

Case Report

HAIRY CELL LEUKEMIA: A CASE REPORT

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ABSTRACT

Hairy Cell Leukaemia (HCL) is defined, according to the WHO classification, as a mature (peripheral) B-cell neoplasm. HCL accounts for between 2–3% of all leukaemia cases, with about 600 new cases diagnosed in the U.S. each year. In HCL, therapy is extremely effective in controlling the disease and may even achieve complete remission. The cell biology, pathogenesis and molecular genetic defects are not well understood. Its association with other malignancies has been implicated. In the present case we have discussed clinical and morphological features of a patient suffering from HCL. We have summarized classic morphologic features as seen by light and immunohistochemical findings. Diagnostic criteria and differential diagnosis are also discussed.

Key Words: *Hairy Cell Leukaemia, Pancytopenia, Immunohistochemistry*

INTRODUCTION

HCL is a rare chronic B cell lymphoproliferative disorder characterized by splenomegaly, pancytopenia and bone marrow infiltration by atypical lymphocytes with circumferential prominent hairy cytoplasmic projection (Goodman, 2003). HCL has been known by several names including leukemic reticuloendotheliosis, lymphoid marrow fibrosis, medullasplenic histiolymplocytosis of primitive appearance and reticulum cell leukaemia (Bouroncle, 1958). The median age at the time of diagnosis is 52 years with a male to female ratio of 4:1 (Staines, 1993). A number of non-malignant disorders have been associated with HCL. There are also several reports suggesting an association with systemic immunologic disorder such as scleroderma, polymyositis and various vasculitides. A humoral or cell mediated response initiated by leukemic cell is thought to be responsible for HCL (Nighat, 2011). Here we report a case of HCL where the patient presented with pancytopenia and splenomegaly.

CASES

An 82-year-old male patient presented with complaints of giddiness and easy fatigability since 8-10 days. There were no other significant complaints. On examination pallor was present. On abdominal examination there was massive splenomegaly. No peripheral lymphadenopathy was present. A chest X-ray showed no lymphadenopathy. Haematological examination revealed a Haemoglobin concentration of 6.1gm/dl, total leukocyte count (TLC) of $2.1 \times 10^9/L$, red blood cell (RBC) count $3.12 \times 10^{12}/L$ and platelets $61 \times 10^9/L$. The peripheral blood smear demonstrated leucopenia, microcytic, hypochromic red cells and a differential count of 38% segmented neutrophils, 48% lymphocytes, 2% eosinophils, 2% monocytes and 10% atypical lymphoid cells. These atypical cells displayed round to oval bland nuclei with light basophilic cytoplasm containing numerous wispy hairy projections on outer surface [Figure 1]. Repeated bone marrow aspiration demonstrated a dry tap. Bone marrow biopsy revealed a moderately cellular marrow having abnormal aggregates of predominant atypical lymphoid cells with typical fried-egg appearance with abnormal areas of fibrosis [Figure 2]. Histochemistry demonstrated increased network of reticulin fibres. Immunohistochemical stains revealed positivity for CD19, CD20, CD25, CD103 and CD11c [Figure 3] and were negative for CD23, CD10 & CD5.

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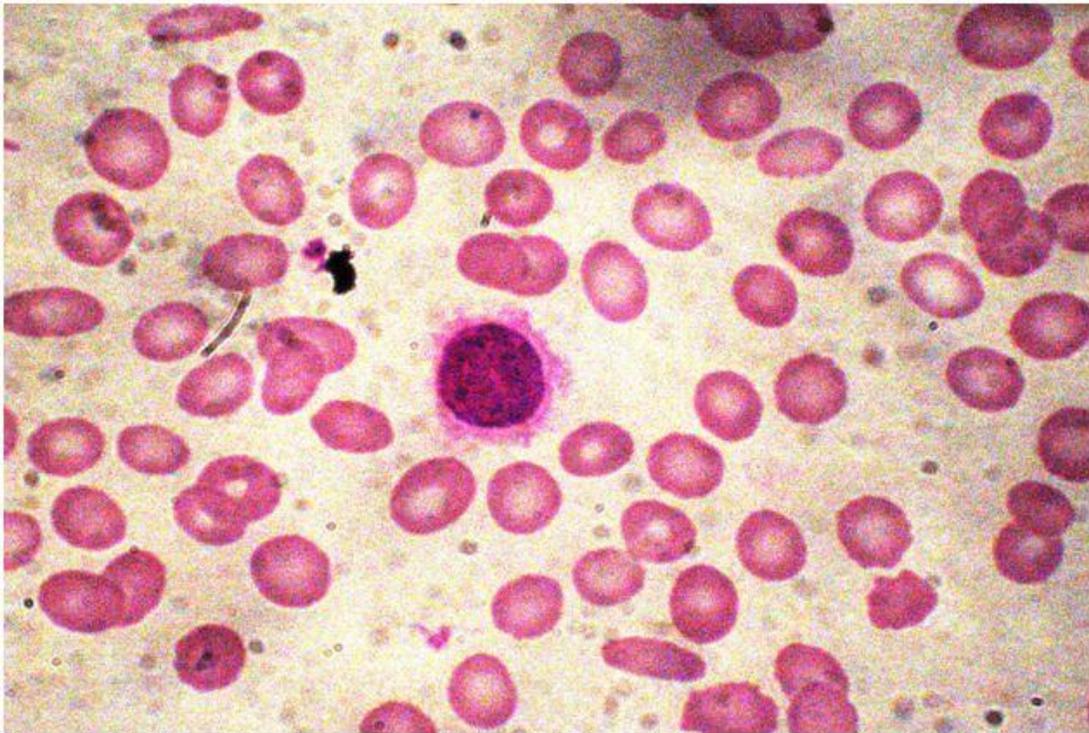


