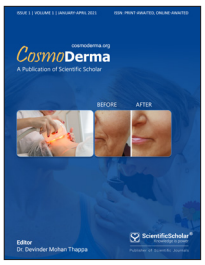
www.cosmoderma.org

ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

CosmoDerma



Case Report

Leukemia cutis: An atypical manifestation of myelodysplastic syndrome

Maruti Mohan Jarag[1], Richa Sharma[1], Rachita S. Dhurat[1]

[1]Department of Dermatology, LTMMC and LTMGH, Mumbai, Maharashtra, India.

***Corresponding author:**
Rachita S. Dhurat,
Professor and Head,
Department of Dermatology,
LTMMC and LTMGH,
Mumbai, Maharashtra, India.

rachitadhurat@yahoo.co.in

Received : 20 March 2023
Accepted : 08 May 2023
Published : 27 May 2023

DOI
10.25259/CSDM_71_2023

Quick Response Code:

ABSTRACT

Leukemia cutis is the infiltration of neoplastic leukocytes or their precursors into the epidermis, dermis, and subcutaneous tissue, resulting in various cutaneous manifestations. A young girl presented with multiple painful hard indurated nodules and plaques over the abdomen, back, breast, and bilateral lower limbs for 4 months. Biopsy showed a dense lymphocytic infiltrate seen with large atypical lymphocytes. Immunohistochemistry (IHC) revealed positivity for CD34, C-KIT, and Glycophorin A. Peripheral smear revealed 32% blast cells. Bone marrow aspirate were suggestive of hyper-cellular marrow with myeloid preponderance. IHC showed myeloperoxidase (MPO) negativity which led to the diagnosis of leukemia cutis secondary to myelodysplastic syndrome with blast showing myelomastocytic differentiation with MPO negative acute leukemia.

Keywords: Leukemia cutis, Immunohistochemistry, Mylodysplastic syndrome, C-KIT mutation, Acute leukemia

INTRODUCTION

Leukemia cutis is defined as the cutaneous infiltration of leukemic cells, manifesting as violaceous red-brown or hemorrhagic papules, nodules, plaques, and patches. Rarely, bullae and ulcers may be seen. The most common malignancy leading to leukemia cutis is acute myeloid leukemia (AML). Leukemia cutis may be the first evidence of underlying hematological malignancy. In approximately 30–40% of cases of myelodysplastic syndrome (MDS), patients progress to acute leukemia, defined by the presence of at least 20% blasts in the bone marrow. Cutaneous infiltration is often the first marker of this progression. Their early recognition and accurate diagnosis can play an important role in the subsequent clinical management of patients with MDS.[1] The frequency of leukemia cutis in MDS is unclear. However, it is believed to be a rare phenomenon linked with poor prognosis and rapid disease progression.

A diagnosis of leukemia cutis generally portends a poor prognosis and strongly correlates with additional sites of extramedullary involvement. The dermatologist is often instrumental in the diagnosis of leukemia cutis. Accurate diagnosis has tremendous prognostic significance and may establish a diagnosis in cases in which leukemia cutis is the harbinger of a systemic leukemic process. This can alter the appropriate treatment regimen for a patient.[2] We report the case of a 16-year-old female patient who presented with multiple painful nodules and plaques over the abdomen, breast, and back, subsequently diagnosed as MDS.

CASE REPORT

A 16-year-old girl presented with multiple painful hard indurated nodules and plaques over the abdomen, back, breast, and bilateral lower limbs for 4 months [Figure 1]. There was no history of fever, night sweats, and weight loss. The patient had no known comorbidities. General examination showed pallor with no peripheral lymphadenopathy. Complete blood count showed gross derangement with hemoglobin 4.7 g/dL, RBC count 1.18 × 1012/L, and platelet count 9000/μL. Peripheral smear showed 32% blast cells. Biopsy from hard indurated nodule showed normal epidermis and extensive infiltrate extending from papillary dermis to reticular dermis [Figure 2]. On higher magnification dense, lymphocytic infiltrate was seen in dermis [Figure 3]. Large atypical lymphocytes having a hyperchromatic nucleus and high nucleus to cytoplasmic ratio was seen through the dermis [Figure 4]. Based on these findings, differential diagnoses kept were leukemia cutis and adult Langerhans cell histiocytosis. To differentiate between these two conditions, immunohistochemistry (IHC) markers were done of which CD1a, S100, and CD207 were found to be negative thus ruling out adult Langerhans cell histiocytosis. Few CD34-positive cells were seen which were negative for tdt. C-KIT (CD117) positivity was seen suggesting an increase in myeloid precursors. Glycophorin A (CD235a) positivity indicated erythroid series of cells suggestive of leukemia of myeloid series. Bone marrow aspirate showed hyper-cellular marrow with myeloid preponderance and shift to left along with megaloblastocytosis and dysgranulopoeisis. IHC showed myeloperoxidase (MPO) negativity. Cytogenetics was done which was negative for BCR-ABL gene t (9:22) and t (8:21). Karyotyping showed monosomy of chromosome 7. Hence a final diagnosis of leukemia cutis secondary to MDS with blast showing myelomastocytic differentiation with MPO-negative acute leukemia was made. 18 fluorodeoxyglucose positron emission tomography scan revealed the involvement of supra and infra diaphragmatic lymph nodes, skin, bone marrow, and spleen. Upper gastrointestinal endoscopy showed duodenal nodularity. The patient underwent the first cycle of chemotherapy with cytarabine and daunorubicin with reasonable improvement [Figure 5]. She was further planned for bone marrow transplantation, unfortunately patient succumbed after the first cycle of chemotherapy.

