Dermatology Practical & Conceptual

A Single Skin Lesion as Expression of Widespread Disease: A Case of Systemic Anaplastic Large Cell Lymphoma With a Cutaneous Onset

Stefano Caccavale[1], Vittorio Tancredi[1], Andrea Ronchi[2], Francesca Pagliuca[2], Sebastiano Pellerone[1], Paola Vitiello[1]

1 Dermatology Unit, Department of Mental and Physical Health and Preventive Medicine, University of Campania Luigi Vanvitelli, Naples, Italy

2 Pathology Unit, Department of Mental and Physical Health and Preventive Medicine, University of Campania Luigi Vanvitelli, Naples, Italy

Key words: systemic anaplastic large cell lymphoma, ALK, skin, dermoscopy, early diagnosis

Citation: Caccavale S, Tancredi V, Ronchi A, Pagliuca F, Pellerone S, Vitiello P. A Single Skin Lesion as Expression of Widespread Disease: A Case of Systemic Anaplastic Large Cell Lymphoma with a Cutaneous Onset. *Dermatol Pract Concept*. 2023;13(3):e2023142. DOI: https://doi.org/10.5826/dpc.1303a142

Accepted: February 3, 2023; **Published:** July 2023

Funding: None.

Competing interests: None.

Authorship: All authors have contributed significantly to this publication.

Corresponding author: Vittorio Tancredi, MD, Dermatology Unit, Department of Mental and Physical Health and Preventive Medicine, University of Campania Luigi Vanvitelli, Via Sergio Pansini, 5, 80131 Naples, Italy; phone: +39 3313919797; fax: +39 081 987252; E-mail: tancredivittorio@ymail.com

Introduction

Anaplastic large cell lymphomas (ALCL) are a subgroup of non-Hodgkin T-cell lymphomas which includes anaplastic lymphoma kinase (ALK)-positive ALCL, ALK-negative ALCL and primary cutaneous ALCL.

Case Presentation

A 44-year-old woman was referred to our Department for a large, rapid-growing, eroded, erythematous nodule of her right arm (Figure 1A).The lesion had developed about one month before, diagnosed as a suppurate sebaceous cyst by her general practitioner and treated with topical antibiotics and ichthyol for several weeks. Dermoscopic differential diagnosis included amelanotic melanoma, basal cell carcinoma, sarcoidosis and B cell lymphomas (Figure 1B).

A complete excisional biopsy was performed. Histopathological examination revealed a diffuse lymphoid population and immunohistochemistry was positive for CD4, CD7, ALK, CD30, partially positive for CD3, and negative for CD2, CD5, CD8, granzyme, CD20, CD15, CD8 (Figure 2, A-D). A diagnosis of ALCL was made. Moreover, histologic features, and in particular ALK expression, were more deponent for a cutaneous manifestation of a systemic lymphoma.

Meanwhile, the patient began to complain about systemic symptoms, such as night sweating and weight loss.

