Annular Atrophic Lichen Planus Responds to Hydroxychloroquine and Acitretin

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PRACTICE **POINTS**

- Annular atrophic lichen planus (AALP) is a rare variant of lichen planus, presenting with distinct atrophic plaques with elevated borders on the trunk and extremities.
- Oral prednisone halts disease progression but does not improve existing lesions.
- Hydroxychloroquine and acitretin may prove to be beneficial systemic therapy in the treatment of resistant AALP.

Annular atrophic lichen planus (AALP) is a rare variant of lichen planus. The clinical presentation of AALP shows distinct atrophic plaques with elevated borders on the trunk and extremities. Histopathologic findings generally reveal a lichenoid dermatitis in active lesions with a distinct loss of elastic fibers in the center of the lesions. We report a unique case of AALP, which highlights the chronicity of the eruption. Our patient showed early signs of improvement with hydroxychloroquine and acitretin, suggesting a role for systemic therapy in the treatment of AALP.

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Annular atrophic lichen planus (AALP) is a rare variant of lichen planus that was first described by Friedman and Hashimoto[1] in 1991. Clinically, it combines the configuration and morphological features of both annular and atrophic lichen planus. It is a rare entity. We report a case of AALP in a 69-year-old black man. The clinical and histopathological presentation depicted the defining features of this entity with a characteristic loss of elastic fibers corresponding to central atrophy of active lesions.

Case Report

A 69-year-old black man with a history of hepatitis C virus infection and hypothyroidism presented to the dermatology clinic with a pruritic rash on the trunk, extremities, groin, and scalp of 4 months' duration. He denied any new medications, recent illnesses, or sick contacts. Physical examination demonstrated well-demarcated violaceous papules and plaques on the trunk, extensor aspect of the forearms, and thighs involving 10% of the body surface area (Figure 1A). The lesions were annular with raised borders and central depigmented atrophic scarring (Figure 1B). The examination also revealed several large hypopigmented atrophic patches and plaques in the right inguinal region and on the dorsal aspect of the penile shaft and buttocks as well as a single atrophic plaque on the scalp. No oral lesions were seen. An initial punch biopsy was consistent with a nonspecific lichenoid dermatitis (Figure 2), and the patient was prescribed triamcinolone ointment 0.1% for the trunk and extremities and tacrolimus ointment 0.1% for the groin and genital region.

The patient continued to develop new annular atrophic skin lesions over the next several months. Repeat punch biopsies of lesional and uninvolved perilesional skin from the trunk were obtained for histopathologic confirmation and special staining. Lichenoid dermatitis again was noted on the lesional biopsy, and no notable histopathologic changes were observed on the

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The authors report no conflict of interest.

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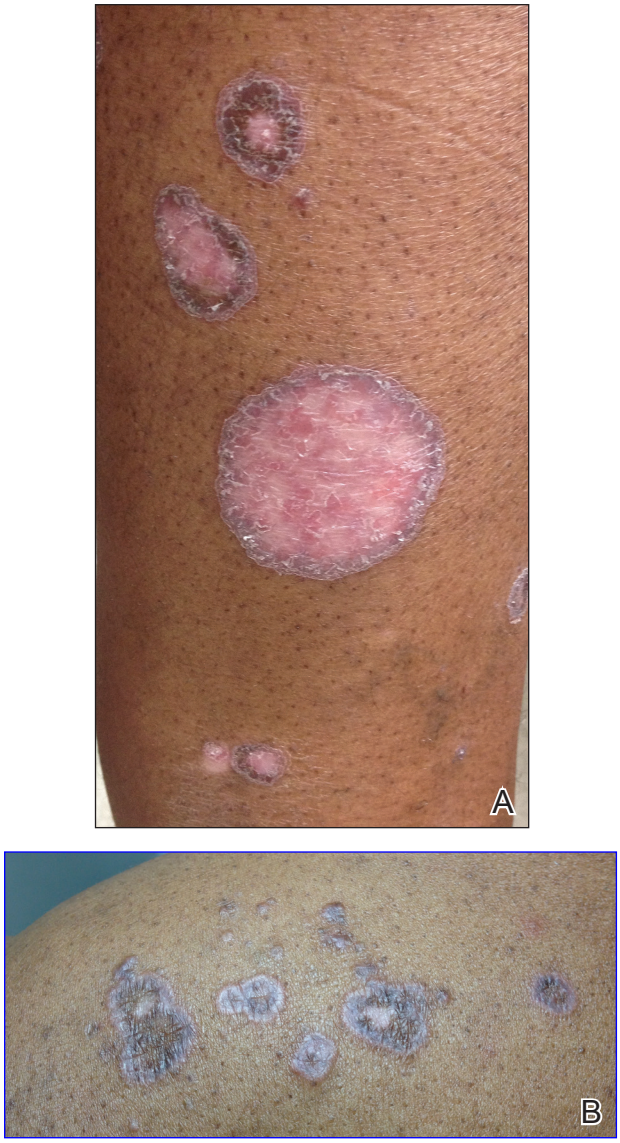

FIGURE 1. Annular atrophic lichen planus with well-demarcated, hyperpigmented, violaceous annular plaques on the left posterior thigh (A) and posterior trunk (B) with elevated borders and central depigmented atrophic scarring.

perilesional biopsy. Verhoeff-van Gieson staining for elastic fibers was performed on both biopsies, which revealed destruction of elastic fibers in the central papillary dermis and upper reticular dermis of the lesional biopsy (Figure 3A). The elastic fibers on the perilesional biopsy were preserved (Figure 3B).

