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Case Report

Hairy cell leukemia: a rare case report

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ABSTRACT

Hairy cell leukemia (HCL) is a rare chronic B-cell leukemia accounting for 2% of all the leukemias and occurs more frequently in the elderly males. The etiology is unknown but possible relationships to radiation exposure, exposure to benzene, to farm animals and to commercial herbicides and pesticides have been identified. Familial predisposition among first degree relatives has been noted. It is characterized by distinctive cytoplasmic "hair like" projections on the cell surface of lymphoid cells, pancytopenia and splenomegaly. We report a rare case of 29 year old female, farm labourer presenting with fever, fatigue and weakness for 1 month. On examination the patient had hepatomegaly, massive splenomegaly and inguinal lymphadenopathy. After peripheral smear examination diagnosis of HCL was made which was confirmed by bone marrow aspiration examination, bone marrow biopsy and immunohistochemistry (IHC).

Keywords: Hairy cell leukemia, Pancytopenia, Splenomegaly, Immunohistochemistry

INTRODUCTION

Hairy cell leukemia (HCL) is a rare chronic B-cell leukemia accounting for 2% of all the leukemias and occurs more frequently in the elderly males. It is characterized by a triad of splenomegaly, pancytopenia and monocytopenia. The characteristic cells are mononuclear having abundant cytoplasm exhibiting thin projections that extend circumferentially over the cell surface with round or oval nuclei infiltrating the bone marrow diffusely and involving the red pulp of the spleen. Only a small number of leukemic cells are seen in the peripheral blood (PB) in a classical case.[1-3]

The distinction between HCL and other chronic B-cell lymphoproliferative disorders is clinically important because of differing treatment protocol and an indolent clinical course.

Patients with HCL are highly sensitive to purine analogues such as cladribine. Apart from morphology, HCL has a characteristic immunophenotypic profile and light scatter characteristics. The tumor cells express B cell-associated markers i.e. CD19, CD20, CD22 and CD79b. Coexpression of CD103, CD11c and CD25 is considered unique for HCL and is often used as an absolute criterion for establishing the diagnosis of HCL. However, atypical immunophenotypes have been reported in an otherwise morphologically classical HCL.[5,8] Here we present a rare presentation in a young female.

CASE REPORT

A 29 year old female presented with history of fever, fatigue and weakness –on and off for 1 month: there were no other significant complaints. On examination mild

pallor was present. Abdominal examination revealed enlarged, firm, nontender splenomegaly 10 cm below the costal margin, hepatomegaly 5 cm below the costal margin and inguinal lymphadenopathy.

Hematological examination revealed haemoglobin (Hb) concentration of 3.1gm/dl, a total leukocyte count of 30.8x10³/µL, red blood cell (RBC) count of 1.07x106/L, platelet count of < 10x103/µL, hematocrit 10.3%, MCV 96.3fL, MCH 29pg, MCHC 30.1g/dL and a reticulocyte count of 0.7. A peripheral smear demonstrated dimorphic population of microcytic hypochromic and macrocytic normochromic red cells with anisopoikilocytosis+, few elliptocytes and tear drop cells with a differential count of 01% polymorphs, 03% lymphocytes and 96% atypical lymphoid cells with round to oval nuclei, moderate cytoplasm and frayed margins with hair like projections (Figure 1 & 2).

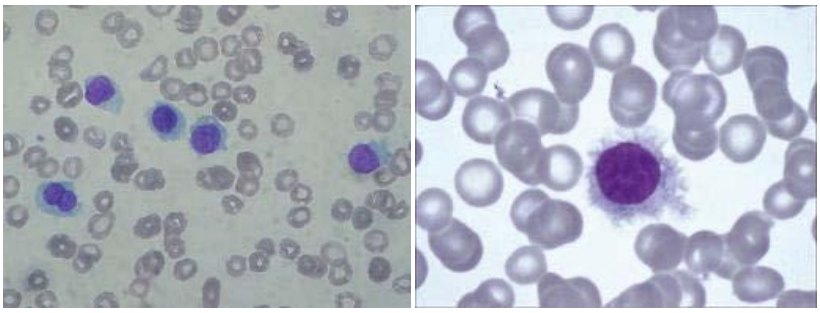

Figure 1 & 2: Photomicrographs showing atypical cells with light basophilic cytoplasm containing numerous hairy projections on the outer surface in a peripheral smear.