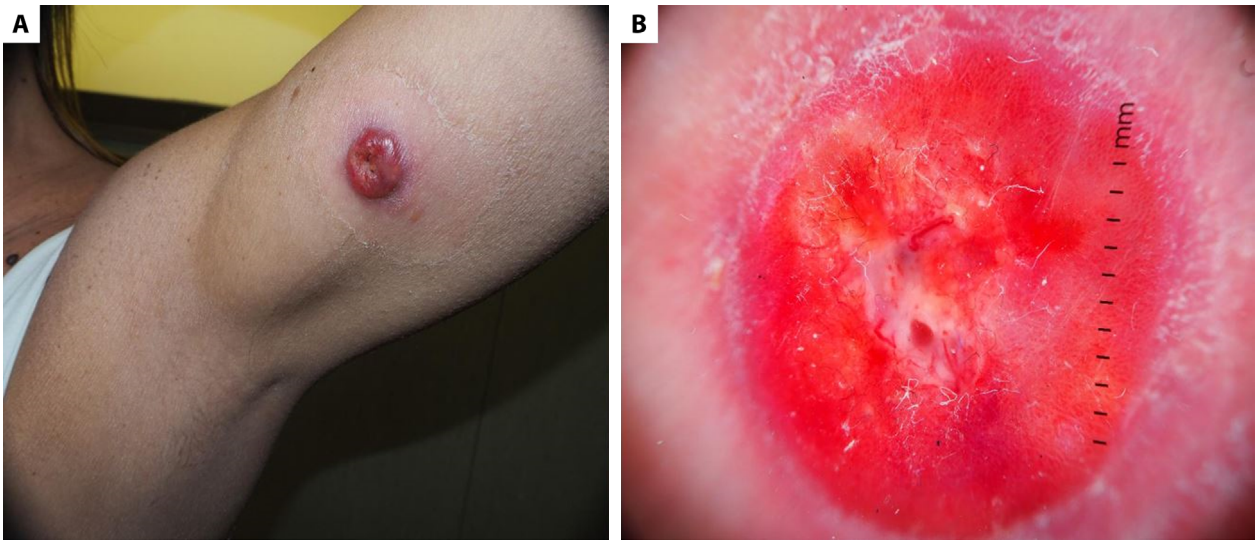

Figure 1. (A) Dermatologic examination revealed a large red nodule, with eroded surface of the right arm. (B) Dermoscopy was characterized by a diffuse structureless red zone with some orange and white areas. Vessels were polymorphic in shape, sizes with a random distribution: some were linear, others were curved and coiled.