The clinical presentation and histopathological findings confirmed a diagnosis of AALP. The patient was prescribed a short taper of oral prednisone, which halted further disease progression. The patient was then started on pentoxifylline and continued on tacrolimus ointment 0.1% with minimal improvement in existing lesions. These medications were discontinued after

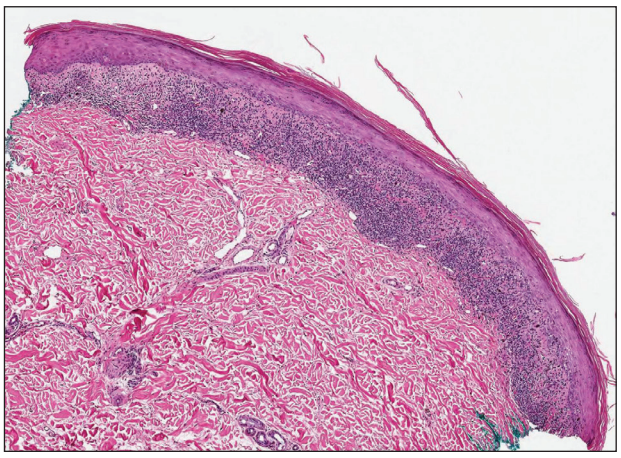

FIGURE 2. Punch biopsy from the left posterior thigh showed compact hyperkeratosis, hypergranulosis, Civatte bodies, and focal acanthosis with an underlying lichenoid dermatitis composed of a lymphohistiocytic infiltrate partially obscuring the dermoepidermal junction (H&E, original magnification ×40).

3 months. Hydroxychloroquine 400 mg once daily was administered, which initially resulted in some thinning of the plaques on the trunk; however, further progression of the disease was noted after 3 months. Acitretin 25 mg once daily was added to his treatment regimen. Marked thinning of active lesions, hyperpigmentation, and residual scarring was noted after 2 months of combined therapy with acitretin and hydroxychloroquine (Figure 4), with continued improvement appreciable several months later.

Comment

Lichen planus is a common pruritic inflammatory disease of the skin, mucous membranes, hair follicles, and nails with a highly variable clinical pattern and disease course that typically affects the adult population.[2] There are many clinical variants of lichen planus, which all demonstrate lichenoid dermatitis on histology. Annular lichen planus is an uncommon variant most commonly seen in men with asymptomatic lesions involving the axillae and groin.[2] Atrophic lichen planus is another variant demonstrating atrophic papules and plaques on the trunk and extremities.[3] Annular atrophic lichen planus is the rarest variant of lichen planus, incorporating features of both annular and atrophic lichen planus.

The first case of AALP involved a 56-year-old black man with a 25-year history of annular atrophic papules and plaques on the trunk and extremities.[1] The second case reported by Requena et al[4] in 1994 described a 65-year-old woman with characteristic lesions on the right elbow and left knee. Lipsker et al[5] reported a third case in a 41-year-old man with a history of Sneddon syndrome who had lesions typical for AALP for 20 years. In all of these cases, histopathologic examination revealed

