Case Report

Hairy Cell Leukemia Presenting with Duodenal Involvement Duodenum: A Case Report

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Abstract

Context: A rare case of adult hairy cell leukemia (HCL) with duodenal involvement is presented. **Case Report:** The patient was a 48-year-old man, who had a history of hairy cell leukemia. Three days after completion of 2-chlorodeoxyadenosine (CDA) chemotherapy, the patient started experiencing abdominal pain. An extensive gastroenterological workup culminated in the patient getting an esophagogastroduodenoscopy (EGD) that revealed duodenal infl ammation and biopsies were taken. The duodenal biopsy was positive for chronic inflammatory infiltrate, primarily consisting of atypical lymphocytes and plasma cells with tartrate-resistant acid phosphatase (TRAP) positivity, and hence a diagnosis of duodenal involvement with HCL was made. Repeat bone marrow biopsy done 2 weeks after finishing chemotherapy revealed residual disease. At the 3-month follow-up, the patient was asymptomatic with a normocellular marrow and no residual disease. Repeat abdomen computerized tomography (CT) scan at completion of therapy showed resolution of duodenal thickening and spleen size of 12 cm. Currently, patient is in clinical remission for 6 years with 4-6 monthly follow-up visits and continues to do well. **Conclusion:** This case is presented to highlight the first case report of HCL with duodenal involvement that was successfully treated with CDA.

Keywords: Hairy cell, Leukemia, Duodenum, Chlorodeoxyadenosine

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Introduction

Hairy cell leukemia (HCL) is a chronic B-cell lymphoproliferative disorder that obtains its name from the fine hair-like projections seen on the cell surfaces under microscopy. Clinically, HCL is associated with splenomegaly and pancytopenia while the cells infiltrate the bone marrow, liver, and spleen. However, unlike most mature B-cell lymphomas, HCL shows minimal lymph node involvement and absence of chromosomal translocations.[1] The nature of its spread is attributed to its unique homing properties being limited to the blood compartments.[2] Tartrate-resistant acid phosphatase (TRAP) positivity is diagnostic of HCL.[3]

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Skin metastases, Sweet syndrome and panniculitis have been rarely reported.[4-6] We hereby report a very rare case of duodenal involvement in HCL.

Case Presentation

A 48-year-old Hispanic male presented to the emergency room (ER) with colitis in March 2008. Hematological workup revealed pancytopenia (hemoglobin, 6.9 gm; leukocyte count, 4800/uL; platelet count, 23,000/uL). Computerized tomography (CT) scan abdomen revealed a massive splenomegaly (17 cm). Bone marrow aspiration and biopsy showed hypercellular marrow diffusely infiltrated by small lymphocytes with round to bean shaped nuclei with reticulated chromatin pattern and abundant cytoplasm. Immunohistochemical staining showed monoclonal kappa expressing B cell population positive for CD19, CD22, and CD45 and TRAP [Figure 1] while negative for CD23, CD10, CD5, and CD3. Reticulin stain showed diffuse increase in reticulin fibers in the marrow. A subsequent diagnosis of HCL was made and the patient received outpatient 2-chlorodeoxyadenosine (CDA) 0.1 mg/kg for 5 days.

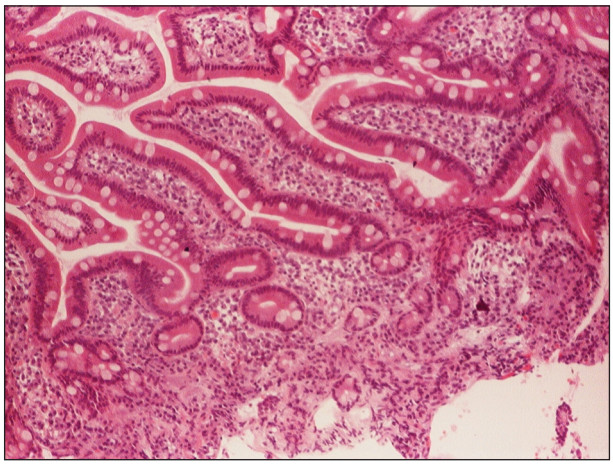

Figure 1: Bone marrow biopsy demonstrates numerous hairy cells admixed with other elements

