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A Case Report: CD8 + Aggressive Cutaneous Epidermotropic T Cell Lymphoma (ACLAE), A Rare and Ill-Defined Subtype

Imane Boujguenna[1]; Mohamed Amine Haouane[1]; Fatimazahra Marhoume[2]; Imane Bahbouhi[3]; Kenza Kandri Rody[3]; Ouafa hocar[3]; Said Amal[3]; Hanane rais[1]; Anass Belbachir[1,2]

[1]*Department of Pathological Anatomy FMPM-UCAM-CHU Mohammed VI Marrakech-Morocco.*

[2]*Regenerative Medicine Center FMPM-UCAM-CHU Mohammed VI Marrakech-Morocco.*

[3]*Department of Dermatology FMPM-UCAM-CHU Mohammed VI Marrakech-Morocco.*

***Corresponding Author(s): Imane Boujguenna**

University Cadi Ayyad Faculty of Medicine and Pharmacy of Marrakech, Morocco.

Email: imane.boujguenna1992@gmail.com

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Abstract

Background: Aggressive" CD8+ cutaneous epidermotropic T-lymphoma is a rare entity belonging to the group of cutaneous lymphomas with a cytotoxic T/NK phenotype and which is characterized by: Rapidly evolving tumor lesions at the onset, keratinocyte necrosis, a cytotoxic T immunophenotype and an unfavorable prognosis.

Case presentation: In this study, we report the case of a A 36-year-old patient with no notable pathological history, presenting a rash made up of plaques evolving for 6 months, associated with scalp lesion and alopecia.

Conclusion: LCEA CD8 + is a lymphoma with a poor prognosis, despite the use of aggressive chemotherapy drugs. The use of gemcitabine, pralatrexate, brentuximab, as well as allogeneic stem cell transplantation seems to open up new therapeutic avenues.

Introduction

CD8 + aggressive cutaneous epidermotropic T lymphoma is a rare entity that represents less than 1% of all T lymphomas, which explains the lack of well-defined diagnostic criteria. It is characterized by its aggressiveness and its often fatal course even under treatment [1,2].

The aim of this study is to focus on the diagnosis difficulty of epidermotropic cytotoxic cutaneous T lymphoma, to insist on the contribution of biopsies from the various affected sites and to demonstrate the advantage of the correlation between clinical, flow cytometry and histology in earlier ACLAE diagnosis to initiate appropriate chemotherapy.

Case Presentation

A 36-year-old patient with no notable pathological history, presenting a rash made up of plaques evolving for 6 months **(Figure 1)**, associated with scalp lesion and alopecia evolving for 3 months **(Figure 2)**.

The clinical examination aimed a quasi-erythroderma, dry, site of infiltrated hyperkeratotic plaques, a palmoplantar keratoderma, with an ulcerative, bleeding scalp tumor of 4cm, with alopecia.

Examination of the lymph node areas found multiple lymphadenopathy in the cervical, axillary and inguinal areas.

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The quantification of the blood lymphocyte subpopulations by flow cytometry has revealed a majority of CD45+/CD3+ T-type lymphoid population with an expression of CD8+ (cytotoxic T lymphocyte) higher than CD4+ (T helpers) **(Figure 3)**.

Histological examination of the skin biopsy taken from the scalp was in favor of a dense lymphoid cell infiltrate of perivascular arrangement with epidermotropism ulcerating the epidermis almost entirely **(Figure 4)** and intense CD8 +, CD3 + expression **(Figures, 5 and 6)**, moderate expression of granzyme B and TIA-1, minimal expression of CD4, profile compatible with a CD8 + LCAE. The other biopsies taken from the forearms and hands were in favor of palmoplantar keratoderma.

Thoraco-abdominal-pelvic CT revealed multiple axillary lymphadenopathy as well as subpleural micronodules.

The patient was put on CHOP protocol multidrug therapy (cyclophosphamide, doxorubicin, vincristine, prednisolone), and has already received 2 cures. She is still being treated.

