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Case report

Pigmented actinic lichen planus: a case report

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Abstract

Actinic lichen planus (ALP) is a rare photosensitive subtype of lichen planus (LP) with four major forms recognized: annular, pigmented (melasma-like), dyschromic, and classic lichenoid. The prevalence is highest among dark-skinned younger females residing in tropical and subtropical regions. There are very few reports of ALP across Europe, with most of the cases among individuals living in warm countries or in people of Middle Eastern and Indian ancestry. We report a case of a 68-year-old white man that presented with a 9-year history of a mildly pruritic solitary hyperpigmented patch on the tip of his nose. Histopathological examination demonstrated signs of classic LP with epidermal atrophy, pigmentary incontinence, and signs of solar elastosis. Based on these findings, a diagnosis of pigmented ALP was established. Topical pimecrolimus and tretinoin along with rigorous photoprotection proved effective, with mild residual hyperpigmentation after 6 months of treatment. Many differential diagnostic possibilities should be considered for such a lesion. Nevertheless, a biopsy and correlation of histopathological and clinical findings can shorten the time from onset to a proper diagnosis. Treating both the hyperpigmented and inflammatory component of this dermatosis is necessary, as well as strict long-term photoprotection to prevent recurrences.

Keywords: actinic lichen planus, pigmented lesion, photosensitivity, clinicopathological correlation, topical therapy

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Introduction

Actinic lichen planus (ALP) is a rare photo-distributed subtype of lichen planus (LP) with an incidence estimated at one case per million inhabitants per year (1). Over the years, the condition has been described under various names, including *lichen planus tropicus*, *lichen planus subtropicus*, *summertime actinic lichenoid eruption*, and *lichenoid melanodermatitis* (2). Katzenellenbogen was the first to use the term *lichen planus actinicus,* emphasizing the role of UV radiation as the main contributing factor (3). Although the clinical appearance varies from annular plaques and melasma-like patches to classic lichenoid papules (4), the histological findings of ALP almost always exhibit the typical characteristics of classic LP (5, 6). The majority of cases are reported in young adult females with darker skin (Fitzpatrick III and IV) of Middle Eastern or Indian origin (5–8). Since the first reports in the 1960s, very few cases have been reported across European countries (9). The extremely low incidence among whites, even in sunny countries, supports the potential role of genetics as another contributing factor (10). Clinical and histological presentation of ALP can resemble other photodermatoses, drug eruptions, and granulomatous diseases (8, 11–13). Due to an uncertain etiology and the rarity of the disease, there are no treatment guidelines and there is no consensus on appropriate topical and/or systemic therapy. Various strategies have been reported, but hydroxychloroquine with intralesional or topical glucocorticoids and sunscreens have shown the greatest efficacy (1, 12, 14).

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In settings of rare occurrence, with a broad spectrum of clinical presentations and no clear guidelines, and with many cases refractory to conventional therapy, ALP may present a significant diagnostic and therapeutic challenge.

Case report

A 68-year-old man with Fitzpatrick skin type II presented to our clinic with a 9-year history of a mildly pruritic hyperpigmented patch on the tip of his nose. He stated that since the onset the lesion had gradually increased in size and become more pigmented. Over the years, the patient observed a pattern of slight regression (lightening) during the winter months and worsening (lesion darkening) through the summer months. He denied using any drugs or supplements before the onset of the disease. He started taking tamsulosin, a treatment for benign prostatic hyperplasia, four years after the lesion began. He had no other significant medical history, and he denied using any other systemic medication or topical application of any creams or ointments.

Clinical examination revealed a slightly elevated irregular-shaped violaceous-brown hyperpigmented patch on the tip of the nose with discrete central atrophy (Fig. 1). The mucous membranes and nails were unaffected. On dermoscopic examination, structureless, irregular brownish-gray pigmentation was present, more prominent around the follicular openings, with diffuse peppering and discrete follicular plugging. At the first visit, the patient rejected a skin biopsy, and he was therefore evaluated clinically and with a detailed history for potential fixed drug eruption and sarcoidosis. Angiotensin-converting enzyme and chitotriosidase serum levels were normal, and X-rays were unremarkable. Because all these findings and history were inconclusive, the patient agreed to a skin biopsy.

A punch biopsy taken from the lesion showed atrophy of the epidermis with mild hyperkeratosis, focal hypergranulosis, and basal vacuolar degeneration. In the dermis, there was scattered superficial, perivascular and perifollicular lymphocytic infiltrate with marked pigmentary incontinence, rare melanophages, and prominent signs of solar elastosis (Fig. 2).

Based on the clinical presentation, seasonal variations, and histopathological findings, a diagnosis of pigmented ALP was made. The patient was treated with topical 0.1% mometasone furoate ointment twice daily for 2 weeks, followed by 1% pimecrolimus cream and 0.05% tretinoin cream applied in the morning and evening, respectively. The patient was instructed to use topical sunscreens and to strictly avoid sun exposure. Three months after starting the treatment, there was significant improvement in terms of infiltration and pigmentation (Fig. 3A). At the last visit, 6 months after starting the treatment, mild hyperpigmentation persisted (Fig. 3B). Given that our patient responded positively to the prescribed treatment combination, with regression of the infiltration as well as less hyperpigmentation, we continued the therapy with rigorous photoprotection.

