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An Atypical Figurate Erythema With Seasonal Recurrences

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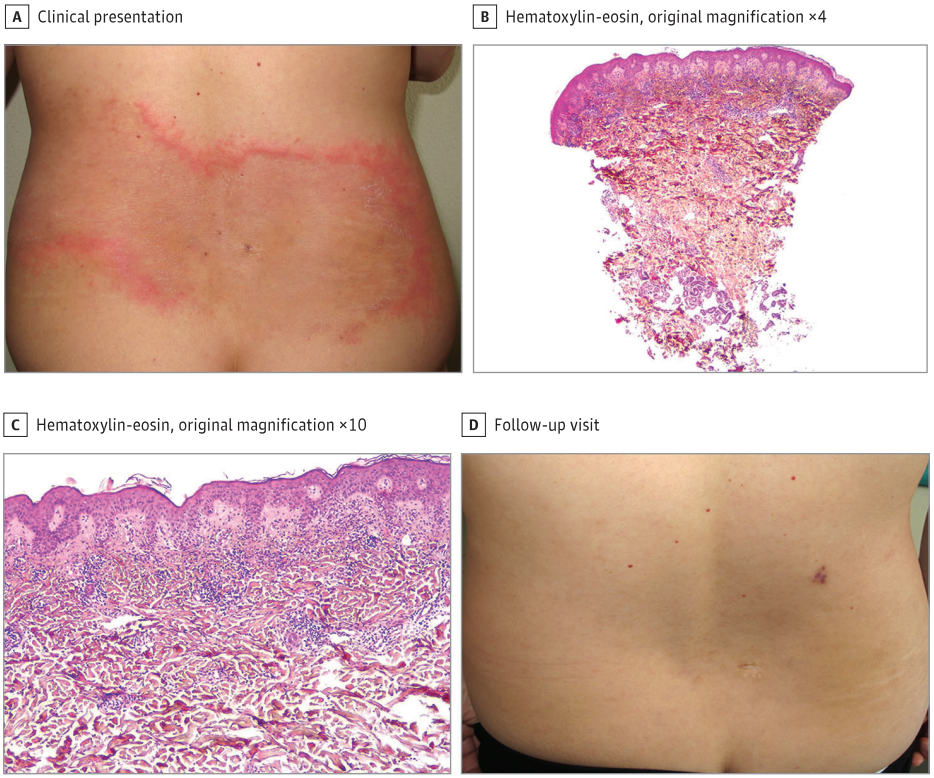

Figure. A, Large erythematous semicircular plaque with central postinflammatory hyperpigmentation on the lower back. B, Absence of epidermal changes, mild edema of the papillary dermis, and a superficial sparse perivascular inflammatory infiltrate. C, Inflammatory infiltrate composed mainly by lymphocytes and few neutrophils. D, Follow-up visit showing clinical resolution in October.

A healthy woman in her 50s presented with a large annular erythematous plaque on her lower back that had appeared in June (1 month previously) and had centrifugally expanded. She complained about mild pruritus that had not responded to conventional antihistamine therapy or topical steroids. She could not recall an arthropod bite prior to the skin eruption and denied having traveled outside Spain in the past few years. She reported a 20-year history of the same annular plaques recurring on the trunk every single year by the beginning of summer and spontaneously resolving by the beginning of autumn. Such lesions had never appeared on the face, hands, or feet. She did not relate these lesions to any particular hobby or application of any skin care product. Physical examination revealed a large, 35-cm semicircular erythematous plaque on her lower back with central clearing and a distinctive, well-demarcated papular border (**Figure**, A and B). After obtaining informed consent, 2 biopsy specimens were taken from the papular border of the plaque for both hematoxylin-eosin stain and direct immunofluorescence (Figure, C).

WHAT IS YOUR DIAGNOSIS?

A. Granuloma annulare

B. Erythema papulosa semicircularis recidivans

C. Deep erythema annulare centrifugum

D. Erythema chronicum migrans

Diagnosis

B. Erythema papulosa semicircularis recidivans

Microscopic Findings and Clinical Course

Histopathologic examination revealed no epidermal changes, mild edema of the papillary dermis, and a superficial sparse perivascular inflammatory infiltrate composed mainly of lymphocytes and few neutrophils (Figure, C). Periodic acid–Schiff staining was performed and did not show fungal microorganisms in the skin surface. Direct immunofluorescence studies were negative for IgG, IgM, IgA, complement, and fibrin deposition.

Complete blood cell count and biochemical studies did not show any significant finding. Results for all serological analyses, including *Borrelia burgdorferi* antibodies, proved to be negative.

