QUIZ

MJS PUBLISHING

Extensive Scaly Erythema with Recurring Ulcers and Crusts: A Quiz

Danfeng ZHU[1#], Bingjie CHEN[2#] and Ying ZHOU[2*]
*[1]Department of Dermatology, The Fifth People's Hospital of Hainan Province, China, and [2]Institute of Dermatology and Venereal diseases, Department of Dermatology and Venereal diseases, Affiliated Hospital of Guangdong Medical University, No. 57 Renmin South Road, Xiashan District, Zhanjiang, Guangdong Province, 524001 China. *E-mail: zhouy10000@126.com*
[#]These authors contributed equally to this article.

A 29-year-old man presented to the dermatology clinic with a 1-year history of papular abscesses and crusts, alongside a 10-year history of generalized erythema and desquamation. He had previously been diagnosed with "eczema" at an outside hospital and was treated with glucocorticoids and antihistamines. While these treatments provided temporary relief, his symptoms recurred following discontinuation, resulting in a fluctuating disease course. The patient had no history of syphilis or HIV/AIDS, and laboratory evaluations revealed normal liver and kidney function as well as a normal electrocardiogram.

On physical examination, extensive, ill-defined erythema was observed on the patient's face, neck, and trunk (Fig. 1A). The erythematous regions exhibited scattered atrophic scars, some of which demonstrated heterochromia and significant surface desquamation. Additionally, scattered dark-red papules, dark-brown nodules with areas of surface necrosis, and some black, oyster-like crusts were noted (Fig. 1B). Scattered brown blood blisters were also present on both hands (Fig. 1C). Biopsies were obtained from a blood blister on the right hand and a nodule on the abdomen for histopathological examination and immunohistochemical analysis.

What is your diagnosis?

1: Lichenoid pityriasis

2: Primary cutaneous anaplastic large cell lymphoma

3: Lymphoid papulosis type E with mycosis fungoides

4: Extranodal NK/T-cell lymphoma

See next page for answer.

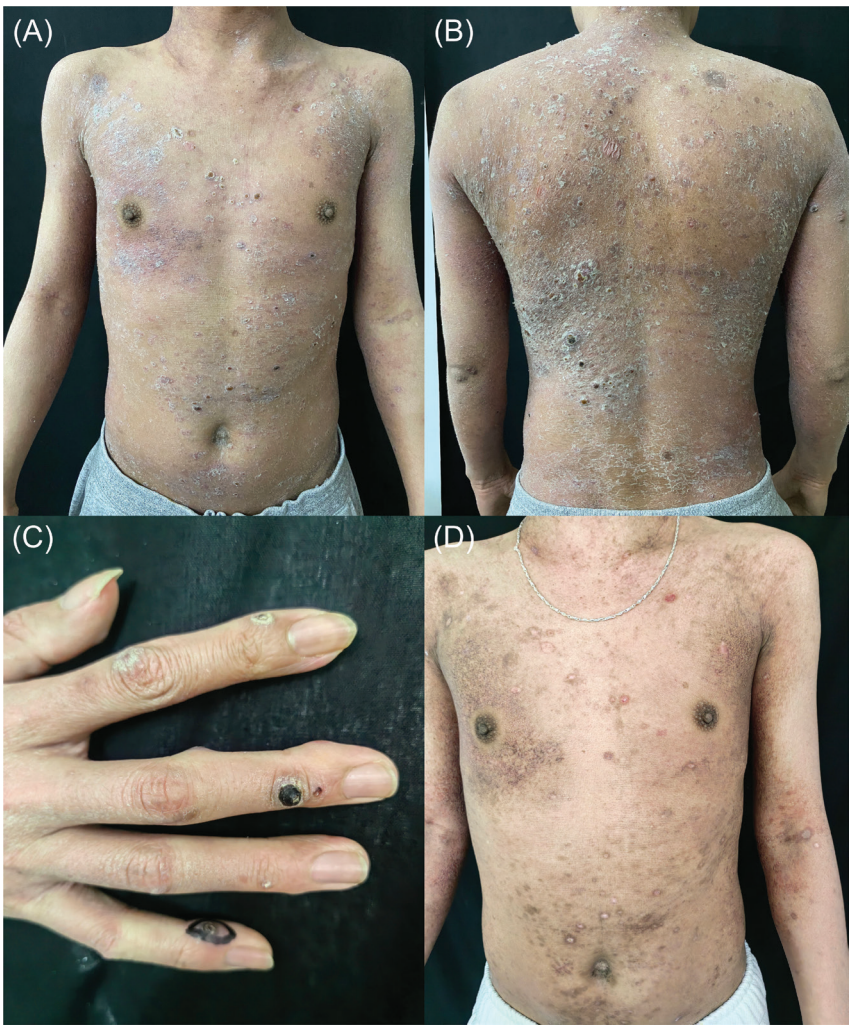

Fig. 1. Clinical image. (A) Clinical image showing extensive, ill-defined erythema on the patient's face, neck, and trunk. (B) Scattered dark-red papules, dark-brown nodules with areas of surface necrosis, and some black, oyster-like crusts were noted. (C) Scattered brown blood blisters on both hands. (D) The improvement in the patient's condition after 3 weeks of medication.

