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Case Presentation

Nail lichen planus in a patient with alopecia totalis

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Abstract

A 67-year-old man with a three-year history of non-scarring alopecia that progressed to alopecia totalis despite intralesional glucocorticoid injections is presented. He developed 20-nail dystrophy that was recalcitrant to antifungal and anti-inflammatory treatments. Biopsy of the nail matrix showed histopathologic features of lichen planus. Alopecia totalis and isolated lichen planus of the nails are uncommon subtypes of common dermatologic disorders. Rarely reported concurrently, we provide a review of the literature of their association, which is most likely attributed to their autoimmune pathogenesis.
Case synopsis

History: A 67-year-old otherwise healthy man was referred for the evaluation of progressive hair loss and nail changes. Three years prior to presentation, he developed asymptomatic and progressive thickening of all 20 nails. Six months later, he began to develop asymptomatic, non-scarring hair loss in round patches. He did report having similar patches of hair loss years earlier that resolved, which he associated with the onset and decline of stress. His new hair changes initially showed some incomplete improvement with the use of intraleosional triamcinolone acetonide but stabilized after six months. One year after initial presentation, he began to notice loss of hair on the arms. Over the next three months, he started losing abdominal hair; his anxiety level appreciably worsened and his scalp-hair loss progressed to complete involvement. A year later, eyelash and eyebrow hair began to fall out. Throughout this period, he continued to receive intraleosional injections for the scalp and topical minoxidil and ketoconazole shampoo were added, each without benefit.

Over this course of time, his nails were treated with separate courses of topical ciclopirox, ketoconazole, urea, fluocinolone, calcipotriene, and hydroxypropyl-chitosan, all without improvement. Several nail clippings were sent for culture and for potassium hydroxide examination, each showing no evidence of fungi. Upon presenting to the Skin and Cancer Unit, the decision was made to perform a nail matrix biopsy.

Physical examination: There was diffuse hair-loss of the entire scalp and eyebrows, with partial hair-loss of the eyelashes, arms and trunk. Follicular ostia remained intact, with no evidence of scars, erythema, or scale. All 20 nails showed onychodystrophy, with converging longitudinal ridges, lateral nail thinning, and some proximal nail folds with a dorsal pterygium.

Laboratory data: A complete blood count, comprehensive metabolic panel, lipid panel, anti-nuclear antibody, anti-Ro antibody, 25-OH vitamin D, thyroid-stimulating hormone, and urinalysis were normal or negative.

Histopathology: There is a band-like proliferation of lymphocytes within the superficial, subungual, soft tissue. Lymphocytes are present at the dermoepidermal junction where there are rare necrotic keratinocytes. A periodic acid-Schiff with diastase stain fails to show a thickened basement membrane or evidence of fungi.

Diagnosis: Nail lichen planus in a patient with alopecia totalis

Comment: Lichen planus (LP) and alopecia areata (AA) represent two of the most common diseases encountered by dermatologists. However, their subtypes of nail LP and alopecia totalis (AT) are encountered much less commonly. Therefore, as would be expected, their co-occurrence has rarely been documented in the literature. Cutaneous LP may be found in 0.2 to 1% of the population, with nail disease in 10% of cases and rarely as an isolated phenomenon [1-3]. Similarly, AA is prevalent in 0.1 to 0.2% of the population, but its progression to AT is rare [4]. Like LP, AA commonly affects the nails and even accounts for 3.3 to 12% of cases of 20-nail dystrophy [5]. However, as demonstrated in our patient, it should not be assumed that trachyonychia in a patient with AA is attributable to this underlying disease.

There have be no documented cases of a patient with AT and isolated nail LP. However, there has been one case of AA with isolated nail LP and another with AA and both cutaneous and nail LP [6-7]. Two cases have been reported with cutaneous LP
arising with a patch of AA and many cases of comorbid AA and cutaneous LP alone have been reported, often in the setting of other autoimmune phenomena [8-14]. In fact, one study reports that an individual with AA has a statistically significant increased risk of developing LP, with a relative risk of 2.7 [15]. On the other hand, hair loss in the patient with cutaneous and/or nail LP is not always secondary to AA but instead may represent another variant of LP, namely lichen planopilaris [16].

This association between AA and LP is most likely secondary to immune dysregulation, with both being linked to an autoimmune pathogenesis [17-18]. The commonality in the autoimmunity between these two entities is that both are driven by CD8+ lymphocytes [8]. Nevertheless, why this dysregulation may become isolated to distinct sites, such as the hair and nails, while not affecting visceral organs or even other cutaneous structures is yet to be fully understood. What is clear, however, is that two seemingly related clinical ailments in one patient might represent distinct autoimmune diseases, even when both of these conditions are uncommon.

References