JAMA Dermatology Clinicopathological Challenge

Unilateral Atrophic Hyperpigmentation

Konstantin Dumann, MD; Jennifer Haiduk, MD; Mirjana Ziemer, MD

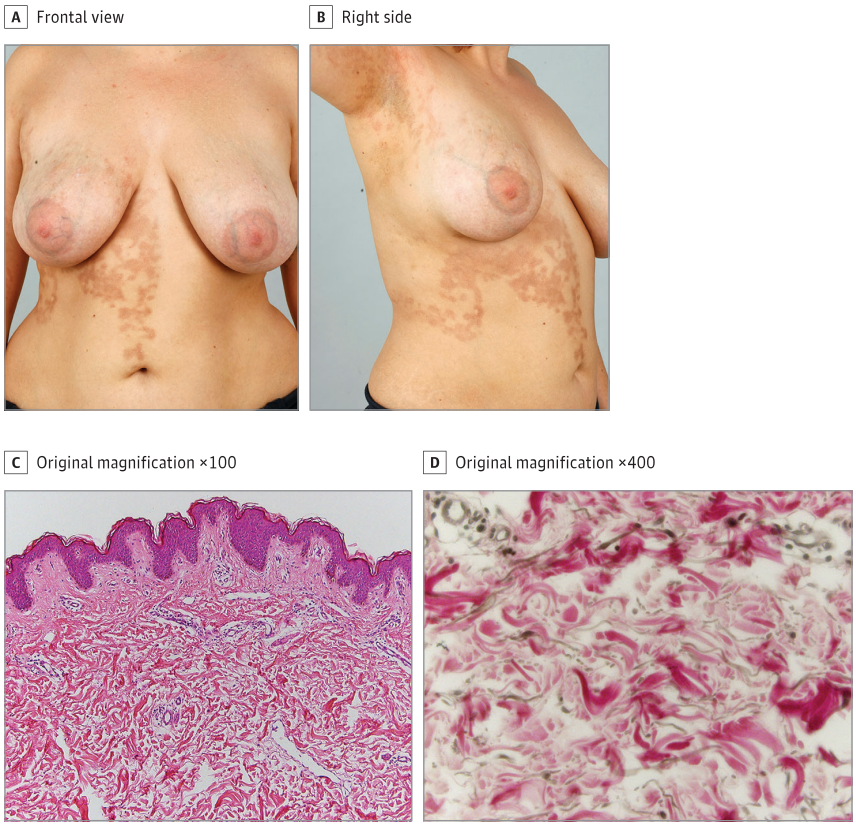

Figure. Clinical images show slightly atrophic hyperpigmented macules following the lines of Blaschko on the right side of the (A) trunk and (B) upper arm. Histopathologic images of punch biopsy specimen show (C) normal epidermis and dermis with a slight hyperpigmentation of the basal epidermis layer (hematoxylin-eosin stain), and (D) preserved elastic fibers in the reticular dermis (Elastic van Gieson stain).

A woman in her late 20s presented with an asymptomatic hyperpigmentation on the right side of the trunk, right upper arm, and right leg, strictly following the lines of Blaschko (**Figure**, A and B). A slight atrophy of the hyperpigmented skin was palpable without any signs of induration.

The first onset was in her childhood, at the age of 6 years, with a small hyperpigmentation on the right side of her lower chest remaining stable for years. In her early 20s the area of hyperpigmentation started to spread slowly over the above-mentioned areas and became more and more intense. Intravenous penicillin G therapy with 10 million units 3 times daily over 1 week combined with a UV-A1 phototherapy in 18 sessions with a maximum dose of 40 J/cm^2 and a cumulative overall dose of 670 J/cm^2 showed no effects. A punch biopsy was obtained (Figure, C and D).

WHAT IS YOUR DIAGNOSIS?

A. Epidermal nevus

B. Linear atrophoderma of Moulin

C. Lichen striatus

D. Segmental incontinentia pigmenti

Diagnosis

B. Linear atrophoderma of Moulin

Discussion

Microscopic examination showed a hyperpigmentation of the basal layer of the epidermis. In the dermis there were only very sparse superficial perivascular lymphocytic infiltrates. The dermis otherwise remained normal, and elastic fibers were preserved. Significant papillomatous epidermal hyperplasia was not apparent, and the specimen was minimally inflammatory, without evidence of lichenoid or interface change, or significant perivascular or perieccrine lymphocytic infiltrates (Figure, C and D). Together with the clinical

image and histopathologic analysis, the diagnosis was linear atrophoderma of Moulin.

In 1992 Moulin et al[1] described a segmental hyperpigmented atrophy named atrophoderma of Moulin. The age at onset usually is in childhood or adolescence. The clinical picture shows atrophic hyperpigmentations following the lines of Blaschko. Ultrasonographic imaging suggests that the clinical atrophy is not associated with dermal changes as seen in scleroderma, but rather owing to subcutaneous atrophy. Microscopic findings therefore are unspecific and mostly reveal a basal hyperpigmentation without inflammatory or sclerotic changes. Most likely atrophoderma of Moulin is identical to published segmental forms of atrophoderma idiopathica Pasini-Pierini.[2,3] Atrophoderma idiopathica Pasini-Pierini—first described in 1923 and 1935 by the 2 authors subsequently—was discussed as a nonatrophic macular hyperpigmented variant of morphea. However, Canizares et al[4] believed that it differs sufficiently from morphea and should be classified as a distinct entity. It is noticeably more frequent in female patients. Manifestation usually also begins in childhood or adolescence. The dermal atrophy manifests as single or multiple sharply demarcated, round or oval, hyperpigmented, nonindurated patches of varying sizes ranging from a few millimeters to several centimeters. The lesions may be discrete or confluent and are usually asymptomatic and do not show any signs of inflammation. These patches are marked by a slight depression of the skin. Histopathologic changes are minimal and not diagnostic. The epidermis is usually normal or slightly atrophic. A mild perivascular infiltrate consisting of lymphocytes may be present. When compared with adjacent normal skin, dermal thickness is reduced. Adnexal structures are not affected.[4,5]

