INTRODUCTION

Hairy cell leukemia (HCL) is a relatively rare form of leukemia characterized by an insidious onset, massive splenomegaly without lymphadenopathy, pancytopenia, dry bone marrow aspirations and the presence of abnormal mononuclear cells with circumpetalent prominent hairy cytoplasmic projections in the peripheral blood. The highly suggestive nature of such situations (splenomegaly with few atypical lymphoid cells) led to the examination of bone marrow for morphological findings characteristic of hairy cell leukemia (HCL) with an increased reticulin framework. Diagnosis was confirmed when flow cytometry was positive for hairy cell markers including CD11c, CD103 and FMC-7, and negative for splenic lymphoma with villous lymphocyte (SLVL) markers including CD19 and CD25. Accurate diagnosis of this entity and its differentiation from other lymphoproliferative disorders is essential in view of therapeutic and prognostic considerations. Biomed. Int. 2011; 2: 36-38. ©2011 Biomedicine International, Inc.

Key Words: Hairy cell Leukemia, lymphoproliferative disorder, splenomegaly
fibrosis. Reticulin staining on a marrow biopsy demonstrated increased reticulin fibers. Flow cytometry revealed positivity for the immunophenotypic markers CD19 (87%), CD20 (89%), CD11c (96%), CD103 (hairy cell marker) (84%), FMC–7 (85%) and HLA-DR (98%), confirming the diagnosis of hairy cell leukemia. The patient responded well to the chemotherapeutic agent 2-chlorodeoxyadenosine (Cladrabine), resulting in a reduction of spleen size and normalization of blood counts.

DISCUSSION

Hairy cell leukemia is a rare disorder, accounting for 2% of all leukemias. HCL was first recognized by Ewald in 1923, who described the condition as leukemische reticuloendotheliose. The disease is more common in Caucasians and particularly frequent in Ashkenazi Jewish males, with an overall male to female ratio of approximately 4:1. The median age of onset is 52 years. HCL has been classified into three types: HCL-classic, variant HCL (HCL-V, type II HCL) and Japanese variant HCL (HCL-J). It is important to diagnose these entities accurately as they have different clinical and biological features, particularly regarding the response to α-interferon. Morphological evaluation of a peripheral blood smear is an extremely valuable tool in screening for HCL. The disease may go undetected when very low levels of hairy cells are present in the peripheral smears.

The basic mechanisms involved in the pathogenesis of HCL are poorly understood. Recent studies have demonstrated that the hairy cells are mature B cells. They have mutated IgH genes, suggesting post-germinal center, antigen-acquainted memory cells. They have low mitotic cycling rates and thus a protracted clinical course, but a highly activated cytokine transcription apparatus including a cytokine storm, which is responsible for diverse clinical and morphological features.

Differential diagnosis of HCL include B-Chronic Lymphocytic Leukemia (CLL), Prolymphocytic leukemia and T-cell lymphoproliferative disorders such as Hepatosplenic γδ T-cell lymphoma and Splenic B-cell lymphoma including splenic lymphomas with villous lymphocytes (SLVL). The cells of CLL differ from hairy cell leukemia as they have more coarsely clumped chromatin and round or ovoid nuclei. Hairy cells are intermediate-sized lymphocytes that possess round to oval nuclei and an abundant light blue agranular cytoplasm with characteristic microfilamentous (‘‘hairy’’) projections. They strongly express CD103, CD22 and CD11c. These cells typically infiltrate the bone marrow, the spleen and to a lesser extent the liver, lymph nodes and skin. Recently, immunohistochemical demonstration of Annexin A has been reported to be a 100% specific marker for HCL. Prolymphocytic leukemia occurs in elderly male individuals with a median age of 70 years. Patients usually present with peripheral lymphadenopathy, leucocytosis, TRAP inactivity, CD5+, CD19+, CD25-, CD103- and CD10-.

Until the mid-1980s, splenectomy was the predominant therapy for HCL, providing improvement of cytopenias in the majority of patients and a normalization of blood counts in 40% to 60% of individuals. Treatment has been revolutionized with the advent of interferon (IFN)-α and purine analogues (PA), which provide an overall response rate of 75-100%. Rituximab has also been used to treat relapsed/refractory HCL with an overall response of 80%. In the present case, the patient presented with a normal leucocyte count, which is not a characteristic feature of HCL. However, this case revealed few atypical lymphoid cells in the peripheral smear, leading to further investigations, confirming the diagnosis of HCL with normal leucocyte counts. Therefore, it necessary to perform a peripheral smear examination with the normal leucocyte counts, even with a single lineage cytopenia.

An erroneous diagnosis of other lymphoreticular disorders can lead to aggressive chemotherapy, which is contraindicated and hazardous for patients with hairy cell leukemia. Careful attention to morphological details is important for early diagnosis, particularly when low percentages of hairy cells are present.
in the peripheral blood and the bone marrow. Early diagnosis of HCL is important to ensure that patients obtain maximum benefit from new therapeutic agents that have greatly improved prognosis in this rare disorder.

REFERENCES