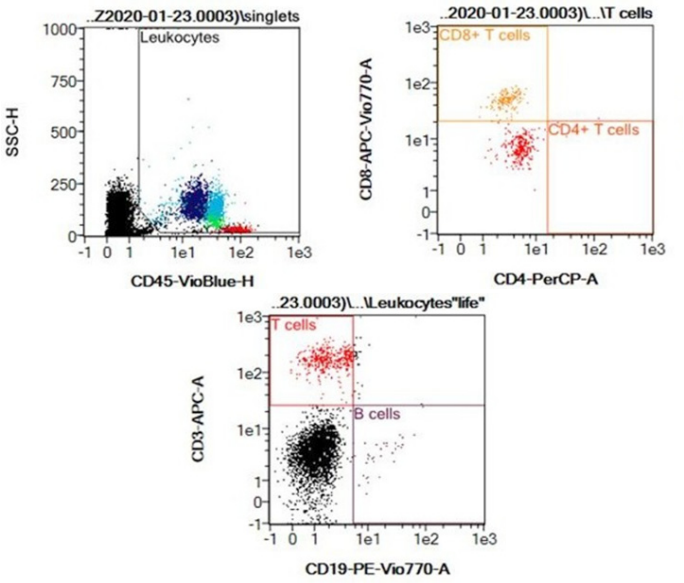

Figure 3: Immunophentyping of T lymphocyte subpopulations.

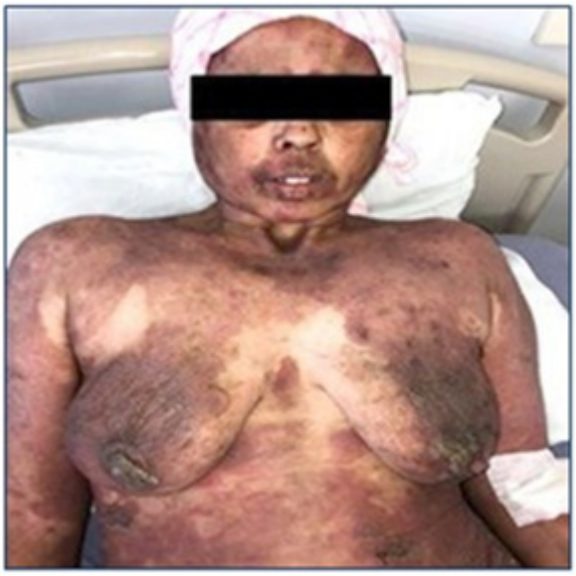

Figure 1: Quasi-erythroderma, dry, site of infiltrated hyperkeratotic plaques.

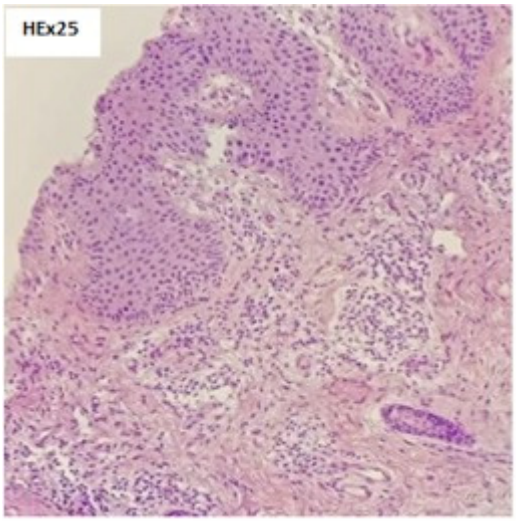

Figure 4: Dense lymphoid cell infiltrate of perivascular arrangement with epidermotropism ulcerating the epidermis.

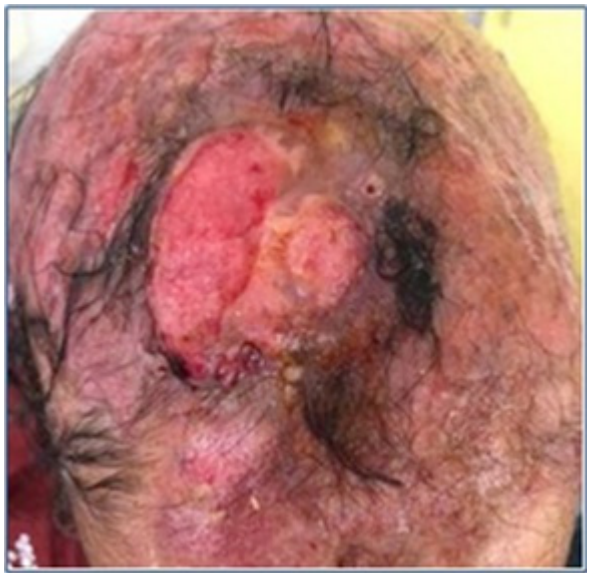

Figure 2: Ulcerative budding scalp tumor with alopecia.

