Systemic lupus erythematosus with Autoimmune Hemolytic anemia in a 37-year old patient with Beta-thalassemia trait

Majed Momin[1], Amitha G[2], Anamika Aluri[3], Sandhya Rani C[4]

[1,4]Consultant Pathologist, [2]Consultant Microbiologist,[3]Consultant Biochemist and Head Biochemistry lab. Yashoda Hospital, Malakpet Nalgonda X-roads, Hyderabad – 500036.

Address for correspondence: Dr.Majed Momin, Consultant Pathologist, Department of Laboratory Medicine, Yashoda Hospital, Malakpet, Nalgonda X-roads, Hyderabad, Telangana, India.

Email id: majedmomin878@yahoo.co.in

ABSTRACT

The occurrence of Systemic lupus erythematosus (SLE) with Autoimmune hemolytic anemia (AIHA) has been only rarely reported in patients with Beta thalassemia trait.We here report a case of SLE presented with history of fever, giddiness associated with headache for three months and on physical examination revealed rash all over the body. On routine hemogram parameters indicated anemia with low blood indices, leucopenia, reticulocytosis, positive coombs test. In view of prolonged fever with rash, LE cell preparation, anemia with erythrocytosis and low blood indices Hemoglobin Electrophoresis was advised which showed positive LE cells in peripheral blood and hemoglobin electrophoresis suggestive of Beta thalassemia trait. Physicians should be alerted to the possible association of hemolytic anemia underlying SLE & Beta thalassemia trait so as not to delay correct diagnosis and initiation of appropriate treatment. We present this case as the occurrence of connective tissue diseases, in particular SLE, has only been rarely reported in patients with thalassemia minor. Peripheral smear findings with hemolytic picture gives clue for underlying SLE and Beta thalassemia minor.

Keywords: Auto immune hemolytic anemia, Systemic lupus erythematosus, Beta thalassemia minor.

INTRODUCTION

Anemia of varying degree and leucopenia is a quite common finding in SLE, but only rarely has an association with concomitant haemoglobinopathies, in particular, beta thalassemia minor. Anemia can take any form – the anemia of chronic disease which is normocytic and normochromic, iron deficiency anemia, or a Coombs' positive hemolytic anemia[1]. Additionally, co-existent hemoglobinopathies such as sickle cell anemia[2] and thalassemia trait[3,4] must be considered. The workup includes iron studies, haemoglobin electrophoresis depending on the patients' indices, and other markers of hemolysis, reticulocyte count, serum haptoglobin levels and lactate dehydrogenase (LDH). Hemolytic anemia occurring in 10 - 15%, is rarely severe enough to require transfusion.

SLE is a chronic inflammatory autoimmune disorder that more commonly affects women[3]. It is a multi - organ disease and may affect any organ system. Several decades ago, the diagnosis of lupus included the LE cell assay. Autoimmune serology techniques such as anti-nuclear antibody staining have replaced this technique. Beta thalassemias are a group of hereditary blood disorders characterized by anomalies in the synthesis of the beta chains of hemoglobin resulting in variable phenotypes ranging from severe anemia to clinically asymptomatic individuals.Carriers of thalassemia minor are usually clinically asymptomatic but sometimes have mild anemia[4].

CASE REPORT

A 37 year-old female referred to our hospital with history of low grade fever, giddiness associated with headache and retrosternal pain from last three months. Physical examination revealed an ill looking female with mild pallor, body temperature 98.4 degree F, pulse rate 90/min, blood pressure 130/70mm of Hg & respiratory rate was 20/min. Systemic examination was unremarkable. Routine investigations were done and blood picture was microcytic hypochromic with moderate anisopoikilocytosis showing elliptical and few target cells, mild polychromasia and spherocytes seen on peripheral blood smear. Malarial antigen,Typhi-Ig M, Weil Felix test and stool for ova and cyst were negative. Direct coomb's test came positive with differential positive for IgG(warm antibody). KFT and coagulation study came normal. Table 1 showing the list of Investigations done in the present case.