Figure 1: Peripheral blood smears from the patient demonstrating presence of hairy cell. Note the presence of fine, outward cytoplasmic projections. (Leishman stain X1000)

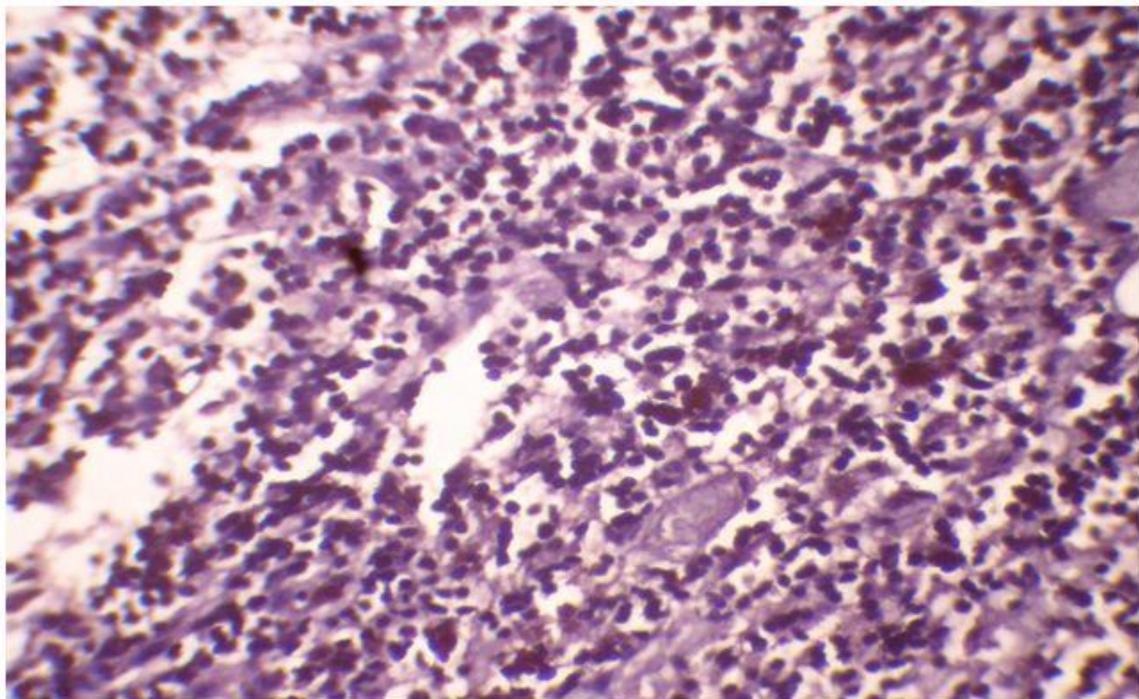


Figure 2: Diffuse replacement of the bone marrow by small cells with clear cytoplasm and round central nucleus showing fried egg-like appearance; (Hematoxylin and Eosin stain, X400)

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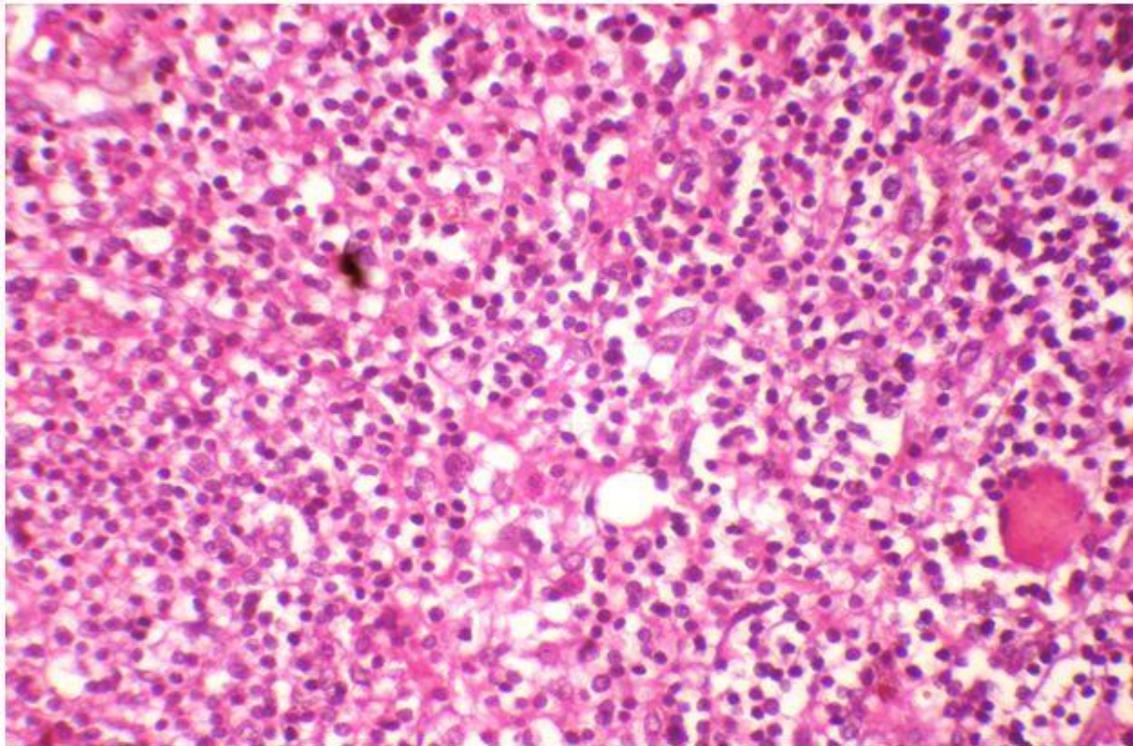