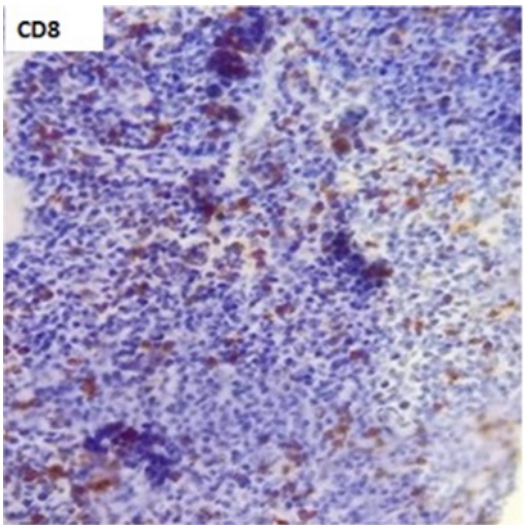

Figure 5: Moderate and diffuse membrane expression of anti-CD8 anticoprs tumor cells.

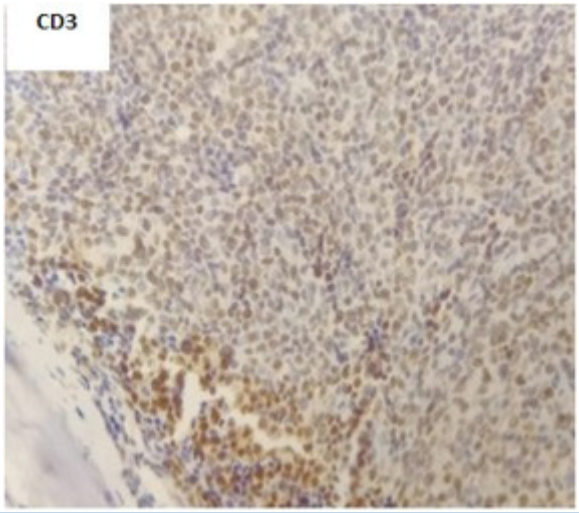

Figure 6: Moderate and diffuse membrane expression of anti-CD3 anticoprs tumor cells.

Discussion

CD8+ aggressive cutaneous epidermotropic T lymphoma (LCAE) is a rare and poorly characterized variant, still considered to be a temporary entity according to the WHO (World Health Organization) classification of 2017. About twenty cases have been reported in the literature [2,3,4].

Clinically, it presents as ulcerated, necrotic, sometimes hemorrhagic plaques, nodules or tumors, which develop rapidly and aggressively, giving rise to extra-cutaneous damage. Our observation illustrates a clinical presentation also associating a quasi-erythroderma, and a palmoplantar keratoderma [4,9].

Histologically, the infiltrate is composed of lymphocytes of varying sizes. Epidermotropism is constant. Skin immunostaining shows tumor CD8+T lymphocytes expressing cytotoxic proteins, which probably explains the local and general aggressiveness of the disease, the angiodestructive nature of the infiltrate and the necrotic lesions [2,3,9].

The differential diagnosis arises with other indolent cutaneous T lymphomas, mainly pagetoid reticulosis and CD8+ mycosis fungoides [8].

There is no standardized treatment to date. The first-line treatment generally involves the CHOP (cyclophosphamide, doxorubicin, vincristine, prednisolone) or EPOCH (Etoposide, Prednisone, Oncovin, Cyclophosphamide hydrochloride dexorubicin) protocols, but generally results in treatment failure. Recent studies suggest switching to second-line therapies such as gemcitabine, pralatrexate, brentuximab, as well as allogeneic stem cell transplantation, which may have allowed complete remission in a few rare cases [5,6,7]. Recurrence is the rule with a median survival not exceeding 34 months [4,8,9].

Conclusion

LCEA CD8 + is a lymphoma with a poor prognosis, despite the use of aggressive chemotherapy drugs. The use of gemcitabine, pralatrexate, brentuximab, as well as allogeneic stem cell transplantation seems to open up new therapeutic avenues.

Declarations

Ethics approval and consent to participate: Yes

Consent for publication: Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Availability of data and material: Data Availabiles

Competing interests: The authors declare no conflict of interest

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Authors' contributions

Imane. Boujguenna and Mohamed Amine Haouane: drafting of the manuscript

Anass Belbachir and Hanane Rais: correction of the manuscript.

Fatimazahra Marhoume and Anass Belbachir: analysis of cytometry data.

Imane Bahbouhi, Kenza Kandri and Ouafa hocar: clinical management of the patient.

All authors contributed to the conduct of this work.

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