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Case Report

Marginal Zone Lymphoma Presenting in a Patient With Autoimmune Hemolytic Anemia: A Case Report

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ABSTRACT

While autoimmune hemolytic anemia (AIHA) is expected in non-Hodgkin's lymphomas, it is less common in marginal zone lymphoma. Sixty-year-old male patient was followed up with hemolytic anemia in our clinic and while the etiology was investigated, scattered nodes in the mediastinum and abdomen compatible with lymphoproliferative disease were observed on CT, and positive detection of CD5, CD19, CD20, CD22, CD45, HLA-DR, FMC-7 on flow cytometry sent from peripheral blood and a diagnosis of low grade B cell lymphoma (MZL) was made with the result of bone marrow pathology. We found it appropriate to present our MZL case because of its response to steroid treatment and its rare presentation with AIHA.

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Introduction

Autoimmune hemolytic anaemia (AIHA) is an anaemia group characterized by the production of antibodies against autologous erythrocytes.[1] It is divided into primary and secondary.[1] Secondary AIHA can usually be seen in lymphoproliferative diseases, measles, EBV and CMV infection, leukaemia, thymoma, and colon cancer.[2] The lymphoproliferative illness is the most common B-cell chronic lymphocytic leukaemia relationship, but the prevalence in other non-Hodgkin's lymphoma is lower.[3] One of these lymphomas is marginal zone lymphoma (MZL).[4] Herein, we presented a case of the MZL with AIHA

Case Report

A sixty-year-old male patient applied to our clinic with the complaint of fatigue for a month. He had a history of using dual antituberculosis drugs due to pulmonary tuberculosis 18 years ago and mild hepatosplenomegaly on physical examination. Peripheral lymphadenopathy could not be detected. In the laboratory tests at the time of diagnosis, hemoglobin 5.8 g/dL, MCV 104 fl, leukocyte 9.65x10^9/L, neutrophil 5.85x10^9/L, lymphocyte 3.19x10^9/L, platelet 3x10^9/L, corrected reticulocyte 8.39%, sedimentation rate 93 mm/hour, LDH 455 U/L, total/direct bilirubin 1.91/0.7 mg/dL, direct coombs (IgG and Cd3) 3+. anti-nuclear antibody (ANA) detected negative. We observed spherocytes in the erythrocyte series in the patient's peripheral smear *(Figure 1)*.

With AIHA diagnosis, the patient received 1 mg/kg/day methylprednisolone treatment. There was no finding of active disease in the consultation of chest diseases due to a history of tuberculosis. Thorax computed tomography (CT) showed a few lymph nodes below 1 cm in the mediastinum, and small intestine mesentery around the celiac trunk, and extensive lymph nodes in the left paraaortic area consistent with a lymphoproliferative disease on abdominal CT. In the first-month follow-up examinations of the patient, his anaemia improved, but lymphocyte increase developed. The flow cytometry sent from peripheral blood showed that CD5, CD19, CD20, CD22, CD45, HLA-DR, and FMC-7 were positive *(Figure 2)*. With the pre-diagnosis of non-Hodgkin's lymphoma, PET-CT was performed to screen for high involvement lymphadenomegaly. PET/CT in the excision according to lymphadenopathy was not detected. A diagnostic bone marrow biopsy was performed on the patient. The bone marrow pathology result was reported as low-grade B-cell lymphoma (MZL) partially suppressed under steroid therapy. The patient with hemoglobin 12.6 g/dL with treatment was considered steroid responsive. It was planned to stop by decreasing the steroid dose.

Discussion

MZL has first described in 1992.[5] The frequency of MZL appears to be around 1-2% of non-Hodgkin's lymphomas.[5] The median age at diagnosis is approximately 65 years, ranging from 30 to 90, without gender predominance.[6]

