

Leukaemia Cutis Presenting as Leonine Facies

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Summary

Acute leukaemia may rarely present as diffuse papules, nodules and plaques forming leonine facies. Leukaemia cutis generally carries a poor prognosis, and responds less well to chemotherapy. We described a case of acute myelomonocytic leukaemia presenting as leonine facies as a result of extensive cutaneous infiltration. The patient did not achieve haematological remission following standard induction chemotherapy and succumbed 6 weeks after the diagnosis was made.

Key Words: Acute myeloid leukaemia, Acute myelomonocytic leukaemia, Leonine facies, Leukaemia cutis

Introduction

Leukaemia is associated with a wide variety of specific and non-specific cutaneous manifestations. The specific lesions of leukaemia cutis result from infiltration of the epidermis, dermis, or subcutaneous fat by neoplastic leucocytes and can have various appearances and presentations.

Case Report

A 60-year-old woman presented with a 2-week history of purplish nodules over her face. The nodules began on her cheeks and continued to enlarged and increased in number. By 3 weeks, the lesions started to coalesced and had involved most of the face and scalp. The skin lesions were not pruritic, painful or tender. There was no history of fever or drug ingestion. Physical

examination revealed violaceous, indurated nodules and plaques over the forehead, cheek, nose and earlobes (Fig. 1). There was thickening of the skin of the face and loss of the eyebrows that resulted in leonine facies. The eyelids and conjunctivae were infiltrated by similar lesions resulting in inability to open the eyes. A nontender erythematous nodule measuring 2cm in diameter was noted on the upper margin of the left nipple. There were no gum hypertrophy or visible oral lesions. No significant enlargement of the liver, spleen or lymph gland was detected.

The haemoglobin was 9.0 g/dl, platelets $39 \times 10^9/l$ and leucocytes $97.1 \times 10^9/l$ with 65% blast cells. Peripheral blood smear showed myeloblasts that represent 65% of the total nucleated cells. Biopsy of the nodules over the forehead and near the left nipple disclosed diffuse infiltration of the dermis by malignant-appearing mononuclear cells

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(Fig.2). The mononuclear cells expressed CD13, CD33, CD11c, CD68, HLA DR, MPO and EMA. Immunophenotypic studies of the skin and bone marrow infiltrates were consistent with acute myelomonocytic leukaemia (FAB-M4). There was no histological evidence of acute febrile neutrophilic dermatosis (Sweet's syndrome), vasculitis or fungal infection.

The patient received induction chemotherapy with cytosine arabinoside and daunorubicin resulting in some resolution of the skin lesions and 30% blast cells in the peripheral blood 2 weeks later (leucocyte count $14.2 \times 10^9/l$). She presented one month after the start of chemotherapy with fever and worsening of the skin lesions. Almost the entire skin was diffusely reddish and thickened, and the face looked like facies leontina (leonine facies). The peripheral blood blast cell count was 85% (leucocyte count $78.8 \times 10^9/l$). The patient developed gram-negative septicaemia associated with profound neutropenia that lead to her death soon after admission.



Fig 1. Photograph of the patient showing the skin lesions over the face that resembled leonine facies

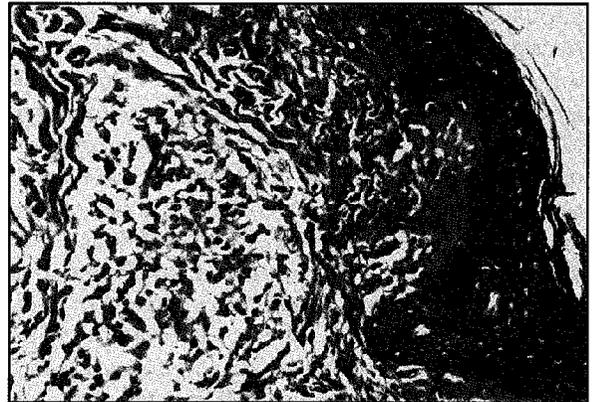


Fig 2. Section of the skin biopsy specimens revealed diffuse infiltration of the dermis by malignant-appearing mononuclear cells (haematoxylin-eosin, orig x20)

Discussion

Cutaneous involvement with acute leukaemia is fairly uncommon and usually a relatively late event. Our patient presented with skin infiltrates without signs of other organ involvement. Skin lesions in acute leukaemia may be specific, called leukaemia cutis, or non-specific, called leukaemids that include leukocytoclastic vasculitis, Sweet's syndrome, urticaria, erythroderma and erythema nodosum. In a study of 42 patients with specific leukaemic infiltrates, Su *et al*¹ found that the most common lesions to be multiple papules and nodules (60%) and infiltrated plaques (26%), with less frequent occurrence of macules, ulcers, ecchymoses, and palpable purpura. Infiltrate of the face, if severe enough, results in facies leontina (leonine facies) - formerly a classic features of advanced promatous leprosy.

Leukaemia cutis occurs in about 20% of patients with acute monocytic leukaemia (FAB-M5)², and is uncommonly seen in other forms of AML. Leukaemia cutis has been known to localise at sites of trauma, burns; intramuscular injections, recent surgery, including Hickman catheter's sites². It typically manifests as red or violaceous

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papules or nodules, mainly on the face, and can become purpuric with coexisting thrombocytopenia, and is generally asymptomatic. Dermal infiltrates can form plaques and arciform lesions that resemble mycosis fungoides, and infiltrates of the face, if severe may result in leonine facies. Skin infiltration favours the lower dermis and subcutaneous fat with prominent involvement of adnexal structures, nerves, vessels of the superficial and deep plexus as noted in the present case. Skin involvement is not related to the circulating white-cell count but the skin can act as a sanctuary for leukaemic cells. Leukaemia cutis generally carries a poor prognosis, with a high incidence of extramedullary disease at other sites including the meninges, and with imminent bone marrow relapses and serial skin relapses. As observed in the present case and a previous report ³, standard induction chemotherapy for AML is less effective in patients who have

cutaneous involvement. Nevertheless, with curative therapy directed at skin, bone marrow, and other sites of extramedullary involvement, long-term disease-free survival is possible ³.

Diffuse nodule and plaque forming leonine facies has been reported recently in a patient with adult T-cell leukaemia ⁴. We herein described a patient with AML who presented with extensive skin nodules and plaques that resembles a leonine facies as the initial clinical manifestation of the disease. Leonine facies has classically been described in patients with lepromatous leprosy and occasionally seen with amyloid or myxedematous skin infiltrate. To the best of our knowledge, this report represents one of the few cases of AML presenting as leonine facies. This case illustrates that leukaemia cutis, albeit uncommon should be included in the differential diagnosis of a leonine facies.

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