Besides intravenous penicillin and UV therapy, as in this case, oral potassium para-aminobenzoate (POTABA), high-dose vitamin E (400 IU/d), topical clobetasol propionate, and topical calcipotriol are reported to show a positive effect. However, there are no evidence-based guidelines or standard therapies for atrophoderma Moulin.[6]

In sum, atrophoderma Moulin is a segmental genotypic and phenotypic mosaicism. Although discussed as a segmental variant of morphea or atrophoderma idiopathica Pasini-Pierini, its etiology still remains unclear.

Epidermal nevi distributed along the lines of Blaschko show a broad spectrum of clinical manifestation. They vary from linearly distributed elevated hyperkeratotic, verrucous or erythematosquamous plaques to solely hyperpigmented macules. A mostly hyperpigmented variant is Becker nevus. It manifests with hyperpigmented macules in childhood but becomes clinically more apparent after puberty owing to the growth of adnexal structures, especially follicular-sebaceous units, possibly related to the androgen stimulus.[7] Atrophy, however, is not reported so we could exclude this differential diagnosis.[7]

Lichen striatus can result in postinflammatory hyperpigmentation. However, before developing a hyperpigmentation lichen striatus usually presents linearly distributed erythematosquamous lichenoid or even psoriasiform papules and plaques. Histologically, epidermal changes show spongiosis and interface dermatitis. In late stages, melanophages remain in the upper dermis as a result of pigment incontinence from epidermal keratinocytes. Inflammatory or palpable lesions have never been mentioned in this patient.[8]

Incontinentia pigmenti could easily be excluded clinically as well as histologically. It usually starts with a blistering inflammation of the skin evolving through 4 stages ending with postinflammatory linear hyperpigmentation with melanophages in the upper dermis. In most patients, early inflammatory stages are present at birth or develop in the first months of life.[9] Segmental postzygotic mosaicisms have been reported especially in male patients, where incontinentia pigmenti is normally lethal.[9,10]

ARTICLE INFORMATION

Author Affiliations: Department of Dermatology, University Hospital Leipzig, Leipzig, Germany.

Corresponding Author: Konstantin Dumann, MD, Department of Dermatology, University Hospital Leipzig, Philipp-Rosenthal-Str 23, 04103 Leipzig, Germany (konstantin.dumann@medizin.uni-leipzig.de).

Published Online: March 11, 2020. doi:10.1001/jamadermatol.2020.0069

Conflict of Interest Disclosures: None reported.

Additional Contributions: We thank the patient for granting permission to publish this information.

Self-assessment Credit: This article is eligible for journal-based self-assessment (1 credit) for Maintenance of Certification (MOC) from the American Board of Dermatology (ABD). After completion of an activity, please log on to the ABD website at www.abderm.org to register your credits. This may be done after each exercise or after accumulating many credits.

REFERENCES

1. Moulin G, Hill MP, Guillaud V, Barrut D, Chevallier J, Thomas L. [Acquired atrophic pigmented band-like lesions following Blaschko's lines]. *Ann Dermatol Venereol*. 1992;119(10):729-736.

2. Norisugi O, Makino T, Hara H, Matsui K, Furuichi M, Shimizu T. Evaluation of skin atrophy associated with linear atrophoderma of Moulin by ultrasound imaging. *J Am Acad Dermatol*. 2011;65(1):232-233. doi:10.1016/j.jaad.2009.12.005

3. Tan SK, Tay YK. Linear atrophoderma of Moulin. *JAAD Case Rep*. 2016;2(1):10-12. doi:10.1016/j.jdcr.2015.10.005

4. Canizares O, Sachs PM, Jaimovich L, Torres VM. Idiopathic atrophoderma of Pasini and Pierini. *AMA Arch Derm*. 1958;77(1):42-58. doi:10.1001/archderm.1958.01560010044007

5. Muntyanu A, Redpath M, Roshdy O, Jfri A. Idiopathic atrophoderma of Pasini and Pierini: case report and literature review. *Clin Case Rep*. 2018;7(2):258-263. doi:10.1002/ccr3.1958

6. Wongkietkachorn K, Intarasupht J, Srisuttiyakorn C, Aunhachoke K, Nakakes A, Niumpradit N. Linear atrophoderma of Moulin: a case report and review of the literature. *Case Rep Dermatol*. 2013;5(1):11-14. doi:10.1159/000346747

7. Danarti R, König A, Salhi A, Bittar M, Happle R. Becker's nevus syndrome revisited. *J Am Acad Dermatol*. 2004;51(6):965-969. doi:10.1016/j.jaad.2004.06.036

8. Johnson M, Walker D, Galloway W, Gardner JM, Shalin SC. Interface dermatitis along Blaschko's lines. *J Cutan Pathol*. 2014;41(12):950-954. doi:10.1111/cup.12412

9. Greene-Roethke C. Incontinentia pigmenti: a summary review of this rare ectodermal dysplasia with neurologic manifestations, including treatment protocols. *J Pediatr Health Care*. 2017;31(6):e45-e52. doi:10.1016/j.pedhc.2017.07.003

10. Alabdullatif Z, Coulombe J, Steffann J, Bodemer C, Hadj-Rabia S. Postzygotic mosaicism and incontinentia pigmenti in male patients: molecular diagnosis yield. *Br J Dermatol*. 2018;178(4):e261-e262. doi:10.1111/bjd.16092