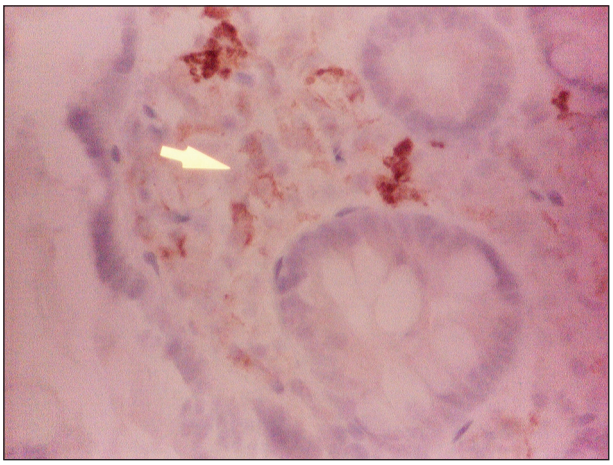

Figure 2: TRAP stain of HCL showing bright granular cytoplasmic positivity in the leukemic cells infiltrating the duodenum

As supportive management, he also received 2 units of packed red cells. He was readmitted 3 days after completion of chemotherapy with febrile neutropenia, severe abdominal pain, and skin rash. Infectious etiology work up and biopsy of the skin rash were negative. Broad-spectrum antibiotics and antifungals were given empirically. A CT scan of abdomen revealed thickening of the duodenum. Subsequently an esophagogastroduodenoscopy (EGD) revealed duodenal ulcerative inflammation and biopsies were performed. The duodenal biopsy was positive for chronic inflammatory infiltrate, primarily consisting of atypical lymphocytes and plasma cells with TRAP positivity and hence a diagnosis of duodenal involvement with HCL was made [Figure 2]. Repeat bone marrow biopsy done 2 weeks after finishing chemotherapy revealed residual disease. At the 3-month follow-up, the patient was asymptomatic with a normocellular marrow and no residual disease. Labs showed a leukocyte count of 2800/uL, hemoglobin at 10 gm%, and platelet count of 246,000/uL. Repeat abdomen CT scan in March 2009 showed resolution of duodenal thickening and spleen size of 12 cm. Currently, the patient has been in clinical remission for 6 years with 4-6 monthly follow up visits and continues to do well.

Discussion

HCL is rare B-cell lymphoproliferative disorder with annual incidence of 3 cases per million populations. HCL is predominately seen in males and Caucasians with a median age of presentation being 52 years. HCL arises from late-activated memory B-cells. The World Health Organization (WHO) classifies HCL as mature B-cell neoplasm with predilection for splenic involvement and certain immunophenotypic characteristics.[7] Classically, diagnosis of HCL was confirmed by TRAP activity, although the standard practice today is immunophenotyping by flow cytometry where HCL is characterized by the expression of B-cell antigens CD19, CD20, and CD22 in addition to coexpression of the surface antigens CD11c, CD25, and CD103. Hairy cells generally lack CD5, CD10, CD21, and CD23. The classical cytologic features of the HCL include mature lymphocyte of a small size and round nuclear contour with a condensed chromatin and indistinct nucleolus, abundant pale cytoplasm, and circumferential cytoplasmic projections. Microscopically, HCL cells have fine hair-like projections and abundant cytoplasm. The finding of hairy cells having nuclei widely separated by abundant cytoplasm, resulting in the so-called "fried-egg" appearance, can sometimes help to confirm the diagnosis. Clinically, these patients present with splenomegaly, pancytopenia, and the bone marrow is usually involved in all patients with HCL. Typical sites that are involved by HCL are the bone marrow and splenic red pulp; the disease can also be more widespread and involve extramedullary sites such as the central nervous system, gastrointestinal and urogenital tracts, heart, lungs, skeletal muscle, skin, thymus, and thyroid.[8-10] Involvement of the bone marrow in HCL with associated reticulin fibrosis results characteristically in hypocellular aspirate smears or dry tap. Our patient presented with residual bone marrow disease at the time of diagnosis of duodenal HCL.

This, to our knowledge, is the first case report of HCL manifesting clinically with duodenal involvement. The patient was successfully treated with CDA.

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