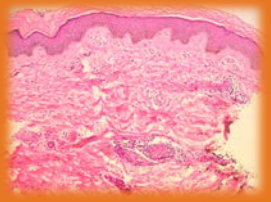
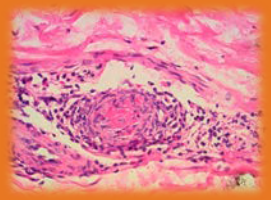
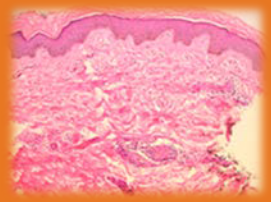
Case Presentation 16 - *Young Woman with Diffuse Lower Extremity Red-Brown Macules a Telltale Sign of Recently Described Macular Lymphocytic Arteritis*

Case Author: Cynthia M. Magro, MD
Contributing Author: James Y. Wang, MD MBA , Garron Solomon, MD and Chris G. Adigun, MD, FAAD

Weill Cornell Medicine Dermatopathology







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Introduction

Macular lymphocytic arteritis (MLA) was recently described in 2008 as a distinct form of larger vessel subcutaneous vasculitis separate from benign cutaneous polyarteritis nodosa (BCPAN). It clinically manifests as asymptomatic reticulated hyperpigmented macules on the lower extremity of women Histopathologically, the condition is characterized by a deep dermal and/or subcutaneous lymphocytic vasculitis of small-to-medium caliber arteries exactly recapitulating the type of vessels affected in BCPAN. Vascular thrombosis is prominent and explains the reticulated morphology of this condition from a clinical perspective.

Clinical History

A 33 year old healthy woman presented with a sudden onset of hundreds of reticulated red and brown nonscaly macules and very thin papules on her bilateral lower extremities with minimal extension to involve the lower trunk and bilateral forearms. (figure 1). Darier's sign was negative. The rash developed over the course of six weeks and had remained without clinical improvement over 3 months. The clinical differential diagnosis at presentation was pityriasis lichenoides chronica and lichen planus pigmentosus.

Histopathological Features

Histopathologic evaluation revealed a striking necrotizing thrombogenic lymphocytic arteritis involving a deeper seated reticular dermal blood vessel (figure 2).

Discussion

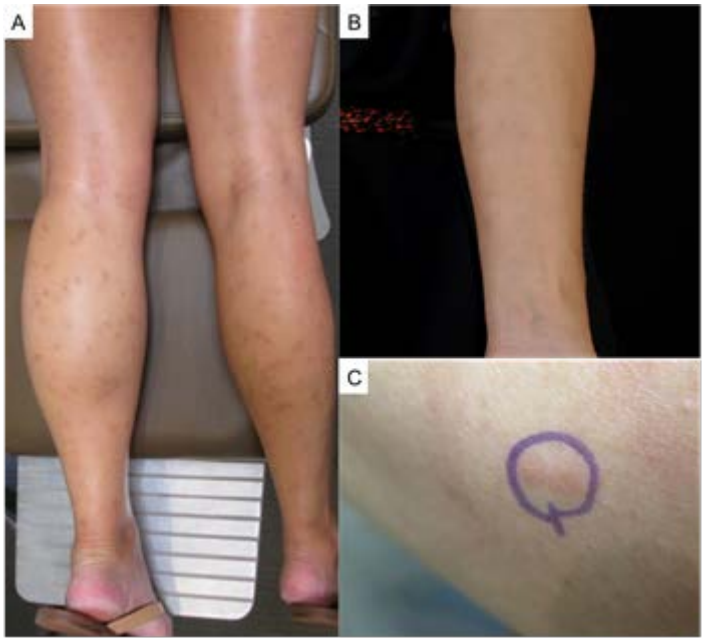
The distinct clinical pathological entity of macular lymphocytic arteritis presents as non-palpable hyperpigmented macules with a livedoid background and a histopathologically arteritic process associated with prominent angiocentric lymphocytic infiltrates and vascular thrombosis defining what is best described as a lymphocytic thrombogenic arteritis. Due to the prominent and almost ubiquitous pattern of thrombosis, this entity has fallen under the alternative designation of lymphocytic thrombophilic arteritis.

At one point, MLA was erroneously concluded to be a later stage lesion of BCPAN. However, macular lymphocytic arteritis is distinct from BCPAN. At all phases in its evolution, the infiltrate is lymphocytic. The vascular thrombosis is characteristically a very intrinsic component of this arteritic process. Clinically, patients tend to be relatively young females. The lesions of macular lymphocytic arteritis do not produce the classic nodular lesions of benign cutaneous polyarteritis nodosa, but rather manifest as macules.

