Case Report

Leukemia cutis

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ABSTRACT

Patients with leukemia may show involvement of the skin. This skin involvement can be due to infiltration of skin by leukemic cells or it may be a part of nonspecific cutaneous manifestations. Leukemia cutis is the infiltration of neoplastic leucocytes or their precursors into the skin resulting in extensive clinical manifestations. Described mostly in acute myeloid leukemia and acute myelocytic monocytic leukemia, it is rare in chronic myeloid leukemia and is seen mostly during the blast crises. Its presence signals poor prognosis.

Key words: Acute myelomonocytic leukemia, chronic myeloid leukemia, leukemia cutis, Sweet syndrome

INTRODUCTION

Cutaneous involvement in leukemic patients has been described. This skin involvement may be due to infiltration by neoplastic leucocytes or their precursors which is termed as leukemia cutis or it could be as a result of nonspecific inflammatory reactions. Leukemia cutis is seen mostly in association with monocytic, myelomonocytic and T cell leukemias.[1,2] Rarely, it has been described in chronic myelocytic leukemia (CML) and is mostly seen during the blast transformation of the case.[1,2] Skin lesions may also represent the first clinical manifestation of this disease accompanied by normal peripheral smear which is termed as aleukemic leukemia.[3] The commonest clinical presentation could range from erythematous or violaceous macules, papules, plaques or nodules.[3] We report a case of cutaneous involvement in a case of CML who was in the accelerated phase (AP).

CASE REPORT

A 58-year-old male visited the Dermatology Department with erythematous macules and papules on the face, anterior chest [Figure 1a], back [Figure 1b] and legs [Figure 1c]. On being questioned he told that he had been diagnosed with CML 2 years back. His bone marrow aspiration (BMA) done at that time was hypercellular with granulocyte hyperplasia. The myelogram was as follows: Blasts - 7%, metamyelocytes - 26%, myelocytes - 27%, band form and polymorphs - 35%, plasma cells - 2%, lymphocytes - 3%. A diagnosis of CML-chronic phase (CML-CP) was made and a bone marrow biopsy was done which was found to show panmyelosis with grade III fibrosis-chronic myeloproliferative disease. The patient was started on imantinib mesylate and was in remission for almost 2 years. Keeping in mind his history a skin biopsy from the lesional site was recommended and a subsequent peripheral smear and BMA was advised. The biopsy on hemotoxylin and eosin showed a diffuse infiltrate of myeloid cells in the dermis with a perivascular and periadenexal predilection [Figure 2a and b]. On immunohistochemistry (IHC) these atypical cells were positive for myeloperoxidase (MPO) [Figure 3]. The peripheral smear showed normocytic normochromic anaemia with thrombocytopenia. Differential leucocyte count revealed 13% blasts. The BMA was hypercellular. Myeloid:erythroid ratio was 10:1. Erythropoesis was normoblastic. No dyserythropoesis was seen. Myeloid series showed a bulge with giant metamyelocytes. The myelogram was as follows: Erythroblasts - 19%, myelocytes - 20%, metamyelocytes - 9%, band forms - 14%, polymorphonuclear neutrophils - 36%, blasts - 12%. Megakaryocytes were seen. A diagnosis of CML-AP [Figure 4] with extensive infiltration of leukemic cells into the skin was made. Patient was advised to take phototherapy along with systemic chemotherapy.

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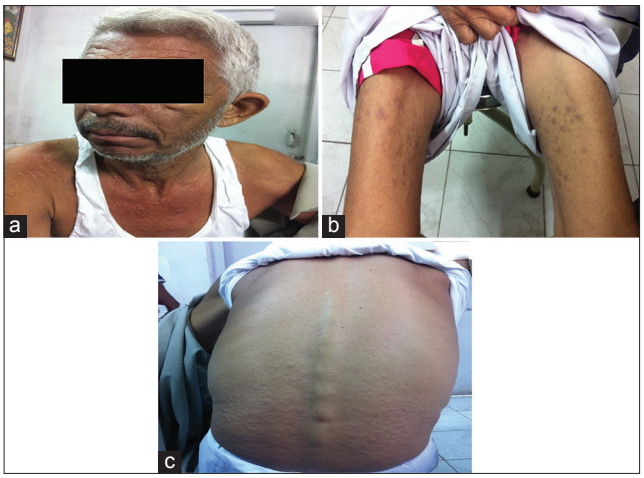

Figure 1: (a) Clinical photograph showing macules and papules on the face and anterior chest wall. (b) Clinical photograph showing macules and papules on the legs. (c) Clinical photograph showing macules and papules on the back

