Erythrodermic Leukemia Cutis in Patient with Myelodysplastic Syndrome with Multilineage Dysplasia

Filippo Viviani[1,2], Miriam Anna Carpanese[1,2], Martina Lambertini[1,2], Cosimo Misciali[1,2], Elena Sabattini[3], Alessandro Pileri[1,2], Emi Dika[1,2]

From the [1]Division of Dermatology, IRCCS Azienda Ospedaliero-Universitaria di Bologna, [2]Division of Dermatology, Department of Experimental, Diagnostic and Specialty Medicine, University of Bologna, [3]Division of Hematopathology, IRCCS Azienda Ospedaliero-Universitaria di Bologna, Bologna, Italy
E-mail: emi.dika3@unibo.it

Indian J Dermatol 2022:67(5):627

Sir,

A 59-year-old man was evaluated for the onset of an asymptomatic diffuse erythroderma with multiple disseminated red/purple papules and patches also involving the face with a localized, violaceous, and firm edema [Figure 1]. Mucous membranes were spared.

Medical history included hypertension and a myelodysplastic syndrome with multilineage dysplasia (MDS-MLD). One month before, a progression to T-cell acute lymphoblastic leukemia (T-ALL) was diagnosed at the bone marrow biopsy. Laboratory examinations showed leukocytosis (34.4×10^9/L) with marked lymphocytosis (91%; absolute count: 31.4×10^9/L). The patient was in treatment with amlodipine, bisoprolol, and prednisolone (25 mg/day), while allopurinol had been suspended on suspicion of a drug reaction. A recent total-body computed tomography showed the neoplastic involvement of multiple lymph node stations, both supra- and infra-diaphragmatic.

A punch biopsy was performed and histology revealed a superficial dermal perivascular lymphocytic infiltrate with small/medium-sized cells and irregular nuclei [Figure 2]. Immunohistochemistry highlighted a phenotype similar to the one found in the peripheral blood [cluster of differentiation 3 (CD3+), CD2+, CD5+, CD7+, CD56–, CD52+, CD4–, and CD8–] but slightly different from the one observed in the bone marrow biopsy (CD4+, CD8–) [Figure 3]. The diagnosis of a cutaneous localization of a T-ALL [leukemia cutis (LC)] was made, likely determined by a more aggressive sub-clonal cell line. Subsequently, prednisone was increased to 1 mg/kg/day and the screening tests for the start of chemotherapy were prescribed, but the patient died a few weeks later.

The cutaneous signs determined by the infiltration of neoplastic leukocytes are described as LC, which is different from leukemids, and defined as nonspecific skin manifestations in patients affected by a hematological disorder without cutaneous localization. LC is rare and its frequency depends on the underlying hematological condition: the most common types are chronic lymphoblastic leukemia and acute myeloid leukemia, monocytic or myelomonocytic.[1] LC frequency appears to be higher in other subtypes of lymphoblastic leukemia: 20–70% of patients affected by T-ALL can present LC and 25% in case of prolymphocytic leukemia.[2] LC shows no preferred sites of involvement but less often appears on palmoplantar and mucosal surfaces, where the most common manifestation is gingival hyperplasia. The cutaneous presentation is polymorphous, but nodular structures are the most frequent: papules, patches, nodules, and larger tumors with sometimes erosions, ulcerations, or purpuric lesions.[3] Skin signs can be both solitary and multiple, while an erythrodermic

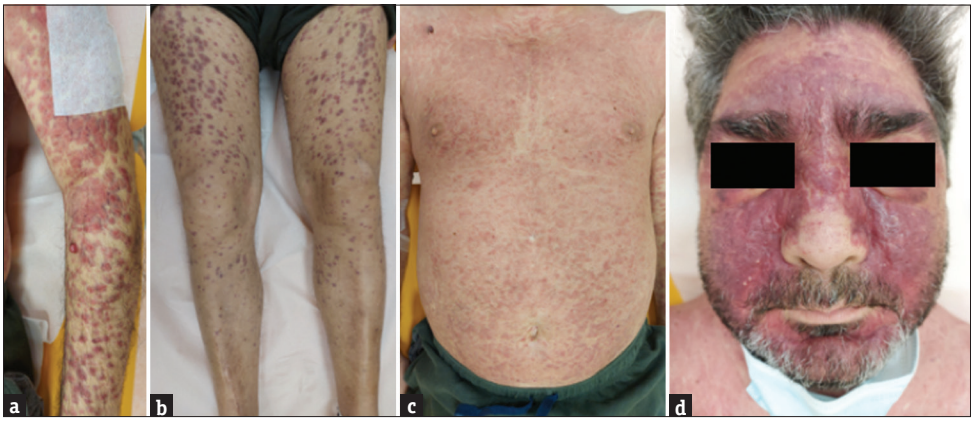

Figure 1: Clinical presentation of the leukemia cutis showing diffuse erythroderma with multiple disseminated red/purple papules and patches involving the limbs (a and b), the trunk (c), and the face with a localized, violaceous, and firm edema (d)

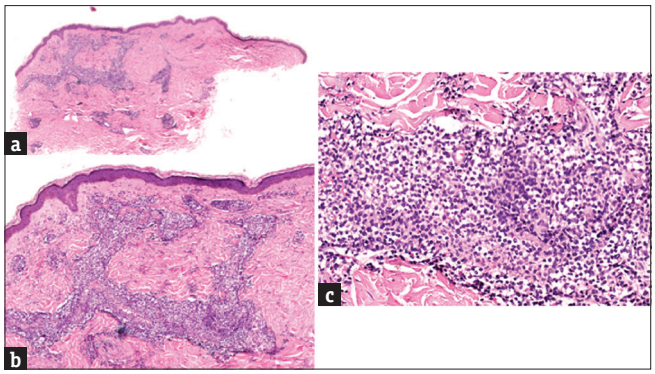

Figure 2: Histology showing a superficial dermal perivascular lymphocytic infiltrate with small/medium-sized cells and irregular nuclei [hematoxylin and eosin, 4× (a), 10× (b), 20× (c)]

presentation may be suspicious of an acute form of leukemia.[4]

