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Persistent Autoimmune Hemolytic Anemia Complicating Splenic Marginal Zone Lymphoma: A Case Report

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INTRODUCTION

Splenic Marginal Zone Lymphoma (SMZL) is a unique subtype of MZL in its involvement of the spleen, bone marrow, and blood and typically requires treatment only if symptomatic. Autoimmune hemolytic anemia (AIHA) is an acquired hemolytic condition that can manifest as either primary or secondary to underlying associated disorders.

BACKGROUND

Marginal zone lymphomas (MZL) are a heterogeneous subtype of indolent B-cell non-Hodgkin lymphomas that encompass a range of distinct entities. They are accounting for 7% of non-Hodgkin lymphoma (NHL)[1]. Splenic MZL, a type of MZL, is characterized by a generally indolent behavior, and only patients with symptomatic presentations should be considered for treatment[2]. AIHA is an acquired hemolytic condition caused by the host's immune system attacking its red cell antigens[3]. The incidence of AIHA is approximately 1 per 100,000 annually[4], with an increased likelihood of occurrence with advancing age. AIHA cases can be classified into warm type (65%), cold type (29%, comprising cold hemagglutinin disease [CHAD] and paroxysmal cold hemoglobinuria [PCH]), or mixed AIHA (5%). Approximately 50% of cases are primary (idiopathic) AIHA, while the other half are secondary to underlying associated disorders[5].

AIM

We report a patient with persistent AIHA following successful treatment of splenic MZL.

CASE REPORT

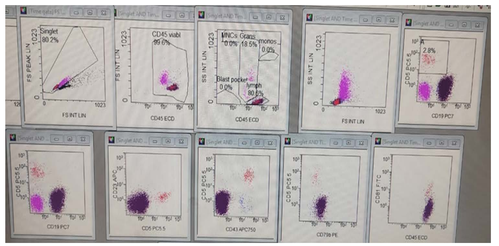
A 70-year-old male patient presented with a one-month history of lethargy, fatigue, and dark urine. Upon physical examination, the patient exhibited pallor, jaundice, and mild splenomegaly. The patient has a known history of diabetes and is currently on insulin therapy. A complete blood count (CBC) revealed white blood count: 49.5 × 10^9/L, lymphocytic count: 39.5 × 10^9/L, hemoglobin: 8.4 g/dL, mean corpuscular volume: 81.1 fl, Platelets: 183 × 10^9/L, Reticulocyte count10%, lactate dehydrogenase (LDH): 466 U/L, total bilirubin: 2.36 mg/dL, and direct bilirubin: 0.47 mg/dL with positive +3 direct antiglobulin test (DAT). Peripheral blood smear showed normocytic normochromic anemia and absolute lymphocytosis. Flow cytometry was conducted on peripheral blood, and bone marrow which identified CD19, 20, CD79b, and kappa restriction-positive cells but negative expression for CD5, CD23, CD43, CD10, CD103, CD11b, CD25, CD38, and IgM. The results are indicative of Splenic MZL with secondary AIHA.

The treatment commenced with oral prednisone 60mg and folic acid daily, along with Rituximab 750 mg weekly for 4 weeks, which proved to be efficacious in treating both conditions and resulted in a switch recovery in hemoglobin level to 11.0 g/dl, together with other hematological parameters and splenic size. However, two months later, the patient's hemoglobin level dropped to 8.1 g/dl while on prednisone 5 mg daily. Splenectomy was not a viable option due to the patient's age and comorbidities, and prednisone 60 mg with azathioprine 100 mg daily was re-initiated, and the patient was scheduled to receive Rituximab every 8 weeks for two years. Ultimately, the patient displayed Hb stabilization to reach 10.0 g/dl with steroid tapering and monthly CBC monitoring for early relapse detection.

SUMMARY

A 70-year-old male patient initially presented with jaundice and mild splenomegaly. Subsequent laboratory investigations and bone marrow examination led to the diagnosis of splenic marginal zone lymphoma with secondary AIHA. The patient responded well to treatment with corticosteroids and Rituximab. However, the patient experienced a recurrence of AIHA and achieved hematological improvement following regular Rituximab therapy administered every 2 months for a period of 2 years.

A



A: **FIgure A:** Gating was done on 80% of total mononuclear cells with bright CD45 , low side scatter, that express CD19 , CD79b negative to dim , CD81 negative to dim , CD123 partial Positive CD20 ..

DISCUSSION

SMZL comprises 2% of all lymphoid malignancies and 20% of MZL[2]. The typical clinical presentation comprises splenomegaly, lymphocytosis, and cytopenia. Additionally, occurrences of autoimmune hemolytic anemia and other autoimmune phenomena are possible[6].The presence of AIHA in patients diagnosed with SMZL is a significant prognostic indicator, as well as a marker of disease activity. Hence, it is crucial to consider AIHA during the risk stratification of SMZL patients. Rituximab is the preferred course of treatment for this condition.

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