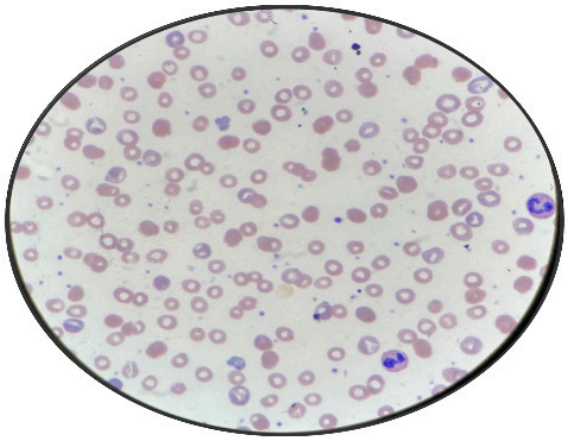
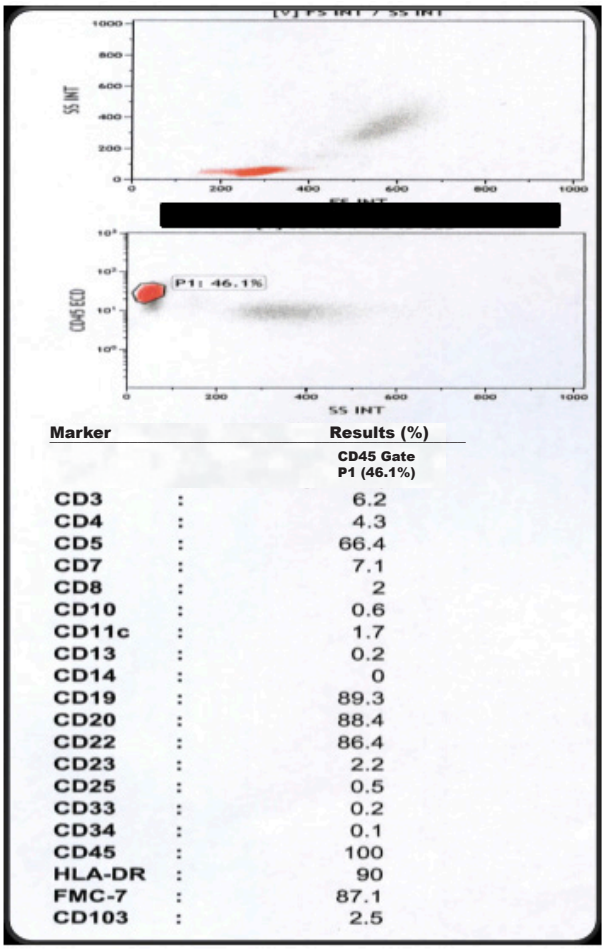


Figure 1. Spherocyte cells in the erythrocyte series in peripheral smear.



Marker	Results (%) CD45 Gate P1 (46.1%)
CD3	6.2
CD4	4.3
CD5	66.4
CD7	7.1
CD8	2
CD10	0.6
CD11c	1.7
CD13	0.2
CD14	0
CD19	89.3
CD20	88.4
CD22	86.4
CD23	2.2
CD25	0.5
CD33	0.2
CD34	0.1
CD45	100
HLA-DR	90
FMC-7	87.1
CD103	2.5

Figure 2. Flow cytometry sent from peripheral blood.

Almost without exception, MZL cases have bone marrow involvement at diagnosis, and roughly one-third have liver involvement.[6] In our case, mild hepatosplenomegaly was present, but no bone involvement was observed. The frequency of AIHA accompanying MZL has been reported as 10% and may be associated with chronic antigen stimulation and autoantibody formation.[2] In the study conducted by Zhou *et al.*[2] in 2020, 11 of 20 patients were followed up with AIHA before diagnosing lymphoma, while 8 were complicated with AIHA during non-Hodgkin lymphoma only one patient was diagnosed with AIHA and lymphoma simultaneously. It has been reported that 2 of these 20 patients belong to the MZL subgroup.[2] In the article published by Hauswirth *et al.*[7], 10 patients with MZL were discussed, and remission was observed after splenectomy in 7 patients and after rituximab treatment in 2 patients. At the same time, only 1 of them responded to steroid treatment.[7]

Conclusions

Our case is rare since MZL presents with AIHA and is responsive to steroids. Since it may be included in the presentation of AIHA lymphomas, patients who apply to the AIHA clinic should be examined for lymphoma.

Conflict of Interests

Authors declare that there is no conflict of interest with regard to this manuscript.

Authors' Contribution

Literature Review, Critical Review, Manuscript preparing held by all authors.

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