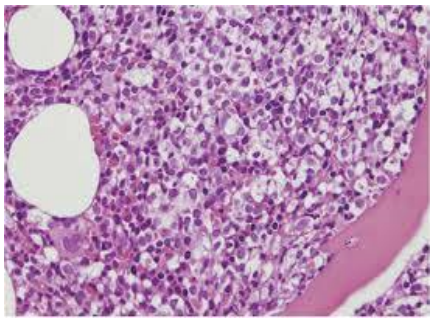

Figure 3: Photomicrograph showing atypical lymphoid cells with cytoplasmic projections in a bone marrow aspirate.

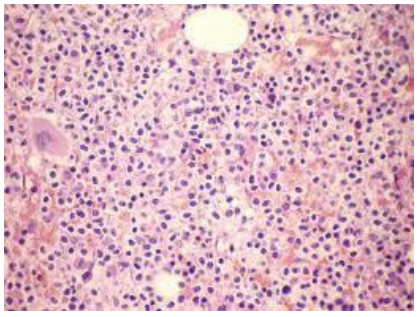

Figure 4: Photomicrograph showing typical fried egg appearance of hairy cells in a bone marrow biopsy.

Bone marrow aspiration shows 82% atypical lymphoid cells with cytoplasmic hair like projections (Figure 3).

Trephine bone marrow biopsy revealed the typical fried egg appearance of hairy cells with fibrosis (Figure 4).

Flow cytometry revealed positivity for the immunophenotypic markers CD19, CD20, DBA44, TRAP, CD11c, CD103 confirming the diagnosis of hairy cell leukemia.

DISCUSSION

Usually patients are in the age group of 45-65 years with male:female ratio of 4:1. They present with fatigue, weight loss and moderate to massive splenomegaly. Hepatomegaly is present in nearly 50% of the cases. Recurrent infections are a major manifestation and important cause of death.[4] The HCL variant makes up 10% of all the HCL cases. The main features are splenomegaly, lymphocytosis and cytopenias without monocytopenia. The circulating cells have a morphology intermediate between prolymphocytes and hairy cells. The immunophenotype shows a mature B-cell phenotype

with expression of B-cell antigens CD11c and CD103 but unlike typical hairy cell the cells are negative for CD25. The histology of bone marrow and spleen shows a pattern of infiltration similar to that in HCL.[5-8] It differs from classical HCL as shown in the table below:

Table 1: HCL vs HCL-V.

Hairy Cell Leukemia (HCL)	HCL-Variant (HCL-V)
Usual age is 50-65 years	Older patients 61-82 years
Pancytopenia is the characteristic feature	TLC is higher, usually > 50 x 109/L
Monocytopenia present	Monocytopenia is not present
Low N/C ratio, finely dispersed chromatin without prominent nucleolus	Higher N/C ratio, more condensed chromatin with a single prominent nucleolus
TRAP +ve cells	TRAP –ve cells/weakly +ve
CD 25+, HC 2+, CD 103, CD 11c+ve, CD 123+	CD 25 –ve, HC 2-, CD 123 –ve
Marrow is not aspirable because of reticulin fibrosis	Marrow is cellular and aspirable
Good response to IFN-α and 2-CDA	Poor response to IFN-α and 2-CDA
Good survival	Poor survival

HCL is a rare, chronic B-cell leukemia characterized by splenomegaly and pancytopenias, usually seen in elderly males. Though pancytopenia is common, occasional patient can present with normal blood counts or leukocytosis. Occasionally characteristic hairy cells are found in peripheral smear, however, bone marrow biopsy is required to make the diagnosis. Immunohistochemistry is used for confirmation using CD 103, CD 20, CD 11c.

Several treatment options are available. The most active agent in this disease is a single course of cladribine also known as 2-CdA. With a single course there is about 80% chance of cure in young people. Adverse effects include myelosuppression and infections.[1,3,7,8]

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