Case Report: A case of hypertrophic lupus erythematosus with negative CD123 staining and absence of transepidermal elimination of elastin [version 2; peer review: 3 approved]

Previously titled: A case of hypertrophic lupus erythematosus with negative CD123 staining and transepidermal elimination of elastin

Matthew Hughes¹, Jerad M. Gardner¹,², Ling Gao¹

¹Department of Dermatology, University of Arkansas for Medical Sciences, Little Rock, AR 72205, USA
²Department of Pathology, University of Arkansas for Medical Sciences, Little Rock, AR 72205, USA

Abstract

We report the case of a 49-year-old male with clinical and histological findings consistent with hypertrophic lupus erythematosus (HLE). HLE must be clinically and histologically differentiated from keratoacanthoma, hypertrophic lichen planus, squamous cell carcinoma and plaque type psoriasis. CD123 positivity and transepidermal elimination of elastin have recently been reported as tools to distinguish HLE. Interestingly, in this case, biopsies of two separate lesions failed to reveal these two features. The etiology of this discrepancy is unknown and further studies are needed to clarify the utility of CD123 positivity and transepidermal elimination of elastin in the diagnosis of hypertrophic lupus erythematosus.
Corresponding author: Ling Gao (lgao@uams.edu)

Competing interests: No competing interests were disclosed.

Grant information: The author(s) declared that no grants were involved in supporting this work.

Copyright: © 2014 Hughes M et al. This is an open access article distributed under the terms of the Creative Commons Attribution Licence, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited. Data associated with the article are available under the terms of the Creative Commons Zero "No rights reserved" data waiver (CC0 1.0 Public domain dedication).

How to cite this article: Hughes M, Gardner JM and Gao L. Case Report: A case of hypertrophic lupus erythematosus with negative CD123 staining and absence of transepidermal elimination of elastin [version 2; peer review: 3 approved] F1000Research 2014, 3:76 (https://doi.org/10.12688/f1000research.3267.2)

Introduction

Hypertrophic lupus erythematosus (HLE) is a rare subset of discoid lupus erythematosus, characterized by erythematous, indurated, verrucous papules and nodules located on sun-exposed areas. HLE must be clinically and histologically differentiated from keratoacanthoma, hypertrophic lichen planus, squamous cell carcinoma and plaque type psoriasis. CD123 positivity and transepidermal elimination of elastin have recently been reported to distinguish HLE.

Report of case

A 49-year-old, unemployed, white male presented with a three-year history of an expanding “rash”. He reported no constitutional symptoms. He had previously been treated with oral prednisone and an unknown topical steroid without improvement and was off all medications at our initial visit. The patient had a past medical history of hepatitis C. He denied a family history of skin or autoimmune diseases. Laboratory work-up was significant for positive anti-nuclear antibodies and anti-Ro antibodies. Physical exam revealed multiple hyperkeratotic, verrucous papules and nodules with white, scaly, cribriform centers overlying patches of depigmentation, erythema and atrophy on his bilateral arms (Figure 1) and anterior legs. His face and scalp had several atrophic, depigmented patches. Two punch biopsies were obtained from separate lesions. Histological sections demonstrated an interface inflammatory pattern with deep peri-vascular and peri-appendageal lymphocytic infiltrate (hematoxylin and eosin, 40× magnification).

Discussion

HLE was first described by Bechet in 1940. Clinical diagnosis can be challenging as HLE can mimic psoriasis or even squamous cell carcinoma. Uitto et al. described two histological patterns of HLE. One resembled hypertrophic lichen planus, while the other was similar to keratoacanthoma. Daldon et al. found that transepidermal elimination of elastin was present in 14 cases of HLE. Recently, Ko et al. reported that a band of CD123 positive cells at the dermal-epidermal junction was characteristic of five cases of HLE.

In this patient, we examined these two recently described histologic features of HLE. Interestingly, both CD123 positivity and transepidermal elimination of elastin were not present in this case (Figure 3). However, the histological and clinical findings were most consistent with HLE. The etiology of this discrepancy is unknown and further studies are needed to clarify the utility of CD123 positivity and transepidermal elimination of elastin in the diagnosis of hypertrophic lupus erythematosus.

There is no definitive treatment for HLE. Options include topical or intralesional steroids, hydroxychloroquine, topical calcineurin inhibitors, topical or oral retinoids, thalidomide and surgical excision. Winchester et al. reported on the efficacy ustekinumab, an inhibitor of IL-12 and IL-23.

This case highlights the discrepancies of CD 123 positivity and absence of transepidermal elimination of elastin in HLE.
Consent
Written informed consent for publication of clinical details and clinical images was obtained from the patient.

Author contributions
Hughes – data collection, manuscript preparation
Gardner – data collection, manuscript preparation
Gao – manuscript preparation, oversight/supervision

Competing interests
No competing interests were disclosed.

