Case Report

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A Febrile Patient with Polyarthritis, Visceromegaly and Cytopenias

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Funding Source: None
Conflict of Interest: None

Received: Dec 21, 2020
Accepted: Mar 09, 2021

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A B S T R A C T

A case report of asymmetrical polyarthritic patient with history of fever of 6 months duration. Clinically also had hepatosplenomegaly. The lab reports revealed Cytopenias with TLC of TLC 1830µl, Hb 9.5g/dl, platelets 44000mL, and high ESR that is 75. Patient was worked up for possible SLE and RA, bone marrow biopsy was done for Cytopenias. While RA, ANF and Anti-DNA came out positive, thus bone marrow revealed hairy cell leukaemia, which was further confirmed by immuno-histochemistry. Patient was started on steroids due to painful arthritis and discharged to be followed up after two weeks and mediation for HCL. Surprisingly, the CBC report reverted significantly normal this period. Now the distinguishing point is, unlikely for hairy cell leukaemia to respond steroids so quickly within 10 days, or this was all SLE related manifestations and HCL was an incidental finding during the workup of pancytopenia. On further follow up, we will do BRAF and flow cytometry for HCL.

Keywords: Arthritis, Hepatomegaly, Splenomegaly, Cytopenias.

Cite this article as: *Kakar ZE, Qazi A, Khan A, Hafeez N, A Febrile Patient with Polyarthritis, Visceromegaly and Cytopenias. Ann Pak Inst Med Sci.2021; 17(1):78-79.* doi. 10.48036/apims.v17i1.413

Introduction

Hairy cell leukemia is a rare hematopoietic stem cell disease that is classified as mature B-lymphocytes with cytoplasmic projections which presents predominantly in elders. The average age of presentation is 55 years and male to female ratio is 5:1.[1] Common presentation involves pancytopenia, although sometimes the patient has a leukemic presentation such as a slight onset of fatigue and other symptoms related to leukemia i.e., a severely enlarged spleen and other associated symptoms.[2] Some patients come with infection of mycobacterium intracellular and vasculitic syndromes. Hairy cell leukemia is usually an indolent disorder; its course is dominant by pancytopenia and recurrent infection. Splenomegaly is almost always present and can be large, the liver is enlarged by 50% and lymphadenopathy is not usually common.[3] HCL is a rare case . Here the unique combination of two disease is the interesting point. Where as HCL looked as a dominating disease initially but worked up led to the main diagnosis of SLE and HCL as a co-incidental disease not requiring treatment

Case Report

A 57 years old lady is diagnosed case of type 2 diabetes mellitus, for last 2 years on oral anti diabetic drugs like metformin 250 mg twice a day with good compliance and control. She was presented with complaint of low grade fever for last 7 months documented up to 101 Fº with associated chills and rigors gradual in onset and used to relieve by taking antipyretics. She had complained occasional episodes of vomiting, significant 14 kg weight loss, associated body aches and pain. These symptoms were associated with generalized body bruises over arms and legs for last 3 months. At that time her blood complete count was done which revealed bicytopenia. Examination revealed that she was markedly pale with no palpable lymphadenopathy was there. She was also having generalized bluish discoloration over the body and has hepatomegaly of size 9cm with splenomegaly of size 6cm. She was also having multi nodular goiter.

CBC was showed pancytopenia, TLC 1210µl, Hb% 8.7 g/dl, platelets 44000 mL, RETICS 0.8%, LFTs & RFTs, alkaline phosphatase (ALP) 449IU/L, TSH 1.38 mIU/L, T3 0.48 mIU/L, T4 0.88 mIU/L, Ferritin 4677 µg/L, BSR 190 mg/dl, HbA1C 8.3%, Coagulation profile with normal limits, ANA positive, Anti-ds DNA positive and serum electrolyte was with normal limits (WNL). This case Has been managed as SLE alone with complete

resolution of clinical issues and recovering of haematological parameters which were initially attributed to HCL

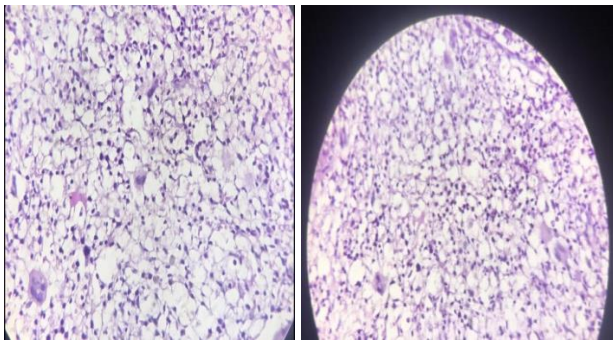

Figure 1. Hairy cell of bone marrow are seen under the microscope

Discussion

A febrile patient was presented with joint pain initially then polyarthritis (asymmetrical) also had hepatosplenomegaly with pancytopenia and raised ESR. Previous studies for RA were negative. But this time work up revealed in the clinical back ground of pancytopenia & investigating collagen vascular disorders like RA and SLE. So this is a rare presentation of hairy cell leukemia (HCL). Clinical manifestations of hairy cell leukemia (HCL) are distinguished from other lymphoproliferative diseases such as Waldenstrom macroglobulinemia & non Hodgkin lymphoma. It also may be confused with other causes of pancytopenia including hypersplenism due to aplastic anemia & paroxysmal nocturnal hemoglobinuria (PNH).[4] Symptoms of hairy cell leukemia are pancytopenia. Anemia is almost universal & 75% of patients with thrombocytopenia & neutropenia. "Hairy cells" tend to be present in small numbers at the edges of a blood smear and have a similar appearance to multiple cytoplasmic specimens. Bone marrow transplantation is usually stimulated (dry tap) and the diagnosis is made by markers of tartrate-resistant acid phosphate (TRAP),[5] in the immunization of cells that produce antigens CD11, CD20, CD103 & CD123. Pathological tests show the complete penetration of red blood cells by hairy cells. This compares with the common consideration of lymphomas involving white pulp of pulse.[4] Treatment is indicated for symptomatic disease i.e., splenic dysfunction, recurrent infections or important cytopenias. So the patients should be cross checked.[6]

Corticosteroids have been used in individual cases, such as in our patient. Of the 46 patients of the Bouroncle series treated with prednisolone, at one point only during the observation period only 5 showed a temporary improvement in bone function and a decrease in the size of the spleen.[7]In the patient, there is improvement in bone marrow as well as symptomatic improvement in the pain in multiple joints. So it is important to mention in our case that it highly unlikely for HCL to response to steroids within 10 days or this was all SLE related manifestations and HCL was an incidental finding during the work up of pancytopenia.

Conclusion

This case describes a rare but a very important as of hairy cell leukemia which is not predicts the disease outcome but also mislead the physician's management. Therefore, the physicians need to be well aware of the unpredictable treatment of HCL. So as to avoid any treatment like chemotherapy, an ongoing research will unveil other potential treatment for this appropriate pancytopenia.

Though we have limitations in the diagnosis of our patient that the V600E mutation in the BRAF gene & flow cytometry.

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