The diagnosis is based upon medical history, clinical presentation, laboratory examinations, and the histopathological report, which usually shows a superficial, diffused, banded dermal infiltrate with epidermotropism or a nodular dermal infiltrate. In the case of patches/plaques, a perivascular infiltrate of small to medium-sized atypical lymphocytes may also be present, while immunohistochemistry is often similar to the one observed in our subject.[3] Patients with MDS-MLD and LC should be treated more aggressively because skin manifestation may highlight a progression to acute leukemia. Aggressive systemic chemotherapy, skin-directed radiotherapy, and allogeneic bone marrow transplantation should be considered, although the feasibility is limited by advanced age, functional status, comorbidities, and donor availability.[5] However, the prognosis remains poor with a mortality rate of 80% at 1 year after diagnosis.[2] Regarding our case, the clinical presentation had suggested as an alternative diagnosis a form of drug reaction with eosinophilia and systemic symptoms, while diffuse Kaposi's sarcoma and mycosis fungoides were excluded on an anamnestic basis. The above-described patient represents an exceptionally rare case of erythrodermic/purpuric LC, with only a few cases reported in the literature.[6-8]

A prompt clinical suspect is key to guide the diagnosis and to offer the best possible treatment in patients like the one we described, whose rare condition (MDS-MLD-derived T-ALL and LC) still presents a bad prognosis.

Declaration of patient consent

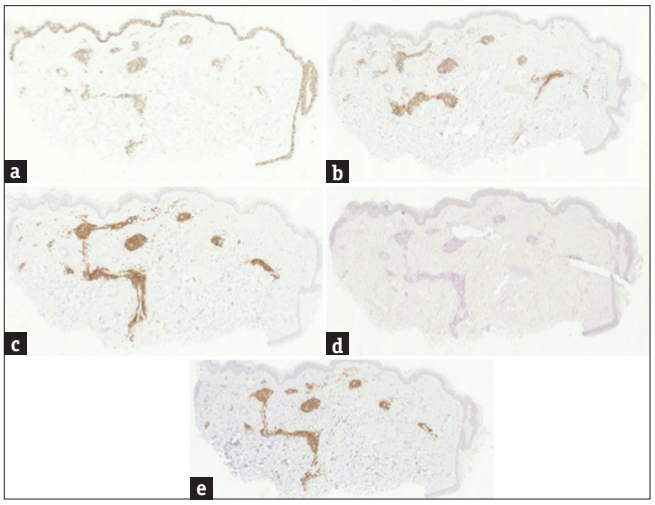

Figure 3: Immunohistochemistry showing positivity for GATA3 (a), CD7 (b), CD3 (c), ITI RED (d), CD5 (e)

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Parsi M, Go MS, Ahmed A. Leukemia Cutis. In: StatPearls. Treasure Island (FL). StatPearls Publishing; 2021. Available from: http://www.ncbi.nlm.nih.gov/books/NBK541136/. [Last accessed on 2021 Apr 21].
2. Martínez-Leboráns L, Victoria-Martínez AM, Torregrosa-Calatayud JL, Alegre de Miquel V. Leukemia cutis: A report of 17 cases and a review of the literature. Actas Dermo-Sifiliográficas Engl Ed 2016;107:e65-9.
3. Miyashiro D, Sanches JA. Cutaneous manifestations of adult T-cell leukemia/lymphoma. Semin Diagn Pathol 2020;37:81-91.
4. Wagner G, Fenchel K, Back W, Schulz A, Sachse MM. Leukemia cutis – epidemiology, clinical presentation, and differential diagnoses. JDDG J Dtsch Dermatol Ges 2012;10:27-36.
5. Patel LM, Maghari A, Schwartz RA, Kapila R, Morgan AJ, Lambert WC, *et al*. Myeloid leukemia cutis in the setting of myelodysplastic syndrome: A crucial dermatological diagnosis:

Myeloid leukemia cutis in the setting of myelodysplastic syndrome. Int J Dermatol 2012;51:383-8.
6. Novoa RA, Wanat KA, Rosenbach M, Frey N, Frank DM, Elenitsas R. Erythrodermic leukemia cutis in a patient with Pre-B-cell acute lymphoblastic leukemia. Am J Dermatopathol 2015;37:650-2.
7. Raj A, Rai R, Rangarajan B. Exfoliative dermatitis with leukemia cutis in a patient with chronic myeloid leukemia: A rare association. Indian J Dermatol Venereol Leprol 2011;77:208-10.
8. Jeong K-H, Lew B-L, Sim W-Y. Generalized leukaemia cutis from a small cell variant of T-cell prolymphocytic leukaemia presenting with exfoliative dermatitis. Acta Derm Venereol 2009;89:509-12.

Access this article online

Quick Response Code:

Website: www.e-ijd.org

DOI: 10.4103/ijd.ijd_240_22

How to cite this article: Viviani F, Carpanese MA, Lambertini M, Misciali C, Sabattini E, Pileri A, *et al.* Erythrodermic leukemia cutis in patient with myelodysplastic syndrome with multilineage dysplasia. Indian J Dermatol 2022;67:627.

Received: March, 2022. **Accepted:** July, 2022.