Figure 3: Immunohistochemical stain, the hairy cells show positivity to antibodies against CD20 antigen (X400).

RESULTS AND DISCUSSION

In the present case, the patient presented with leukocytopenia without peripheral lymphadenopathy and prominent splenomegaly. Further investigation like bone marrow aspiration, biopsy and immunohistochemical stain confirmed the diagnosis of HCL. Careful attention to morphologic details is important to recognize HCL from other variant particularly when low percentages of hairy cell are present.

HCL is a rare disorder accounting for 2% of all leukemias. The annual incidence of HCL is estimated to be 0.3 cases per 10,000 (Mey, 2003). It was first described in 1958 at the Ohio State as a distinct clinical pathological and haematological entity (Bouroncle, 1958). The basic mechanism involved in pathogenesis of HCL is poorly understood. Expression profiling of HCL have demonstrated that they are activated late B-cells. Hairy cells do not morphologically resemble any known cells in normal lymphocyte lineage (Nighat, 2011). Anaemia with pancytopenia is a common hematologic abnormality seen in most patients of HCL. It is important to clinically differentiate between HCL and other chronic B-cell lymphoproliferative disorder because of differing treatment protocol and an indolent clinical course. Two disorder that particularly bear histological resemblance to hairy cell leukaemia are Splenic lymphoma with villous lymphocytes (SLVL), and HCL variant (HCL-V) but these typically lack expression of hairy cell surface antigen (Pettit, 1999). HCL-V is an uncommon variant and accounting for approximately 0.4% cases of chronic lymphoid malignancies and 10-20% of HCL cases. HCL-V has hypercellular marrow that can be easily aspirated. The cells are comparatively smaller than classical HCL cell contain abundant basophilic cytoplasm, high N/C ratio, central round occasionally bilobed indented nucleus with prominent nucleolus (Narat) . The cells are positive for B cell markers and also for CD 103, CD 11c but negative for CD 25, and ANNEXIN 1. SLVL is a rare disorder that comprises more than 1% cases of lymphoid neoplasm. It is a malignant counterpart of splenic marginal zone lymphoma. Morphologically

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the SLVL cells are slightly larger than small lymphocytes with condensed chromatin, inconspicuous nucleoli, and moderate amount of cytoplasm and fine villous projections on cell surface. Immunophenotypically these cells are CD22 positive and negative for CD25, CD103 and CD11c (Swords). Other differential diagnosis includes B- chronic lymphocytic leukaemia (CLL), Prolymphocytic leukaemia. The cells differ from hairy cell leukaemia, as they have more coarsely clumped chromatin and round to oval nuclei (Burke, 1981). They strongly express CD 103, CD20 and CD 11c. These cells typically infiltrate bone marrow, spleen and to lesser extent liver and lymph nodes (Jaffe, 2001). Marked fibrosis produces “dry tap”. Such type of fibrosis is rare in other lymphomas and is also differentiated from myelofibrosis as significant fibroblast proliferation is not seen as is seen in myelofibrosis. Recently immunohistochemical stain of Annexin A has been reported to be 100% specific marker for hairy cell leukaemia (Falini, 2004).

HCL has an indolent course and patient who are asymptomatic requires no therapy. Treatment is indicated for symptomatic or cytopenic patients. Indications include significant neutropenia, thrombocytopenia, symptomatic splenomegaly, constitutional symptoms due to HCL or recurrent infections (Matutes, 1997). We have reported this case of HCL which was suspected on morphology, and diagnosis was confirmed by immunohistochemistry. Recognising the features of this leukaemia and considering it in the differential diagnosis in the right clinical setting is crucial to avoid a misdiagnosis of this most successfully treatable leukaemia.

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