ActaDV Acta Dermato-Venereologica ActaDV Advances in dermatology and venereology

ANSWERS TO QUIZ

Extensive scaly erythema with recurring ulcers and crusts: A Commentary

Acta Derm Venereol 2025; 105: adv43151.
DOI: 10.2340/actadv.v105.43151

***Diagnosis*: Lymphoid papulosis type E with mycosis fungoides**

Lymphomatoid papulosis type E (LyPE) is a self-limiting lymphoproliferative disorder characterized by CD30+ and $CD8^+$ lymphoid cells, accompanied by destructive vascular infiltration and necrosis (1). Clinically, LyPE presents with papular nodules that often ulcerate and scar, typically resolving within several weeks. Mycosis fungoides (MF), the most common primary cutaneous T-cell lymphoma, is derived from $CD4^+$ effectors and follows a slow disease course (2). Its clinical manifestation includes pruritic erythematous patches covered by scales, typically distributed in "swimsuit" areas not exposed to sunlight, and often associated with telangiectasia (3). In cases where LyPE and MF co-occur, a combination of clinical benignity and histologic malignancy is observed, while the combination of immunohistochemical ($CD30^+$) T cell cloning helps us differentiate the diagnosis from other dermatitis and lymphoma.

Microscopic examination revealed abnormal keratinization, central necrosis, surface erosion, and dyskeratotic cells. Additional features included epidermal hyperplasia with mild oedema, nuclear hyperchromasia, basal cell degeneration, and dense lymphoid cell infiltration extending into the dermis (Fig. 2A). Pleomorphic T cells exhibit marked aggressiveness and destructive angiotropic growth, leading to vascular occlusion, thrombus formation, haemorrhage, extensive necrosis, and ulceration (Fig. 2B). Pathological sections demonstrated epidermotropism, with atypical lymphocytes infiltrating the epidermis. The infiltrating cells appeared slightly enlarged, showing the characteristic structure of MF – Pautrier microabscesses (Fig. 2C). Immunohistochemistry identified cells positive for CD30 (Fig. 3A), CD3, CD4 (Fig. 3B), CD56, CD7 and CD8 (Fig. 3C), TIA-1, and Ki-67, and negative for EBV-encoded RNA (EBER). T-cell clonality assessment revealed a monoclonal rearrangement.

The differential diagnosis for this presentation includes primary cutaneous anaplastic large-cell lymphoma (PC-ALCL), extranodal NK/T cell lymphoma (ENKTL), and lichenoid pityriasis. Diagnosis relies on a comprehensive assessment of clinical course, histopathology, and immunohistochemical findings. PC-ALCL is a rare subtype of $CD30^+$ T-cell non-Hodgkin lymphoma, manifesting clinically as localized masses on the face, trunk, and limbs, with positive CD30 and negative CD8 expression. Given the overlapping histopathological, phenotypic, and genetic features of PC-ALCL and LyP, careful clinical differentiation is required. PC-ALCL typically presents as ulcerative erythematous nodules, whereas LyP is marked by recurrent episodes of multiple papular nodular lesions that resolve spontaneously within weeks (4, 5). ENKTL is a rare, aggressive subtype of non-Hodgkin's lymphoma, histologically characterized by vascular destruction, coagulative necrosis, and associated with Epstein–Barr virus infection (6). While LyP also demonstrates vascular destruction, ENKTL's aggressive clinical course and EBER positivity aid in differentiation. The clinical features of LyPE can resemble those of pityriasis lichenoides, with spontaneous resolution of papules and evidence of lymphocytic vasculitis in the dermis on histopathology. Pityriasis lichenoides is more common in younger patients, has a shorter disease course, and lacks nodular lesions; CD30 is also not expressed histologically. In this case, the distinctive histopathological findings – CD3, CD4, CD8, and CD30 positivity – along

