia with haemolytic component, Direct Anti-globlin is mandatory. Instead of monotherapy with corticosteroids, combination of steroids with newer modalities like Rituximab should be instituted early in order to prevent or delay life threatening complications.

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