UNUSUAL PRESENTATION OF HAIRY CELL LEUKEMIA: A CASE REPORT

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Abstract

Hairy cell leukemia is a chronic B cell lymphoproliferative disorder, which is uncommon and constitutes around two percent of hematolymphoid malignancies. HCL commonly involves bone marrow and spleen and rarely peripheral blood. Splenomegaly is a prominent feature and is seen in around 70 to 100% of HCL cases as reported in various case reports. Sometimes the absence of splenomegaly rules out the diagnosis of HCL and is misdiagnosed as aplastic anemia. Thus the aim of our study is to understand the importance and keep a high level of suspicion in such cases. As in present case there was no evidence of splenomegaly clinically or radiology, but the morphologic features on biopsy had suggested HCL which was further confirmed on immunophenotyping. The purpose of this case report is to highlight the importance of the fact that HCL can present even without splenomegaly.

Keywords: Hairy cell leukemia, Splenomegaly, Immunohistochemistry

Introduction

Hairy cell leukemia (HCL) is a chronic B cell lymphoproliferative disorder, which is uncommon and constitutes around two percent of hematolymphoid malignancies. It is common in middle-aged to elderly and presents mainly with Pancytopenia, monocytopenia and splenomegaly. HCL commonly involves bone marrow and spleen and rarely peripheral blood(1).

Splenomegaly is a prominent feature and is seen in around 70 to 100% of HCL cases as reported in various case reports (2,3). Many patients who are otherwise asymptomatic comes for the medical attention because of presence of mass in left side. Sometimes the absence of splenomegaly rules out the diagnosis of HCL and is misdiagnosed as aplastic

anemia and which further delays the diagnosis and proper and timely management.

Thus the aim of our study is to understand the importance and keep a high level of suspicion in such cases based on morphologic features on histology even without the clinical features. As in our case there was no evidence of splenomegaly clinically or on imaging investigation, but the morphological features on biopsy had suggested HCL which was further confirmed on immunophenotyping.

Case Report:

A 72 year old with known case of CAD & HTN presented with 2 months history of generalised weakness and fatigue. He had received 3 units of PBCs in the past. On examination, pallor was present; however he had no hepato-splenomegaly or lymph node enlargement. His complete blood counts

showed pancytopenia with Hemoglobin 8gm/dl , total leucocyte count 1800 per cumm and platelet count of 72000 per cumm. Peripheral blood smear showed occasional atypical cells with hair like projections. Bone marrow biopsy showed intervening hypercellular marrow with interstitial increase in atypical lymphoid cells. These cells are small to intermediate in size, have scant to moderate cytoplasm, round nuclei & clumped chromatin. Trabeculae are normal. Reticulin showed moderate patchy increase. A few lymphoid cells with hairy projections were also seen. Immunohistochemistry(IHC) for Annexin A1 & DBA was positive in these atypical lymphoid cells confirming the diagnosis of HCL. Patient then received standard dose chemotherapy with fludarabine for 10 days in continuous infusion and was disease free later on .

Figure 1: Bone marrow biopsy section (H&E stain, 100X) shows hypercellular marrow.

Figure 2: Bone marrow biopsy (H&E stain, 400X) shows atypical lymphoid cell and few cells show hairy projections.

Figure-3: Immunohistochemical stain on bone marrow biopsy (400x) shows CD 20 immunoreactivity in atypical cells.

Figure 4: Immunohistochemical stain on bone marrow biopsy (400x) shows DBA 44 immunoreactivity in atypical cells.

Figure 5: Immunohistochemical stain on bone marrow biopsy (400x) shows ANNEXIN A1 immunoreactivity in atypical cells.

Discussion:

HCL is a chronic lympho-proliferative disorder that can present with pancytopenia or monocytopenia, and accompanied with splenomegaly and inaspirable bone marrow due to fibrosis. In various differential diagnosis of dry tap and presence of atypical lymphoid cells such as in aplastic anemia, hypoplastic myelodysplastic syndrome, atypical CML, B-Prolymphocytic leukemia and idiopathic myelofibrosis, HCL should always be considered due to its unusual presentation(4).

Amongst all clinical findings, splenomegaly is the most consistent physical finding. HCL is also considered in differential diagnosis in all cases having of splenic enlargement. Hence all such cases with such unusual presentations are challenging for both clinician and pathologists as peripheral smear shows only few of these atypical cells and bone marrow is also unyielding in such cases due to fibrosis. Hence bone marrow biopsy is an important modality and should always be carefully evaluated in all such cases of HCL without splenomegaly as the marrow is hypocellular in such cases.

Various case reports had shown the incidence of HCL without splenomegaly ranged from 0 to 30%. In around 7% of cases of HCL without splenomegaly, bone marrow is hypocellular, suggesting that the disease is in early stage. In some studies bone marrow hypocellularity is reduced upto 18 %(5).

It can be speculated that in near future these hypoplastic variant is on rise as more and more bone marrow examination are done in asymptomatic patients presented with cytopenias and thus pathologists should always keep suspicion of HCL in

such cases. Hence it is to emphasize that early diagnosis is not only beneficial for the patients as they can obtain maximum benefits from the newer drugs but can also improve the prognosis in this rare disorder.

Conclusion:

The idea of this case report is to highlight the importance of the fact that HCL can present even without splenomegaly, therefore a high level of suspicion should be kept in all cases showing atypical hairy lymphoid cells on peripheral smear and thus careful evaluation of marrow biopsies should be done in order to diagnose the disease in early stage and benefit the patients through the newer drugs and improve the overall prognosis of this rare disease.

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