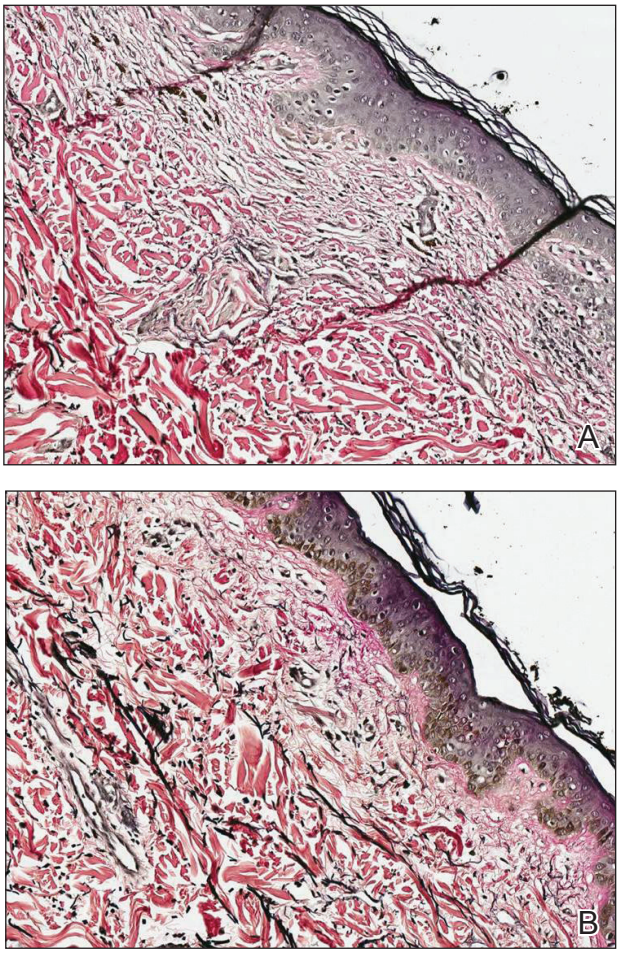

FIGURE 3. Verhoeff-van Gieson staining performed on a punch biopsy of a lesion from the upper back showed a decreased amount of elastic fibers in the dermis (A)(original magnification ×200). Punch biopsy of clinically unaffected adjacent skin was unremarkable with preservation of elastic fibers (B)(original magnification ×200).

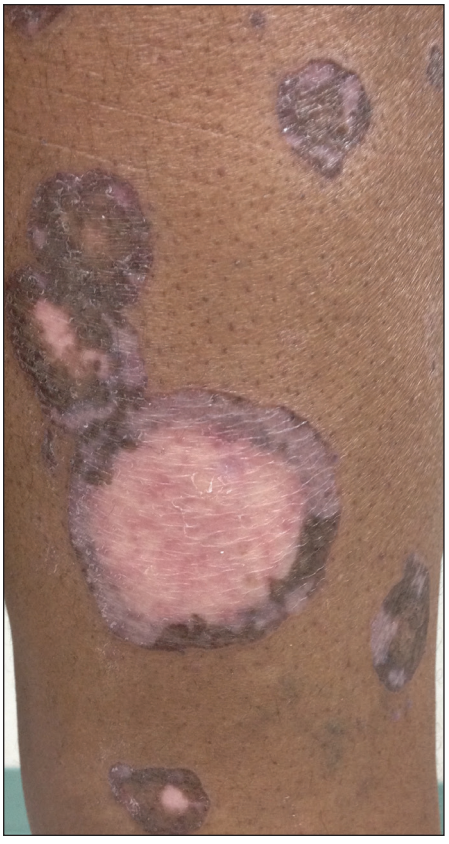

FIGURE 4. Annular atrophic lichen planus with lesional thinning, hyperpigmentation, and residual scarring after 2 months of combined treatment with hydroxychloroquine 400 mg and acitretin 25 mg both once daily.

a lichenoid infiltrate with thinning of the epidermis and loss of elastic fibers in the center of the active lesions.

In more recent cases of AALP, the characteristic findings primarily occurred on the trunk and extremities.[6-10] Treatment with topical corticosteroids failed in most cases and some patients noted moderate improvement with tacrolimus ointment 0.1%. Sugashima and Yamamoto[11] reported a unique case in 2012 of a 32-year-old woman with AALP on the lower lip. She had notable improvement with tacrolimus ointment 0.1% after 6 months.[11]

All of the known cases of AALP to date have occurred in adults, both male and female, presenting with a limited number of annular plaques with slightly elevated borders and depressed atrophic centers.[1,3-11] Disease duration of AALP has ranged from 2 months to 25 years.[11] Histopathologic findings characteristically demonstrate a lichenoid dermatitis of the raised lesional border with a flattened epidermis, loss of rete ridges, and fibrosis of dermal papillae in the lesion center.[7] The elastic fibers are destroyed in the papillary dermis of the lesion center, presumably due to elastolytic activity of inflammatory cells.[1] Macrophages present in the lichenoid infiltrate of acute lesions release elastases contributing to this destruction.[7] Furthermore, elastic fibers appear fragmented on electron microscopy.[1]

The clinical course of AALP has proven to be chronic in most cases and frequently is resistant to treatment with topical corticosteroids, retinoids, phototherapy, and immunosuppressive agents.[3] Treatment administered early in the disease course may provide a more favorable outcome.[11] Lesions characteristically heal with scarring and hyperpigmentation. Our case displayed more extensive involvement than has previously been reported. Our patient showed minimal improvement with topical therapy; however, he demonstrated thinning and regression of active lesions after 2 months of combined treatment with hydroxychloroquine and acitretin. Our use of oral pentoxifylline, hydroxychloroquine, and acitretin has not been previously reported in the other cases of AALP we reviewed. Acitretin is the only systemic agent for lichen planus that has achieved level A evidence, as it previously was shown to be highly effective in a placebo-controlled, double-blind study of 65 patients.[12]

Conclusion

Annular atrophic lichen planus is a known variant of lichen planus characterized by a loss of elastic fibers in the papillary dermis in the center of active lesions. Treatment with topical corticosteroids and phototherapy frequently is ineffective. To our knowledge, there are no studies to date regarding the efficacy of systemic therapy in treatment of AALP. Hydroxychloroquine and acitretin may prove to be beneficial treatment options for resistant AALP. Additional alternative treatments continue to be explored. We encourage reporting additional cases of AALP to further characterize its clinical presentation and response to treatments.

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