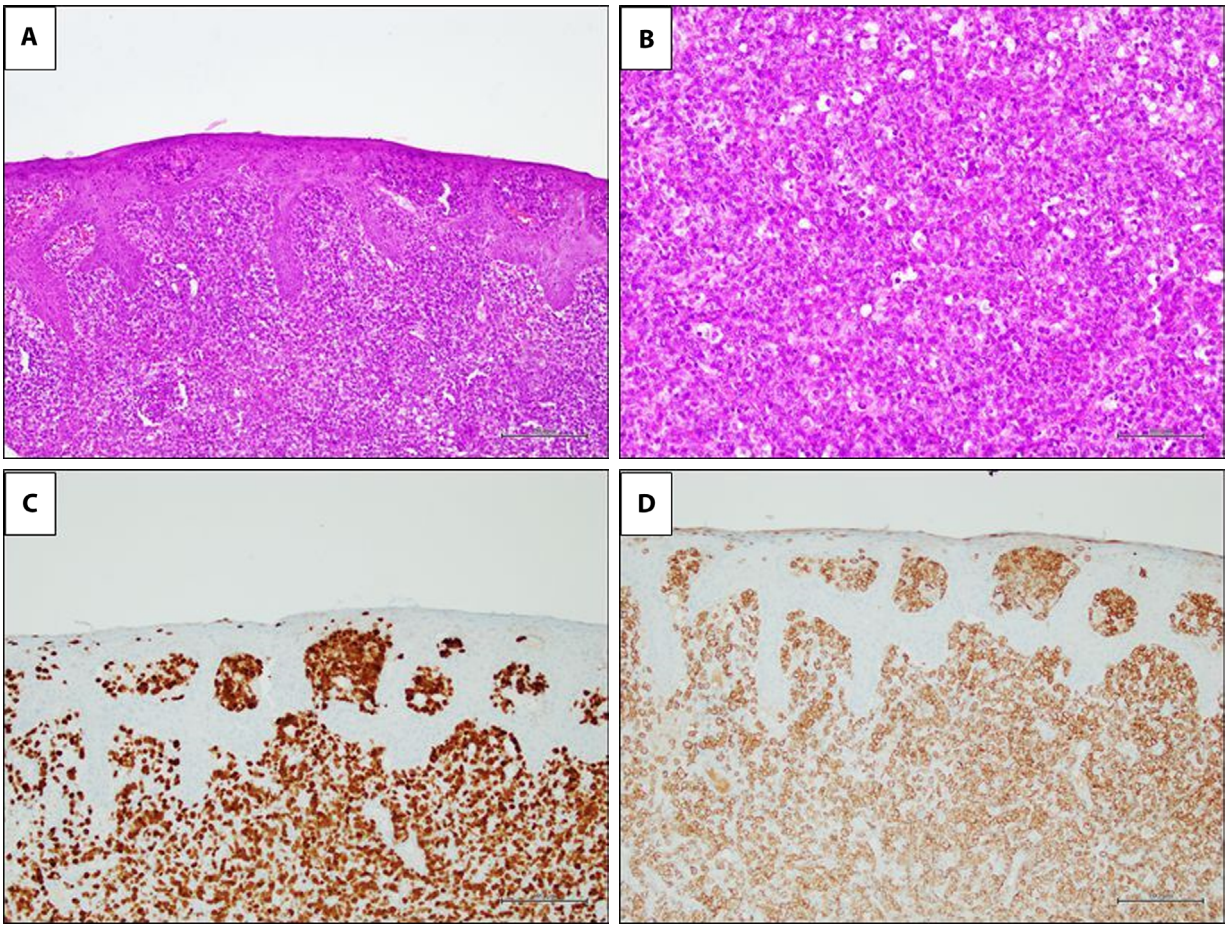

Figure 2. Histological findings. (A) Microscopic examination showing a diffuse lymphoid population filling the dermis (H&E, original magnification 40x). (B) The lymphoid population was constituted by large and atypical cells, with small mature lymphocytes in the background (H&E, original magnification 200x). (C,D) Immunohistochemically, the large neoplastic cells resulted positive for ALK (C) (immunohistochemical stain, original magnification 100x) and (D) CD30 (immunohistochemical stain, original magnification 100x).

Besides, the following cutaneous examination revealed another skin nodule on her right hip, and a lymphadenopathy in her right groin.

CT (computed tomography) scan revealed diffuse large lymphadenopathies in the left axilla and in the right groin. Positron emission tomography (PET) showed a remarkably increased intake of 18–fluorodeoxyglucose in both sites with a maximum standardized uptake value (SUV) of 22.6. Ultimately, PET/CT scan confirmed that cutaneous nodules were secondarisms, thus metastases of a systemic lymphoma. So, according to the TNM system, the disease was a stage IV lymphoma. The patient was hospitalized and admitted to the onco-hematology ward for the management and treatments.

Conclusions

ALCL is a subtype of non-Hodgkin T-cell lymphoma, accounting about 2% of total cases [1]. Among clinical manifestations, diffuse lymphadenopathies are frequently at diagnosis. Systemic symptoms are not uncommon; rarely, some patients have a leukemic presentation [2,3]. Extra nodal disease is possible, and skin is among the most frequently-involved tissues [3]. It's very important for lymphomatous skin localizations to establish if cutaneous lesions have developed in the context of a systemic ALCL or are themselves a primary cutaneous ACLC [2]. Histology is characterized by many possible patterns, but detection of the so-called hallmark cells is constant [1]. These are large CD30 positive tumor cells with bizarre kidney shaped nuclei [1,2]. ALK gene translocation is a feature of this lymphoma and its finding is important to differentiate systemic forms, which are generally positive, from primary cutaneous ALCL, usually ALK negative [3]. When ALK is negative, differentiating primary cutaneous ALCL from systemic ALK negative ALCL is more difficult. Even when integrative exams are negative, often only the course of the disease allows to distinguish the two entities.

Skin involvement is possible during systemic ACLC, but primary cutaneous onset without other symptoms is unusual. A complete physical examination should always be conducted to evaluate other signs of a systemic disease. Our case is a striking example of how even a single skin lesion might be an expression of a systemic and widespread oncological disease.

References

1. Tsuyama N, Sakamoto K, Sakata S, Dobashi A, Takeuchi K. Anaplastic large cell lymphoma: pathology, genetics, and clinical aspects. *J Clin Exp Hematop.* 2017;57(3):120-142. DOI: 10.3960/jslrt.17023. PMID: 29279550. PMCID: PMC6144189.
2. Bennani-Baiti N, Ansell S, Feldman AL. Adult systemic anaplastic large-cell lymphoma: recommendations for diagnosis and management. *Expert Rev Hematol.* 2016;9(2):137-150. DOI: 10.1586/17474086.2016.1122514. PMID: 26581318. PMCID: PMC4876810.
3. Xing X, Feldman AL. Anaplastic large cell lymphomas: ALK positive, ALK negative, and primary cutaneous. *Adv Anat Pathol.* 2015;22(1):29-49. DOI: 10.1097/PAP.0000000000000047. PMID: 25461779.