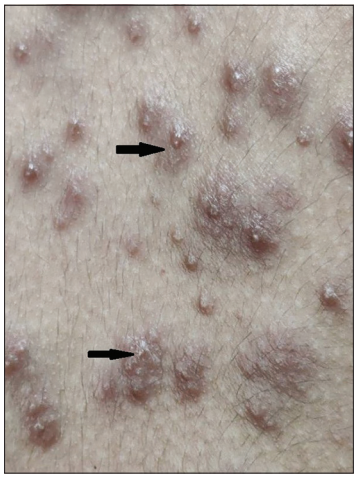


Figure 1: Multiple painful firm to hard indurated hyperpigmented nodules and plaques over abdomen (Black arrow).

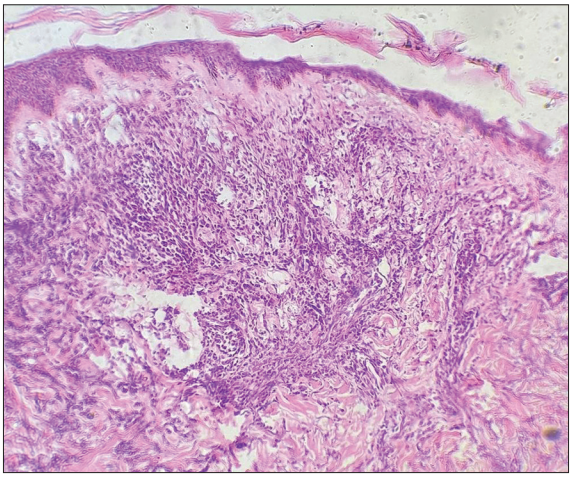


Figure 2: Epidermis appears normal. Dermis shows extensive infiltrate extending from papillary to reticular dermis (H and E ×100).

DISCUSSION

Leukemia cutis is a nonspecific term used for varied cutaneous manifestations of leukemia, often difficult to diagnose clinically. Patients with leukemia cutis usually have concomitant leukemia but occasionally skin involvement occurs earlier than bone marrow and peripheral blood. Thus biopsy can be the first indication of the presence of leukemia. IHC of biopsy specimens helps to establish the diagnosis.

Cutaneous involvement is generally indicative of advanced disease and warrants the investigation of other body sites for extramedullary involvement. Cutaneous manifestations of leukemia can be specific (infiltrates of leukemic cells, or leukemia cutis) or non-specific (inflammatory, paraneoplastic, or secondary to marrow failure).[3]

The clinical appearance of leukemia cutis is variable with erythematous to violaceous papules or nodules being the most common lesions (60%), followed by infiltrated plaques, generalized cutaneous eruption, and erythroderma. They are usually asymptomatic. The

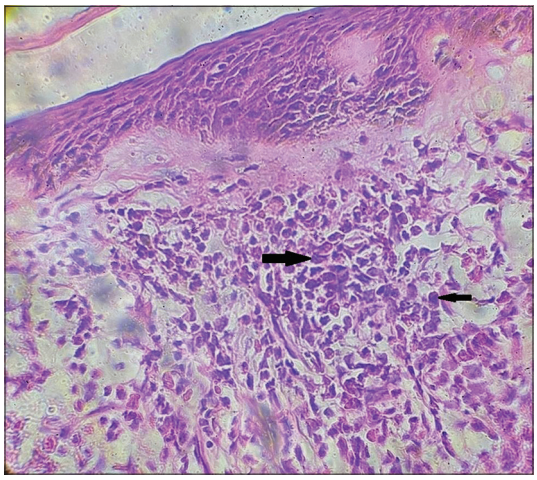


Figure 3: Dermis showing dense atypical lymphocytic infiltrate (black arrow) (H and E ×400).