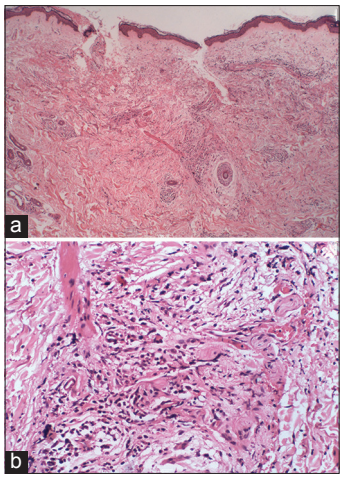

Figure 2: (a) Phtomicrograph showing atypical cells in the dermis. (H and E, ×40). (b) Photomicrograph highlighting the atypical cells in the dermis with a periadenexal and perivascular prediliction. (H and E, ×100)

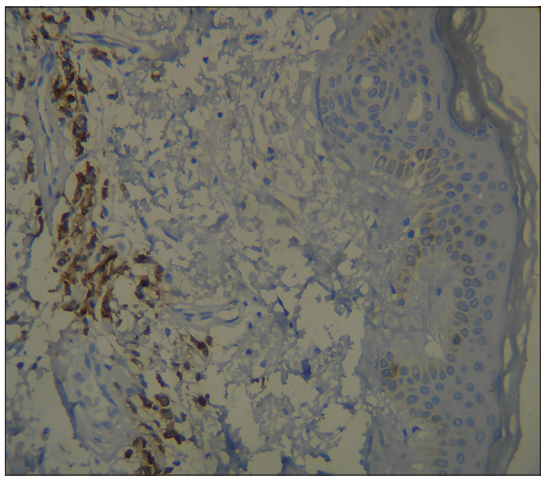

Figure 3: Phtomicrograph showing the atypical lymphoid cells to be positive for myeloperoxidase. (IHC, ×100)

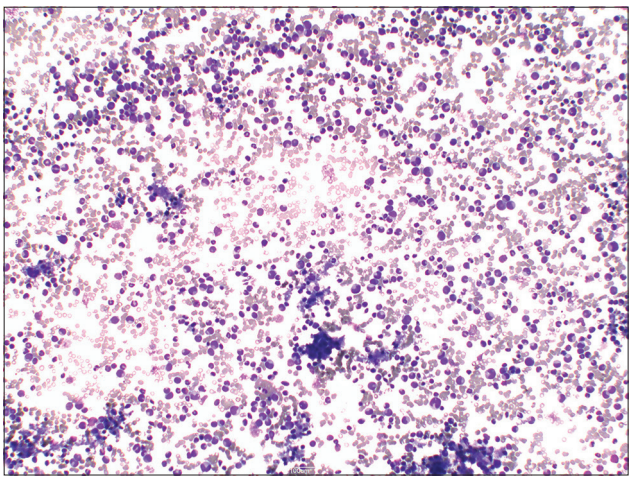

Figure 4: Photomicrograph showing the bone marrow aspirate of the patient (chronic myeloid leukemia - chronic phase). (Giemsa, ×40)

DISCUSSION

Leukemia cutis is the infiltration of the skin by malignant leucocytes and their precursors and signals a grave prognosis. Described mostly in acute monocytic, myelomonocytic and T cell leukemias, very few cases have been reported in CML usually in the blast crises and very rarely in the AP.[1-3] Sometimes skin lesions may be the first clinical manifestation of the disease with a normal peripheral smear. This has been termed as aleukemic leukemia cutis.[1-3] Our patient was a known case of CML-CP on treatment since 2 years. He presented with erythematous macules and papules on the face, chest, back and legs since 2 weeks. Erythematous macules, papules, plaques and nodules is the most common morphological presentation of leukemia cutis with exanthematous eruptions, purpura, bullae, ulcers, leonine facies, stasis dermatitis being the other clinical presentations.[4] The skin biopsies of such patients shows diffuse infiltration of leukemic cells in the dermis with a perivascular and periadenexal predilection. In our patient a similar picture was obtained on histopathological examination with the atypical cells being positive for MPO on IHC.

Leukemia cutis needs to be distinguished from Sweet syndrome which is a cutaneous paraneoplastic manifestation in patients of hematological malignancies which is characterized by the presence of mature neutrophils in the dermis.[5] We ruled out Sweet syndrome as our case showed extensive infiltration of the dermis with immature granulocytes.

The exact nature of leukemia cutis is still a mystery but chemokine receptor (CCR4) and adhesion molecule (cutaneous lymphocyte associated antigen) probably play a role in the cutaneous tropism of neoplastic leucocytes.[2]

Presence of leukemia cutis indicates a poor prognosis with an average survival of 9.4 months.[6-8] Our patient succumbed to the disease after 8 months. We reported this case to draw attention of the dermatologists and pathologists towards this entity as it calls for early diagnosis and treatment and the pathologist is quite instrumental to aid the dermatologist in arriving at the correct diagnosis of this entity.

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