



A CASE REPORT ON CHRONIC LYMPHOCYTIC LEUKEMIA PRESENTING AS AUTOIMMUNE HEMOLYTIC ANEMIA

Internal Medicine

Dr. Manoj Kumar Mathur

Professor, Department of Internal Medicine, MLN Medical College, Prayagraj

Dr. Aman Yadav

3rd Year Resident, Department of Internal Medicine, MLN Medical College, Prayagraj

Dr. Deepanshu Dhaneshary

3rd Year Resident, Department of Internal Medicine, MLN Medical College, Prayagraj

ABSTRACT

Chronic lymphocytic leukemia (CLL) is a disorder of the B cell lineage characterized by monoclonal B cell proliferation. It is characterized by the proliferation and accumulation of morphologically mature but immunologically dysfunctional B-cell lymphocytes. A patient with CLL can present with a variety of symptoms. Up to 10% of patients with CLL have an autoimmune hemolytic anemia. This study reported a case of an elderly female presenting with a complaint of easy fatigability due to anemia, later diagnosed to be a case of autoimmune hemolytic anemia as a result of underlying CLL.

KEYWORDS

Chronic Lymphocytic Leukemia; Autoimmune Hemolytic Anemia; Female; Malignancy

INTRODUCTION

Chronic lymphocytic leukemia (CLL) or small lymphocytic lymphoma (SLL) is an indolent malignancy characterized by increased production of mature but dysfunctional B lymphocytes. The median age at diagnosis is between 67 and 72 years. The exact etiology of CLL is not known. Genetic factors, rather than environmental factors, are the most likely cause of CLL. A few known risk factors include occupational exposure to certain chemicals, radiation exposure, and tobacco use.

The incidence is known to rapidly increase with age. CLL has a slightly higher incidence in male populations than female populations (1.3 to 1 to 1.7 to 1). However, studies have shown that women can have a more aggressive form of the disease than men.

CLL is characterized by the clonal proliferation and accumulation of mature, typically CD5-positive B-cells within the blood, bone marrow, lymph nodes, and spleen. The iwCLL guidelines give clear recommendations on how to establish the diagnosis of CLL. Two widely accepted clinical staging systems exist: the Rai system and the Binet system.

Autoimmune hemolytic anemia is a decompensated, acquired hemolysis caused by the host's immune system acting against its own red cell antigens used by autoantibodies directed against red blood cells (RBCs), with or without complement activation. AIHA can be primary or secondary. The common associations are hematological malignancy, infection, and autoimmune disorders. Treatment of second-degree AIHA requires treatment of the underlying disease.

Case Report

A 54-year-old female patient admitted to the hospital who had presented with an on/off fever since 1 month, mild to moderate grade, got relief from treatment from a local practitioner. A general examination reveals pallor and cervical lymphadenopathy, while an abdominal examination reveals splenomegaly. Clinical and laboratory features were recorded (Hb-6, TLC-55000, and lymphocytes 65%). Routine investigation has shown anemia and lymphocytosis.

On evaluation of lymphocytosis, peripheral blood smears revealed small, mature lymphocytes in excess, with a slightly raised N:C ratio. Flow cytometry reports 63% lymphoid cells gated in bright CD19, which are low-side scatter positive for CD5, CD23, CD200, CD20, CD45, KAPPA, and negative for CD10, FMC7, CD3, CD7, CD8, and lambda, suggesting a diagnosis of CLL. The FNAC of the enlarged cervical lymph node showed monomorphic, small, immature lymphoid cells. The bone marrow aspirate supported the PBS finding showing a small, uniform lymphoid cell (97%).

The patient had anemia; on evaluating for it, it was found to be autoimmune hemolytic anemia positive with IgG-type antibodies (the

Direct Coombs Test, also called a direct antiglobulin test). To guide treatment and prognosis, the patient had undergone FISH for cytogenetic analysis, which revealed trisomy 12 in 70% of the clonal cells studied. There are no abnormalities in the TP53, ATM, or 13q categories of patients at intermediate risk.

The patient was classified using Rai and Binet systems, with Rai III and Binet C being the staging systems.

CLL IPI score of 5 (unmutated IGHV, serum beta 2 microglobulin 3.7, bone stage C)

Based on these results, Ibrutinib (IBR) treatment (420 mg OD) was started. IBR is a first-in-class oral inhibitor of Bruton tyrosine kinase (BTK) approved for the treatment of relapsed CLL. IBR blocks downstream signaling of the B-cell receptor, disrupting stromal microenvironment interactions and B-cell cytokine signaling.

DISCUSSION

In this case report, we have emphasized investigating and treating the underlying cause of anemia in patients with chronic lymphocytic leukemia. The patient improved, and hemolysis subsided after taking Prednisolone 1 mg/kg body weight and tapering 10 mg every 2 weeks. He sustained remission at 10 mg/day in 8 weeks.

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