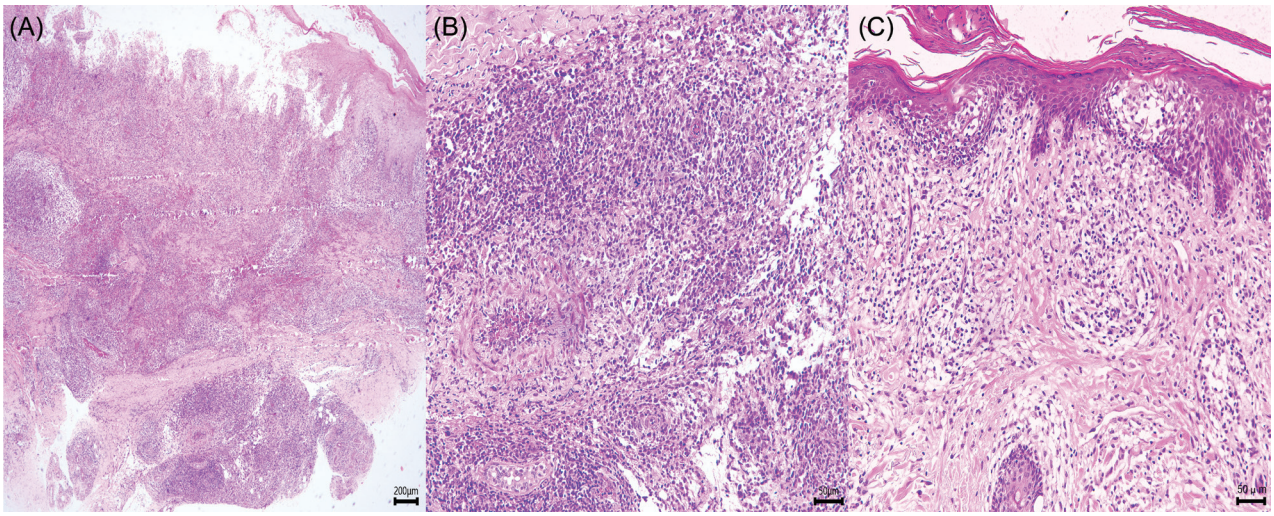

Fig. 2. Histopathological image. (A) Histopathology reveals dyskeratotic cells, epidermal hyperplasia with mild oedema, basal cell degeneration, and dense lymphoid cell infiltration extending into the dermis (H&E 40x). (B) Pleomorphic T-cells exhibit marked aggressiveness and destructive angiotropic growth, leading to vascular occlusion, thrombus formation, haemorrhage, extensive necrosis, and ulceration (H&E 200x). (C) The infiltrating cells appeared slightly enlarged, and Pautrier microabscesses were present (haematoxylin-eosin) (H&E 200x).

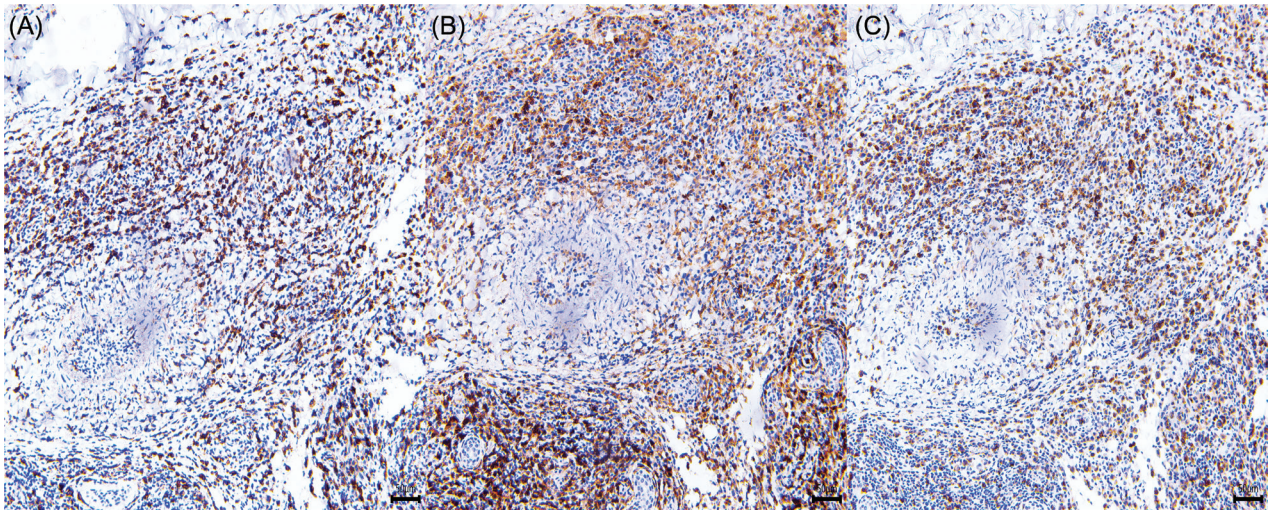

Fig. 3. Results of immunohistochemical analysis. (A) Moderate CD30 positive (original magnification ×200). (B) Moderate CD4 positive (original magnification ×200). (C) Moderate CD8 positive (original magnification ×200).

with clinical manifestations and T-cell clonality, support a diagnosis of LyPE combined with MF.

