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Plasma Cell Leukemia Mimicking Hairy Cell Leukemia: Extended Role of Immunophenotyping in Correct Diagnosis

Introduction

Plasma cell leukemia (PCL) is a scarce and belligerent type of plasma cell dyscrasia. Patients with PCL have a very poor prognosis with median survival measured in months. There is presence of more than 20% plasma cells on peripheral blood film however, morphology can sometimes be misleading and may result wrong diagnosis with delay in treatment. Therefore, it is always important to confirm the probable diagnosis with additional testing.

Plasma cell leukaemia with hairy cell morphology has been reported in some case reports earlier^[1–3]. Chan et al published a case report of plasma cell leukaemia with hairy cell morphology showing good response to Bortezomib based therapies^[4].

Currently, multiparameter flowcytometry technique for immunophenotyping of neoplastic cells is considered to be mandatory for the diagnosis^[5].

CD38 is widely expressed on plasma cells and may be present on hematopoietic cells. Having said that, it should be keep in mind that the uniquely bright intensity of CD38 is typically observed on plasma cells with an expression that is higher than other hematopoietic cell populations. Hence it is considered a "must" for plasma cell profile. In addition, CD138 staining of hematopoietic cells is also a specific feature of plasma cells. We report a case of a patient which showed hair-like projections on morphology of abnormal cells on peripheral smear while immunohistochemistry and immunophenotyping confirmed the diagnosis as plasma cell leukemia.

Case Report

A 68-year-old male, presented with a history of weakness, body aches, and weight loss for 2 months. Physical examination revealed pallor and hepatomegaly. Baseline peripheral blood counts showed Hb 10.7 g/dl, Hct 37.2%, mean corpuscular volume 103.9 FL, mean corpuscular hemoglobin 34.5 PG, white blood cells 56.2×10^9 /L, absolute neutrophil count 2.5×10^9 /L, and platelets 74×10^9 /L. His peripheral blood film showed 60% lymphoplasmacytoid cells with hair-like

projections [Figure 1a and b], and bone marrow aspirate revealed infiltration with plasma cells, which exhibited pleomorphic features and cytoplasmic projections [Figure 1c and d]. Bone trephine biopsy was performed, which showed interstitial infiltration with plasma cells [Figure 1e] and suppressed hematopoietic precursors. These plasma cells were positive for CD 138 [Figure 1f] immunohistochemical stain. Immunophenotyping was also performed which showed positivity for CD 138, CD 56, and CD 38. Gated population was negative for myeloid markers such as CD 13, CD 33, and CD 117 and lymphoid markers such as CD3, CD 5, CD 7, CD 10, CD 11c, CD 19, CD 20, CD 22, CD 23, CD 25, and CD 103 along with cCD79a while population was also negative for CD 34 and terminal deoxynucleotidyl transferase. Hence, the diagnosis of plasma cell dyscrasia (plasma cell leukemia) was made.

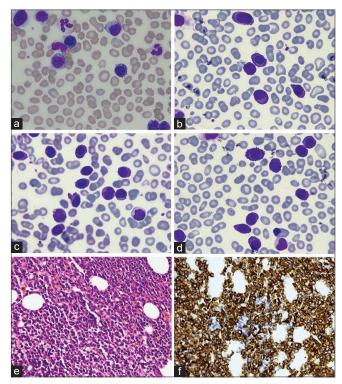


Figure 1: (a and b) Peripheral blood film showing hairy plasma cells. (c and d) Bone marrow aspirate showing infiltration with plasma cells having hair like projections. (e) Bone trephine specimen showing infiltration with plasma cells and suppressed haematopoiesis. (f) Positive staining of plasma cells with CD138 immunohistochemical stain

Memon and Ali: Hairy plasma cells

The biochemical evaluation was done, which showed raised creatinine level (2.8 mg/dl) and lactic dehydrogenase level (1308 IU/l). He was advised treatment but was lost to follow.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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