Koebner phenomenon in leukocytoclastic vasculitis: A case report and an updated review of the literature

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Abstract
Leukocytoclastic vasculitis is the most common form of cutaneous vasculitis. It is a neutrophilic small vessel vasculitis resulting from the deposition of circulating immune complexes. Henoch-Schonlein purpura is a systemic type of leukocytoclastic vasculitis, characterized by immunoglobulin A-mediated blood vessel injury. We present a case of Henoch-Schonlein purpura in an adult female manifesting with a vasculitic rash with Koebner phenomenon.

Keywords
leukocytoclastic vasculitis, vasculitis, Koebner phenomenon, isomorphic response

Introduction
An isomorphic response (or true koebnerization) is not uncommonly seen in psoriasis, lichen planus, and vitiligo. However, it is not commonly seen in vasculitis. This article includes an updated literature review of the Koebner phenomenon (KP) occurring in association with a leukocytoclastic vasculitis (LCV).

Case report
A 52-year-old female with end-stage renal disease (ESRD) of unknown cause was evaluated as an inpatient by the dermatology service for a 4-day history of a purpuric rash on the lower extremities. The patient described generalized pruritus for the past 5 years. On review of symptoms, there was no history of fevers, abdominal pain, arthralgias, hematuria, hematochezia, or melena. No new medications had been introduced prior to the cutaneous eruption.

Of note, the patient reported two episodes of an analogous rash prior to this presentation. The first episode developed during childhood and was associated with abdominal pain. The second episode occurred 10 years ago, and biopsies done at this time showed histopathological findings consistent with LCV.

Physical examination revealed a female with Fitzpatrick skin type III. Over the lower extremities, non-blanchable purpuric papules were observed in a linear arrangement, which was consistent with KP (Figure 1).

Serological investigations revealed an unremarkable autoimmune workup including a normal antinuclear antibody (ANA), perinuclear antineutrophil cytoplasmic antibodies (pANCA), cytoplasmic antineutrophil cytoplasmic antibodies (cANCA), rheumatoid factor (RF), C3, and C4. Workup for Hepatitis C and B were also negative. Punch biopsy from a purpuric area on the left leg demonstrated a small vessel vasculitis on histopathology with positive immunoglobulin A (IgA), C3, and fibrin on direct immunofluorescence (DIF). The clinicopathological diagnosis of Henoch-Schonlein purpura (HSP) was made. The patient was started on prednisone 50 mg once daily for 1 week and was then tapered off the prednisone with significant improvement.

Discussion
The isomorphic response (later called the KP) was first described by German dermatologist, Heinrich Koebner, in 1876.¹ This phenomenon refers to the development of an...

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existing dermatosis in previously normal skin, induced by either internal or external trauma. Isomorphic translates from Latin to “same shape,” with the prototypical disease demonstrating a KP being psoriasis.

Boyd and Nelder classified KP into four different groups. The first group, true koebnerization, includes psoriasis, lichen planus, and vitiligo, whereby KP is reproducible in all patients by a variety of insults. Boyd and Nelder argued that the term KP should be reserved for this group of diseases. The second group, pseudo-koebnerization, includes diseases such as molluscum contagiosum and verruca vulgaris, whereby KP is produced by the seeding of infectious agents along externally traumatized sites, usually from scratching. The third group, occasional lesions, includes diseases such as Darier disease and erythema multiforme, whereby some criteria are met for KP, but not all. Finally, the fourth group, questionable trauma-induced process, includes all disorders that have a questionable association with trauma such as pemphigus vulgaris and lupus erythematosus. Due to the paucity of reports of KP in vasculitis, LCV has been categorized in group four by Boyd and Nelder.

LCV is a neutrophilic small vessel vasculitis resulting from the deposition of circulating immune complexes. HSP is a common small vessel vasculitis, which is characterized specifically by IgA-mediated blood vessel injury affecting multiple sites. A comprehensive literature review of reports published in the English language revealed a total of nine reports on KP in LCV. Seven of the nine articles were reported in adults, while the remaining two articles were reported in the pediatric population. The majority of reports (seven articles) have been published on KP specifically in HSP, with the remaining two articles reporting KP in cases of medication-induced LCV. Of the seven reports of KP in HSP, only four described IgA in vessels on DIF to confirm the diagnosis of HSP. Our case which also showed IgA on DIF would be the fifth documented case of KP in HSP.

In addition, two studies have reviewed the prevalence of KP in patients hospitalized with LCV. One study found that 10% of patients with LCV presented with KP (n = 98), while the other found that 26% of children hospitalized with HSP presented with KP (n = 31). Interestingly, there have also been two case reports on the topic of reverse KP in LCV, in which skin lesions have disappeared at the sites of trauma.5,16

The mechanisms underlying KP in LCV are not well understood. However, it has been hypothesized that the increase in tryptase levels following trauma may play a role. In addition, Papi and Didona postulate that prolonged pressure inducing microcirculatory stasis, scratching inducing mast cell activation, and scars altering vascular anatomy are all plausible mechanisms responsible for KP in cutaneous vasculitis. One of the most commonly cited explanations for KP in vasculitis is the hypothesis that immune complexes aggregate at the site of inflammation induced by scratching.4

The patient presented in this article had generalized pruritus secondary to ESRD. We presume that her scratching due to uremic pruritus produced Koebnerized LCV lesions. A possible explanation for the small number of reported cases of KP in LCV may be because LCV is typically not pruritic. We would predict that more cases of Koebnerization would be seen in LCV if more patients with LCV were simultaneously pruritic from other causes including other systemic illnesses. We also feel that there are now sufficient cases of KP in LCV for this association to be included in the Boyd and Nelder classification as a group one disorder (i.e. true koebnerization).

Further studies investigating the prevalence of KP in various patient populations with LCV are required, as well as further research into the mechanisms that explain this cutaneous manifestation.

Declaration of conflicting interests
The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding
The author(s) received no financial support for the research, authorship, and/or publication of this article.

Informed consent
Informed consent for publication of the case history and the clinical image was provided by the patient.

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