LyPE belongs to $CD30^+$ lymphoproliferative diseases; MF does not belong to $CD30^+$ lymphoproliferative diseases, but can express CD30, especially in the case of large-cell transformation. CD30-induced apoptosis may lead to the self-regression seen in LyP lesions (2). Meanwhile, Viral CD30 binds with high affinity to the CD30 ligand and blocks its interaction with CD30. CD30 is co-expressed on $CD4^+$ T cells that express T helper 2 (Th2) but not Th1 cytokines (7), which may explain why it can be seen in the late stages of MF.

LyP and MF share a common monoclonal T-cell receptor gene rearrangement, suggesting a common origin between the clinicopathologies of the 2 diseases (8). That is, the 2 diseases are different clinical manifestations of the same T-cell clone. Based on this, we consider the possibility of clinical disease based on LyPE based on existing MF risk genes.

The current treatment regimen includes methotrexate tablets (15 mg/week), anti-inflammatory immune modulation, and topical application of halometasone cream and allantoin cream. Regular follow-up visits were conducted post-treatment, and the patient's condition showed signs of improvement (Fig. 1D).

To our knowledge, only 1 prior case of LyPE coexisting with MF has been reported (9). A recent large retrospective study has confirmed a biological link between LyP and MF (10), though type E has been less frequently documented. Our case provides references for future diagnosis and treatment strategies for both diseases.

ACKNOWLEDGEMENTS

Funding sources: This project was supported by Hainan Province Clinical Medical Center.

IRB approval status: The authors thank the patient for granting permission to publish this information.

The authors have no conflicts of interest to declare.

REFERENCES

1. Kempf W, Kazakov DV, Schärer L, Rütten A, Mentzel T, Paredes BE, et al. Angioinvasive lymphomatoid papulosis: a new variant simulating aggressive lymphomas. Am J Surg Pathol 2013; 37: 1–13. https://doi.org/10.1097/PAS.0b013e3182648596
2. Duvic M. CD30+ neoplasms of the skin. Curr Hematol Malig Rep 2011; 6: 245–250. https://doi.org/10.1007/s11899-011-0096-8
3. Jawed SI, Myskowski PL, Horwitz S, Moskowitz A, Querfeld C. Primary cutaneous T-cell lymphoma (mycosis fungoides and Sézary syndrome): part I. Diagnosis: clinical and histopathologic features and new molecular and biologic markers. J Am Acad Dermatol Venereol 2014; 70: 205.e1–16; quiz 21–22. https://doi.org/10.1016/j.jaad.2013.07.049
4. Atmaja B, Millard T, Wotherspoon A, Attygalle AD, Cunningham D, Sharma B. A rare case of primary cutaneous-anaplastic large cell lymphoma (PC-ALCL) localized to the eyelid. Br J Haematol 2023; 200: 539. https://doi.org/10.1111/bjh.18573
5. Yan X, Ren W, Li S, Zhu Z, Gao L, Zhi K. Primary cutaneous anaplastic large-cell lymphoma resembling infratemporal space infection: a case report. BMC Oral Health 2024; 24: 470. https://doi.org/10.1186/s12903-024-04178-w
6. Akbar M, Clasen-Linde E, Specht L. Extranodal NK/T-cell lymphoma, nasal type, with extranasal presentation: a case report and a review of the literature. Acta Oncol 2020; 59: 1480–1487. https://doi.org/10.1080/0284186X.2020.1795250
7. Del Prete G, De Carli M, Almerigogna F, Daniel CK, D'Elios MM, Zancuoghi G, et al. Preferential expression of CD30 by human CD4+ T cells producing Th2-type cytokines. FASEB J 1995; 9: 81–86. https://doi.org/10.1096/fasebj.9.1.7821763
8. de la Garza Bravo MM, Patel KP, Loghavi S, Curry JL, Torres Cabala CA, Cason RC, et al. Shared clonality in distinctive lesions of lymphomatoid papulosis and mycosis fungoides occurring in the same patients suggests a common origin. Human Pathology 2015; 46: 558–569. https://doi.org/10.1016/j.humpath.2014.12.008
9. Kiavash K, Abner SM, Malone JC. New variant lymphomatoid papulosis type E preceding and coexisting with mycosis fungoides: a case report and review of the literature. J Cutan Pathol 2015; 42: 1018–1023. https://doi.org/10.1111/cup.12606
10. Melchers RC, Willemze R, Bekkenk MW, de Haas ERM, Horvath B, van Rossum MM, et al. Frequency and prognosis of associated malignancies in 504 patients with lymphomatoid papulosis. J Eur Acad Dermatol Venereol 2020; 34: 260–266. https://doi.org/10.1111/jdv.16065