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A CASE OF A CHRONIC LYMPHOPROLIFERATIVE DISORDER PRESENTING AS AUTOIMMUNE HEMOLYTIC ANEMIA

Immunohematology

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ABSTRACT

Lymphoproliferative disorders encompass a group of diseases with a highly variable clinical course. This is a case report of a patient who presented with haemolytic anemia initially and was subsequently diagnosed as a chronic lymphoproliferative disorder. He was treated with Rituximab to which he showed a favourable response.

KEYWORDS

INTRODUCTION

Lymphoproliferative disorders are a set of disorders characterized by the abnormal proliferation of lymphocytes into a monoclonal lymphocytosis. The two major types of lymphocytes are B cells and T cell.

Individuals who have dysfunction of their immune system are susceptible to develop a lymphoproliferative disorder because when any of the numerous control points of the immune system become dysfunctional, immunodeficiency or deregulation of lymphocytes is more likely to occur.

The most common type is CLL (chronic lymphocytic leukemia) but other non CLL types such as hairy cell leukemia, B cell lymphoma, T cell lymphoma and splenic lymphoma etc. are also seen.

CASE REPORT

A 50-yr old male, non diabetic, normotensive, non-alcoholic, non-smoker presented with a history of jaundice for 15 days and history of easy fatigability for 10 days. There was no fever, cough, weight loss, abdominal pain, vomiting, clay coloured stools or bleeding from any site. The patient did not notice any swellings on the body. He did not have any prior drug intake or blood transfusion.

On examination, his vital parameters were stable. Pallor and icterus were present. There were no palpable lymph nodes. Liver and spleen were not palpable.

Lab investigations showed Hemoglobin of 3 gm%. Total counts were 17,000 with 71% lymphocytes and 29% neutrophils. Platelet count was 2.7 lakhs. MCV was 120.3 fL. Peripheral smear showed moderate anisocytosis, moderate to marked polychromasia, macrocytes and microspherocytes. Corrected Retic count was 4.3%. Liver function tests showed total bilirubin of 5.4 mg% with direct bilirubin 0.5 mg%. Enzymes and serum proteins were normal. Renal function tests were normal.

Direct Coomb's test was positive and Indirect Coomb's test was negative. G6PD levels were normal. Hemoglobin Electrophoresis showed normal study. Osmotic fragility test was negative. Serum iron was 132 ug/dl, serum ferritin was normal. ANA and dsDNA were negative. HbsAg and anti-HCV were negative.

ECG and chest radiograph were normal. Ultrasongraphy of abdomen showed liver 14cm, spleen 12.7cm and pancreas and kidneys normal.

A provisional diagnosis of autoimmune hemolytic anemia was made. The patient was given intravenous Methylprednisolone 1 gm for 3 days and then put on oral prednisolone 1mg/kg body weight. Patient was transfused two units of packed red cells. Due to repeated autoagglutination, further crossmatching was not possible.

The patient improved gradually over a period of 1 week. His hemoglobin increased to 7.9g% and bilirubin decreased to 3.5mg%. He was discharged as a case of autoimmune haemolytic anaemia. He was put on oral steroids at 50 mg per day (planned to be gradually tapered) and asked to follow up at our out-patient department after a week. At the follow up visit, his repeat complete hemogram showed Hemoglobin of 5.8 gm% . Total count was 32500 with neutrophil count of 43 % and lymphocyte count of 57 %. Platelets were normal. MCV was 120, MCH was 32 and MCHC was 32. Peripheral smear showed moderate anisocytosis, moderate to marked polychromasia, microcytes, and smudge cells.

The patient was readmitted for further evaluation. CT scan of abdomen and thorax was done which showed mild hepatomegaly, intra-abdominal lymphadenopathy along the para-aortic and aorto-caval regions and external iliac vessels, largest of 14 x 6 mm. Bone marrow aspirate revealed erythroid hyperplasia with micronormoblastic erythropoiesis, absence of fat content in fragments, increase in the number of mature lymphocytes (68%) and numerous smudge cells which was opined as a picture being consistent with hemolytic anemia and lymphocytosis of marrow, suggestive of a chronic lymphoproliferative disorder. Immunophenotyping showed positivity for CD19 and CD20 100% and CD22 95% and CD 23 98%. Other CD markers were negative. This was suggestive of a chronic B cell lymphoproliferative disorder.

The final diagnosis therefore was an autoimmune haemolytic anemia with chronic B-cell lymphoproliferative disorder of non CLL type.

Following this,the treatment received by the paient was Prednisolone 40 mg /day initially tapered to 10 mg/day over one month and Inj. Rituximab 500 mg once a week for 4 weeks (4 cycles).Inj. Pneumovac and Revac-B (Hepatitis B) were given. He was also put on supportive treatment.

The patient was reviewed one week after completing this treatment which showed improvement in the total count to 13,000 and Hb to 11.2 gm %.

DISCUSSION

Patients with chronic lyphoproliferative disorders especially B-chronic lymphocytic leukemia (CLL) have a 5–10% risk of developing autoimmune complications which primarily cause of cytopenia. These autoimmune cytopenias can occur at any stage of CLL and do not have independent prognostic significance. The most common autoimmune complication is autoimmune hemolytic anemia with a lower frequency of immune thrombocytopenia and pure red blood cell aplasia and only rare patients with autoimmune granulocytopenia.

Many studies have been done on autoimmune hemolytic anemia complicating CLL but few on it's presence in other lymphoproliferative disorders although the pathogenesis is similar.

In 2013, Zhuang YI et al[1], did a study on autoimmune hemolytic anemia associated with B-cell chronic lymphoproliferative disorders. Out of the 14 patients in the study, all had DCT positive and 8/14 patients were treated with steroids and Rituximab. The rest were given either steroids alone or steroids with chemotherapy. Overall response rate was 100% and of those treated with Rituximab only 1 had a relapse. They concluded that AIHA can occur at any stage of a chronic lymphoproliferative disorder and that Rituximab is an effective therapy. Various other studies have been done which showed efficacy

of Rituximab in autoimmune hemolytic anemia associated with CLL.[2,3,4]

Good response to Rituximab was seen in our patient also.

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