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Case Report

Aleukemic leukemia cutis: a rare case report

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ABSTRACT

Leukemia cutis is a rare case disorder characterised by infiltration of neoplastic leukocytes (myeloid or lymphoid) in clinically identifiable cutaneous lesions. It can occur before the onset of hematological presentation of leukemia or during the disease course. The lesions may be highly variable ranging from flesh coloured to violaceous papules, nodules or plaques. A 52-year-old male patient came to the Dermatology Department with multiple asymptomatic erythematous papulonodular lesions involving the face, trunk as well as the extremities from the past 6 months. Systemic examination and counts were normal. On histopathological examination diffuse dense infiltrate of neutrophils and eosinophils throughout the dermis with extension of infiltrate in the interstitium of reticular dermis. Scattered amidst the infiltrate were several large cells with abundant pale cytoplasm and irregular nuclei. Immunohistochemistry was done, radiotherapy was planned and poor prognosis was explained to the patient. The case is being reported due to its rarity and the role of dermatopathologist in early diagnosis.

Keywords: Leukemia cutis, Neoplastic leukocytes, Irregular nuclei

INTRODUCTION

Leukemia cutis is characterised by infiltration by neoplastic lymphocytes (myeloid or lymphoid) in clinically identifiable cutaneous lesions. It has a highly variable range of cutaneous manifestations, patients usually have a pre-existing systemic leukemia, but occasionally skin involvement occurs before the involvement of the bone marrow or peripheral blood and hence termed aleukemic leukemia cutis.[1] Cutaneous manifestation can be of 2 types: nonspecific leukemid lesions that contain no leukemic cells and the less common leukemia cutis lesions that has leukemic cell infiltrate.[2] The prognosis of leukemia cutis is poor hence early diagnosis with skin biopsy is crucial.[3]

CASE REPORT

A 52-year-old male patient presented to the out-patient department of dermatology with multiple asymptomatic erythematous papulonodular lesions involving the face trunk and extremities from the past 6 months. The papules varied in size from 1×2 cm to 2×2.5 cm. The rest of the systemic examination was normal. There was no history of any bleeding, weight loss or any systemic illness in the past. Hemogram showed Hemoglobin 11.2 gm/dl, total and differential white blood counts were normal. ESR was 27 mm at 1 hr. Liver and renal function tests were normal. HIV ELISA was negative. On histopathological examination there was a diffuse dense infiltrate of neutrophils and eosinophil throughout the dermis with extension of infiltrate in the interstitium of reticular dermis.

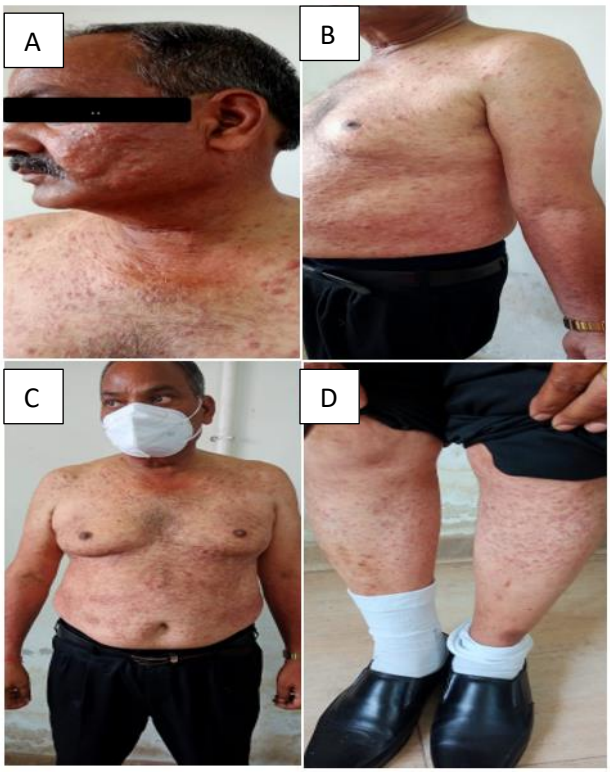

Figure 1: Multiple erythematous papules and nodules present over A) face, B) trunk, C) upper and D) lower limb.