Grant information
The author(s) declared that no grants were involved in supporting this work.

References
Open Peer Review

Current Peer Review Status: ✔️ ✔️ ✔️

Version 2

Reviewer Report 18 June 2014

https://doi.org/10.5256/f1000research.4723.r5159

© 2014 Lu T. This is an open access peer review report distributed under the terms of the Creative Commons Attribution Licence, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Theresa T. Lu
Autoimmunity and Inflammation Program and Pediatric Rheumatology, Hospital for Special Surgery, New York, NY, USA

The authors have now addressed all the concerns.

Competing Interests: No competing interests were disclosed.

I have read this submission. I believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard.

Reviewer Report 17 June 2014

https://doi.org/10.5256/f1000research.4723.r5010

© 2014 Werth V. This is an open access peer review report distributed under the terms of the Creative Commons Attribution Licence, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Victoria P. Werth
Department of Dermatology, Perelman School of Medicine, University of Pennsylvania, Philadelphia, PA, USA

The authors successfully addressed the reviewer comments in this case report.

Competing Interests: No competing interests were disclosed.

I have read this submission. I believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard.
Jordan Reynolds
Department of Anatomic Pathology, Cleveland Clinic, Cleveland, OH, USA

This article is well written but needs a few additions to improve it:
1. Please elaborate on what CD 123 is, and how it is used for diagnosing HLE. It is not common knowledge and should be expanded upon.

2. It might also be worth commenting on this paper by Miyashita A et al. (2013, Acta Derm Venereol) in which the authors showed patients with CD123 positive cells responded better to treatment than patients with low CD123. Do you think any of this could be supported or refuted by your cases?

Competing Interests: No competing interests were disclosed.

I have read this submission. I believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard.

Theresa T. Lu
Autoimmunity and Inflammation Program and Pediatric Rheumatology, Hospital for Special Surgery, New York, NY, USA

This report describes a case of hypertrophic lupus erythematosus based on clinical and histopathologic criteria that is negative for CD123 and elastin elimination. Negative data is important. However, as the emphasis is on the lack of CD123 and the lack of transepidermal elastin elimination, it would be good to show the negative results. For the CD123 stain, it would be good to show a positive control to make sure that the antibody really worked.

Competing Interests: No competing interests were disclosed.
I have read this submission. I believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

Reader Comment 28 May 2014

Jerad Gardner, UAMS, USA

Thank you for your commentary. The second version of this report will contain a histological photo of the positive control and negative CD 123 staining of the biopsy specimen.

Competing Interests: No competing interests were disclosed.

Reviewer Report 14 April 2014

https://doi.org/10.5256/f1000research.3507.r4229

© 2014 Werth V. This is an open access peer review report distributed under the terms of the Creative Commons Attribution Licence, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Victoria P. Werth
Department of Dermatology, Perelman School of Medicine, University of Pennsylvania, Philadelphia, PA, USA

This is a case of hypertrophic lupus erythematosus that is described as unusual in pathologic presentation.

The title needs to indicate absence of transepidermal elimination of elastin. It is currently unclear if transepidermal elimination of elastin was present.

The order of treatment described for HLE is confusing. One would start with hydroxychloroquine, then add quinacrine to hydroxychloroquine, with topicals as adjunctive therapy. Oral retinoids, thalidomide, and immuosuppressives would be options. Given that frequently there are multiple lesions that may actually koebnerize in a surgical scar, one would not include surgical excision as an option.

The report cited in favor of TNF-alpha inhibitor is on ustekinumab, which is not a TNF inhibitor. This needs revision.

Information about the antibody used for CD123 staining, as well as whether frozen or fixed tissue was used, is important. Anti-CD123 staining is not as good on fixed tissue. Were there any positive controls stained simultaneously?

Competing Interests: No competing interests were disclosed.
I have read this submission. I believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

Reader Comment 28 May 2014

Jerad Gardner, UAMS, USA

Thank you for your commentary. The title has been changed to more clearly reflect the absence of transepidermal elimination in this case. The treatments listed in the report were a review of treatment options from the literature. They were not ordered as a suggested line of therapy. Ustekinumab has been reported to improve the plaques of hypertrophic lupus erythematosus. It is an inhibitor of IL-12 and IL-23. This fact has been corrected. We performed CD 123 staining on paraffin embedded tissue which is the method Ko et al. employed in her report on the novel use of CD 123 staining in hypertrophic lupus erythematosus. The second version of this report will contain a figure displaying positive control staining and negative CD 123 staining of the biopsy from this case.

Competing Interests: No competing interests were disclosed.

The benefits of publishing with F1000Research:

- Your article is published within days, with no editorial bias
- You can publish traditional articles, null/negative results, case reports, data notes and more
- The peer review process is transparent and collaborative
- Your article is indexed in PubMed after passing peer review
- Dedicated customer support at every stage

For pre-submission enquiries, contact research@f1000.com