Treatment with topical steroids and antihistamines was maintained daily during the first weeks of summer without an apparent clinical response. However, a subsequent follow-up visit in October confirmed that the lesion had spontaneously resolved without any further treatment a few weeks before evaluation (Figure, D).

Discussion

Erythema papulosa semicircularis recidivans (EPSR) represents a novel cutaneous disorder that manifests as centrifugally spreading semicircular erythematous plaques that typically relapse every year during the warm seasons and resolve in autumn.[1] First described by Song et al[2] in 2012, their case series of 9 middle-aged Chinese men ages 24 to 39 years from the Chongqing or Sichuan provinces presented with similar large figurate erythemas in the trunk that recurred yearly every summer. Since then, a few more cases have been also reported with the same seasonal pattern in young women from other countries.[3] While the disease course of this disorder might share some similarities with annually recurring erythema annulare centrifugum,[4] for some authors its clinical presentation differs slightly enough to consider it an independent entity.[1,2]

Differential diagnosis in figurate erythemas can be challenging. A good clinical history and clinicopathological correlation in this setting is of utmost importance.[5] Other annular erythemas similar to our case must be considered, such as erythema annulare centrifugum, erythema chronicum migrans, granuloma annulare, erythema marginatum, and dermatophytosis.

Erythema annulare centrifugum can present as a superficial or deep variant. Both presentations begin as small papules that slowly expand centrifugally and develop central clearing.[5,6] In the superficial form, a trailing scale is commonly observed at the advancing edge. The size of the plaques is much smaller than in EPSR, and it does not usually relapse in a seasonal pattern, although in some cases this has been described.[4] A biopsy specimen from the peripheral border will commonly show a superficial and deep perivascular infiltrate of lymphocytes surrounding the vessels with a "coat-sleeve" arrangement. Superficial forms may present focal parakeratosis and spongiosis in the overlying epidermis. Such a pattern was not observed in this case.

Erythema chronicum migrans is associated with the first stages of Lyme disease and usually presents as an expanding annular plaque that develops at the site of the bite of a Borrelia-infected tick.[7] Histopathologic analysis usually shows a superficial and deep perivascular and interstitial infiltrate of lymphocytes, plasma cells, and eosinophils. Spirochetes can be identified with the Warthin-Starry silver stain in approximately half of the specimens. Anti–*Borrelia burgdorferi* IgM antibodies can be detected from 3 to 6 weeks into the acute infection, which in this case resulted repeatedly in negative results.

Granuloma annulare is a self-limited dermatosis that can present in many different forms, ranging from localized, generalized, perforating, to subcutaneous variants.[8] Histopathologic analysis commonly reveals collagenolytic granulomas, with areas of necrobiosis in the superficial and mid dermis surrounded by a peripheral rim of palisaded histiocytes and lymphocytes.

According to the initial description by Song et al,[2] the prognosis of their patients with EPSR is marked by a spontaneous resolution after 2 to 5 years of annual recurrences. However, in the patient described herein, the disease period has been even longer, as she has been experiencing seasonal relapses for the past 20 years.

ARTICLE INFORMATION

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REFERENCES

1. Gönül M, Cevirgen Cemil B, Keseroglu HO, Kaya Akis H. New described dermatological disorders. *Biomed Res Int*. 2014;2014:616973. doi:10.1155/2014/616973

2. Song Z, Chen W, Zhong H, Ye Q, Hao F. Erythema papulosa semicircularis recidivans: a new reactive dermatitis? *Dermatitis*. 2012;23(1):44-47.

3. Inoue A, Sawada Y, Ohmori S, et al. Erythema papulosa semicircularis recidivans associated with primary pancreas B cell lymphoma. *Eur J Dermatol*. 2016;26(3):306-307.

4. García Muret MP, Pujol RM, Gimenez-Arnau AM, Barranco C, Gallardo F, Alomar A. Annually recurring erythema annulare centrifugum: a distinct entity? *J Am Acad Dermatol*. 2006;54(6):1091-1095.

5. Ríos-Martín JJ, Ferrándiz-Pulido L, Moreno-Ramírez D. Approaches to the dermatopathologic diagnosis of figurate lesions [in Spanish]. *Actas Dermosifiliogr*. 2011;102(5):316-324.

6. Ziemer M, Eisendle K, Zelger B. Erythema annulare centrifugum: a clinical reaction pattern [in German]. *Hautarzt*. 2010;61(11):967-972.

7. Bhate C, Schwartz RA. Lyme disease, part I: advances and perspectives. *J Am Acad Dermatol*. 2011;64(4):619-636.

8. Piette EW, Rosenbach M. Granuloma annulare: clinical and histologic variants, epidemiology, and genetics. *J Am Acad Dermatol*. 2016;75(3):457-465.