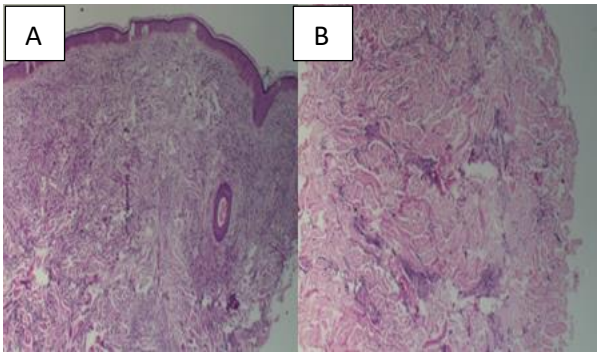

Figure 3: A) Histopathology 10x slide showing dense infiltrate, B) 40X slide showing large cells with pink cytoplasm and irregular nuclei.

Scattered amidst the infiltrate were several large cells with abundant pale pink cytoplasm and irregular nuclei. Immunohistochemistry was done and it was consistent with monocytic differentiation (positive for CD65 and lysozyme). Bone marrow aspiration was performed and revealed a hypercellular bone marrow with trilineage maturation with only 2% blasts. Diagnosis of aleukemic leukemia cutis was made. Patient received total body irradiation with electron beam (total dose 18Gy) after which there was partial regression of the skin nodules. Since complete remission could not be obtained combination chemotherapy with vincristine, cyclophosphamide, doxorubicin and prednisolone is ongoing.

DISCUSSION

Leukemia cutis is characterised by infiltration of the skin by neoplastic leukocytes and their precursors. The migration of leukemic cells into the cutaneous tissue occurs due to skin selective homing of memory T-cells. Hence adhesion molecules , chemokines and integrin may play an important role in the process of cell migration.[1] Common clinical finding include papules, papulonodular, tumour and plaques. Rare findings include erythema, erythroderma, ulceration and blister formation.[2] Atypical findings may mimic leonine facies, leukemic vasculitis, figurate cutaneous lesions and erythema nodosum.[1] The occurrence of skin lesions may develop after the systemic involvement of bone marrow and peripheral blood, may occur concurrently or before systemic involvement. When lesions develop before systemic involvement it is termed as aleukemic leukemia cutis. Leukemia cutis may develop secondary to acute myelogenous leukemia, chronic myelogenous leukemia, acute lymphoblastic leukemia, myelodysplastic syndrome and chronic lymphocytic leukemia.[3] In the above case systemic examination and peripheral blood examination was normal hence it was diagnosed as aleukemic leukemia cutis. The precedence of cutaneous lesions and lack of systemic involvement delayed the diagnosis. Initial diagnosis of a non-specific inflammatory reaction was made but persistence of lesions increased suspicion and biopsy was done. Histologically, a diffuse infiltration of mononuclear cells into the dermis and subcutaneous fatty tissue is evident. The cells have abundant cytoplasm and irregularly shaped nuclei, with fine chromatin and prominent nucleoli. Immunohistochemical markers are required to make definite diagnosis.[4] Differential diagnosis include metastasis of visceral malignancies, Kaposi sarcoma, sarcoidosis, secondary syphilis and systemic lupus erythematosus.[5] Leukemia cutis should be included in the differential of evolving infiltrative cutaneous nodules and plaques and further peripheral blood and bone marrow examination should be done.[6] Both chemotherapy and radiotherapy in the form of whole skin electron beam therapy can used in the form of treatment.[7] In addition to these autologous or allogeneic stem cell transplantation can be done. In this patient radiotherapy was planned. Poor prognosis has been reported in some cases whereas other reports have suggested that it is a heterogenous condition that can result in spontaneous resolution without the need of aggressive chemotherapy.[8] But once the cells start to migrate from the skin to the peripheral blood or bone marrow the mean survival rate is only 3-30 months.[9] Relapse rates are also high. It is important for dermatologists to be aware of this entity so that prompt diagnosis can be made with the help of histopathological examination and immunohistochemical studies so that specific treatment can be started.[10]

CONCLUSION

Above described is a case of aleukemic leukemia cutis, diagnosis was made based on histopathological examination and immunohistochemistry. Poor prognosis was explained to the patient and radiotherapy was planned.

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