The dominant lymphocytic infiltrate in and around vessels, accompanied by vascular thrombosis, suggests its association with an autoimmune-based thrombophilic tendency state. Indeed, a significant percentage of patients with this condition may have underlying antiphospholipid antibodies. There is evidence of C5b-9 deposition in the vessels although the typical Type I rich interferon microenvironment that characterizes classic lupus erythematosus is not seen. C5b-9 is indeed the effector mechanism of vascular thrombosis in the setting of antiphospholipid antibody syndrome and hence the pattern of prominent C5b-9 deposition in the vessels would be expected.

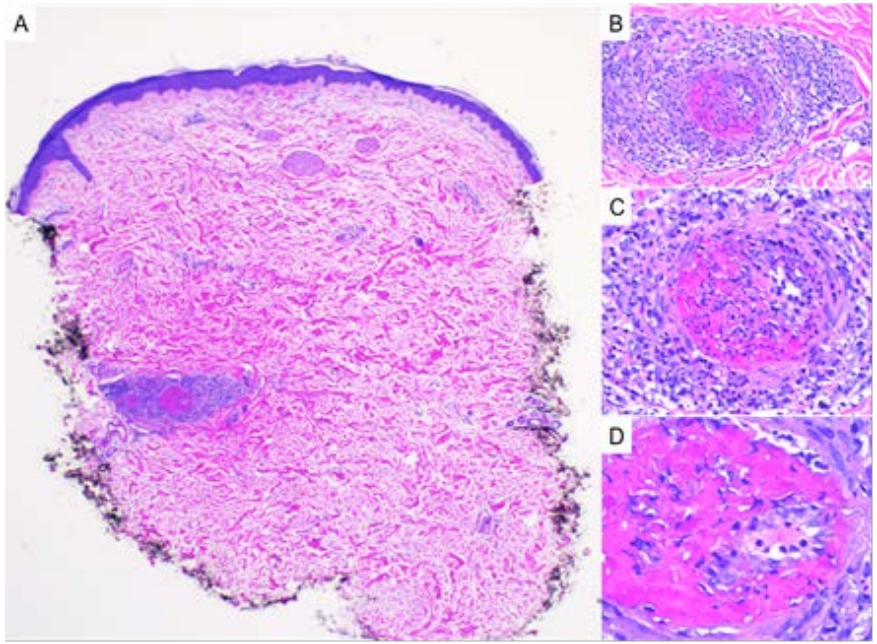
Figure Legend

1



***Figures 1a-c**: Scattered asymptomatic red-brown macules on the extremities are present in our patient. (A) bilateral legs (B) Left forearm (C) Biopsy site.*

2



***Figures 2a-c**: (A) A brisk lymphocytic vasculitis is present in a medium-sized vessel. (B-D) Higher power images showing lymphocytic infiltration of the vessel wall and extensive thrombosis of the vascular lumen.*

Case References

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Under the direction of Dr. Cynthia M. Magro, the Weill Cornell Comprehensive Dermatopathology Service is a leading edge consultation service and CAP-accredited laboratory for dermatologists, plastic and general surgeons and other dermatopathologists. Dr. Magro is an internationally renowned dermatopathologist, educator and author. She is a Professor of Pathology and Laboratory Medicine at the Weill Cornell Medical College in Manhattan, and is board certified in anatomic pathology, dermatopathology and cytopathology. Dr. Magro is an expert in the diagnosis of complex inflammatory skin diseases. Her areas of expertise include cutaneous manifestations of auto-immune disease, systemic viral disease and vasculitis, atypical drug reactions, benign, atypical and overtly malignant lymphocytic infiltrates of the skin, and diagnostically difficult melanocytic proliferations.The award-winning author of *The Melanocytic Proliferation: A Comprehensive Textbook of Pigmented Lesions*, Dr. Magro has recently completed her second book, *The Cutaneous Lymphoid Proliferation, a comprehensive textbook on benign and malignant lymphocytic infiltrates*. She has co-authored over 300 peer reviewed papers and several textbook chapters. Dr. Magro frequently presents courses on inflammatory skin pathology and difficult melanocytic proliferations to the American Academy of Dermatology, the United States and Canadian Academy of Pathology, and the American Society of Clinical Pathology. Dr Magro has consistently been recognized in Who's Who in America®, Castle Connolly's renowned America's Top Doctors – New York Metro Area® edition and in the Super Doctors® list published in The New York Times Magazine.

For more information, consultation, or patient referral please contact:

Cynthia M. Magro, MD
Director
Weill Cornell Comprehensive Dermatopathology Service
Tel. 212-746-6434
Toll-free 1-800-551-0670
ext. 66434
Fax. 212-746-8570
www.weillcornelldermpath.com