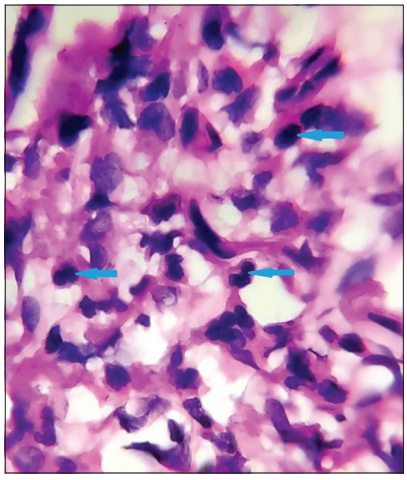


Figure 4: Dermis showing hyperchromatic nucleus and high nucleus to cytoplasmic ratio (Blue arrow) (H and E ×1000).

nodules are typically firm or rubbery in consistency and can become purpuric if the patient is thrombocytopenic. There are also reports of leukemia cutis localizing to sites of herpetic lesions, trauma, intravenous catheters, and recent surgeries. Non-specific cutaneous signs of leukemia are much more common than leukemia cutis, occurring in up to 30–40% of patients with leukemia. The common nonspecific signs are hemorrhagic, including petechiae, purpura, and ecchymosis.

MDS constitutes a spectrum of disorders of ineffective hematopoiesis whose classification by the World Health Organization takes into consideration the presence of unilineage or multilineage dysplasia, an excess of marrow myeloblasts, and specific clonal chromosomal abnormalities.

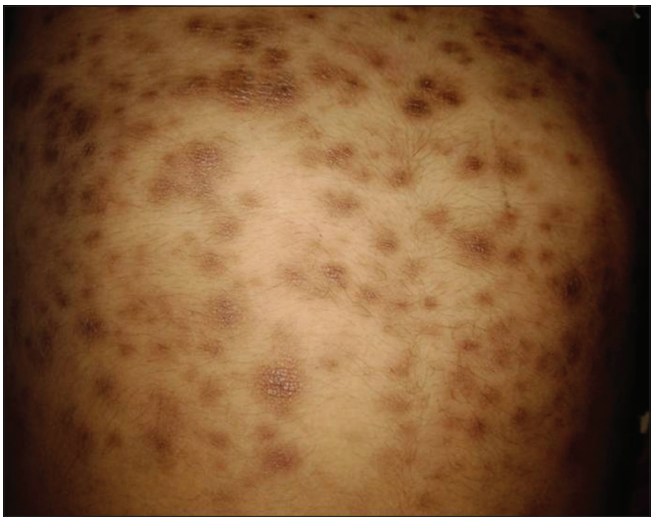


Figure 5: Post one cycle of chemotherapy.

MDS is not a subtype of leukemia; yet, it could also progress into AML or present with cutaneous involvement. The French American British (FAB) classification for MDS is mainly based on the percentage of myeloblast in the bone marrow. The manifestations of leukemia cutis in MDS patients could imply concurrent or approaching transformation to AML.[4] The prognosis is poor, with many patients having other extramedullary diseases and poor survival rates. There are certain hematological and molecular findings associated with the progression of MDS. Poor prognostic indicators include older patient age, thrombocytopenia, refractory anemia, and increased bone marrow blasts. The tendency to progress to leukemia is greater in patients with MDS who have required blood transfusions in the past, have chromosome 7 abnormalities as seen in our patient, or have more than three total genetic abnormalities. CD34-positive blasts in the peripheral blood indicate poor prognosis of MDS. Further, the evolution to AML has been noted to occur more frequently in MDS with basophilia or eosinophilia in the bone marrow. Most patients die within months of diagnosis. Dermatologist plays an important role in early detection and adequate management.

CONCLUSION

A leukemia cutis can present as papule, macule, nodules and plaques. Leukemia cutis is often first indicator of underlying leukemia. Histopathology and immunohistochemistry helps in diagnosis. Early diagnosis and treatment of leukemia cutis leads to favourable prognosis.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Patel LM, Maghari A, Schwartz RA, Kapila R, Morgan AJ, Lambert WC. Myeloid leukemia cutis in the setting of myelodysplastic syndrome: A crucial dermatological diagnosis. Int J Dermatol 2012;51:383-8.
2. Rao AG, Danturty I. Leukemia cutis. Indian J Dermatol 2012;57:504.
3. Martínez-Leboráns L, Victoria-Martínez AM, Torregrosa-Calatayud JL, de Miquel VA. Leukemia cutis: A report of 17 cases and a review of the literature. Actas Dermosifiliogr 2016;107:e65-9.
4. Chang YW, Lee CH, Tseng HC. Leukemia cutis in a medical center in southern Taiwan: A retrospective study of 42 patients. J Formos Med Assoc 2021;120:226-33.

How to cite this article: Jarag MM, Sharma R, Dhurat RS. Leukemia cutis: An atypical manifestation of myelodysplastic syndrome. CosmoDerma 2023;3:81.