Table 1: Showing list of investigations

Name of the Test	Result
WBC count	3400cells/cumm
Platelet count	2.8 lakhs /cumm
Hemoglobin	8.1gm/dl
RBC	5 millions/cumm
MCV	58fl
MCH	18pg
MCHC	28gm/dl
ESR	60mm/hr
Serum Creatinine	1.0mg/dl
SGOT	26U/L
SGPT	32U/L
Alkaline Phosphatase	68U/L
Reticulocyte count	03%
Plasma Glucose level	124mg/dl
Total Iron	45mcg/dl
TIBC	168 mcg/dl
Transferin saturation	20.8%

In view of clinical history of fever with anemia, leucopenia and high ESR, LE cell preparation done which showed numerous LE cells [Figure 1. Round eosinophilic,homogenous mass(LE body) engulfed by neutrophils and monocytes,displacing the nucleus of neutrophil to the rim of the cell]

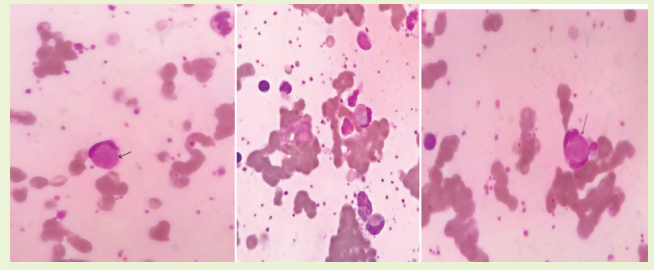

Figure 1: Smears showing rouleaux formation,agglutination of RBC along with LE cell. [Leishman,x40]

Advised battery of tests including ANA, Antibody profile and ANA IF study which came positive for ANA, ds DNA & nucleosomes. ANA IF showed positive with nucleus homogenous pattern and intensity 3+. Bone marrow aspiration done showed erythroid preponderance and normal ironstores. Hemoglobin Electrophoresis reveal increased Hemoglobin A2 suggestive of Beta thalasemia minor. Chest X-ray was normal. Ultrasound abdomen and CT brain showed normal study. Finally patient was treated as a case of autoimmune hemolytic anemia with SLE and Beta thalassemia minor.

DISCUSSION

In the present case report we described the case of 37-year-old woman with SLE and coexistent Beta thalasemia trait. SLE is a chronic inflammatory autoimmune disorder that more commonly affects women[5,6]. It is a multi-organ disease and may affect any organ system.

Anemia, leucopenia, lymphopenia, and thrombocytopenia are the most common hematological findings and in most cases, are derived from autoantibody-mediated peripheral destruction[7,8]. The presence of LE cells in blood in conjunction with the appropriate clinical picture and laboratory values, would contribute to the diagnosis of SLE[9].

Peripheral smear findings, hemolytic anemia with direct coomb's of IgG(warm antibody) and positive LE cell gives clue for underlying collagen vascular disease and Hemoglobinopathies. Further special investigations confirmed diagnosis. The overlap of SLE and Beta thalassemia is of interest, but the limited number of patients that have been reported previously implies that the association is uncommon.The association between SLE and haemoglobinopathies has been poorly explored to date. Sixteen cases have been reported describing the co existence of sickle cell disease[3,10] and SLE, but only Kaloterakis et al reported a case of sickle cell/ß° thalassemia in a patient with SLE.

Diagnosis of coexistent of SLE with Beta thalassemia is necessary to predict treatment outcome. There is increased evidence of SSA antibodies or persistently reduced C3 and C4 levels among those with Beta thalassaemia have some role in increasing the atherosclerotic events and reduced ability to bind the immune complexes (secondary to chronic hypercoagulable state or to the homocysteinemia)[11,12]. Thalassemia patient's may have increased transfusion requirement over time because of alloantibodies and secondary infection due to SLE. Thalassemia with SLE sometimes may develop lupus anticoagulant that can cause intracranial hemorrhage[13].

In conclusion, the prevalence of ß thalassaemia in patients with SLE seems to be lower than in the general population. Basic investigations give clue for underlying diseases. However, when the two conditions coexist, SLE seems to have a more severe course. Early diagnosis and the initiation

of appropriate treatment may decrease morbidity and mortality in these patients.

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