Discussion

We report a case of the pigmented form of ALP, a rare photosensitive subtype of LP, presenting as a slightly indurated violaceus patch on the nose of a 68-year-old male with Fitzpatrick II skin type. The condition is highly unusual among whites residing in central and northern parts of Europe. Diagnosing ALP can be challenging, given that there are no other clinical signs and symptoms indicative of lichen or lichenoid dermatoses (4). Because the pathogenesis of the onset and relapses of ALP is mostly unknown, there is still no consensus on the treatment approach.

The first reports suggested that the condition is restricted to tropical and subtropical regions and that it predominantly manifests in younger individuals with darker skin types (7). Racial predilection to people of Middle Eastern and Indian origin, rather than geographical distribution, favors a genetic basis of the condition (5, 15). However, UV radiation from sunlight and lamps seems to be the main trigger in inducing the lesions (9). UV radiation provokes the expression of altered self-antigens on basal keratinocytes that are responsible for the recruitment of cytotoxic T-cells, resulting in characteristic microscopic features such as basal cell vacuolization (14). Supporting the hypothesis of ALP being a photosensitive reaction in genetically

predisposed individuals is a case of actinic LP following the lines of Blaschko (15). The concept of genetic mosaicism in the acquired Blaschko-linear inflammatory dermatoses implies that different genetic compositions within skin cells may result in the presence of various antigens on them. This antigenic mosaicism could then activate a mosaic T-cell response to external triggers such as sunlight (16).

Accordingly, the lesions of actinic LP mainly develop on sun-exposed areas of the face, neck, and forearms, on the dorsa of the hands, and on the shins (4, 17). Rarely, unexposed skin areas and buccal mucosa can be involved (6). Most patients report onset in spring or summer, and improvement or even complete remission in winter (18). Four morphological patterns of actinic LP have been clinically described in the literature: annular, pigmented (melasma-like type), dyschromic, and classic plaque-like type (1). Our case was compatible with the pigmented type of actinic LP based on the clinical appearance of the lesion. The pigmented form, also called the melasma-like type, is characterized by gray to brown or black patches ranging in size between 0.5 and 5 cm (13) located on the face and neck, with the lateral sides of the forehead being the most common site of presentation (1, 6, 18). Unlike in classic lichen planus, the Koebner phenomenon, scaling, and mucosal or nail involvement are not common in the actinic variant. Pruritus is minimal or absent (1). Our patient presented with a solitary violaceous-brown hyperpigmented patch on the tip of the nose, a site rather uncommon for ALP, which led us to consider cutaneous sarcoidosis, fixed drug eruption, and discoid lupus erythematosus, and differential diagnoses of actinic LP among melasma, lentigo maligna, granuloma annulare, polymorphous light eruption, pigmented actinic keratosis, and erythema dyschromicum perstans (1, 12–14). A detailed medication history and seasonal appearance of the lesion excluded the possibility of drug-associated eruptions (e.g., amiodarone-induced nose pigmentation).

Although dermoscopy might help differentiate actinic LP from other pigmented and nonpigmented lesions, its use is limited because similar patterns are observed in other acquired hypermelanoses. Data on dermoscopic findings in actinic LP are scarce, but so far peripheral or diffuse dots and globules with peppering and perifollicular, annular, linear, or homogeneous cloud-like pigment patterns have been reported (13, 19, 20). Gungor et al. (19) demonstrated that all 10 ALP lesions analyzed in their study had diffuse peppering arising on a brown background with absent Wickham striae and a vascular pattern, a finding consistent with our case. However, there is a significant overlap of dermatoscopic patterns in pigmented actinic LP, melasma, pigmented actinic keratosis, erythema dyschromicum perstans, erythromelanosis follicularis faciei et colli, and lentigo maligna (20). For instance, shared dermatoscopic characteristics of pigmented ALP and pigmented AK comprise linear, annular, and granular structures, characterized by the coalescence of grayish-brown dots and globules around the hair follicles; these have been shown histologically to be due to the presence of aggregates of melanin and macrophages in the papillary dermis. Another common shared dermatoscopic finding is a subtle homogeneous gray or beige halo that surrounds the follicular openings or follicular plugs, also known as inner gray halo. The histologic equivalent of this feature is the inverted cone of “spared” orthokeratotic epidermis, which tends to spread around the follicular openings on the surface of the epidermis. When in doubt, clinicians should seek additional and specific patterns so far described in some of the differential diagnoses. In the aforementioned example, an additional finding present in pigmented AK, but absent in pigmented ALP, is rosettes arranged in a four-leaf clover shape, mainly localized inside the follicular openings, better visible in polarized contact dermoscopy (21). Although more research is required, the use of dermoscopy for this group of disorders appears promising and may act as a supplementary tool. Nevertheless, histologic examination remains the gold standard. The histologic features of actinic LP are quite diverse, but mostly similar to those of classic lichen planus: hyperkeratosis, hypergranulosis, basal vacuolar degeneration, and Civatte bodies are usually present. At the dermal level, there are a bandlike lymphocytic inflammatory infiltrate in the superficial dermis, melanophages, and marked pigmentary incontinence (1). Furthermore, the pigmented form of ALP is often associated with signs of actinic damage in the skin such as epidermal atrophy and solar elastosis in the dermis (8, 13). The punch-biopsy specimens taken from our patient demonstrated significant epidermal thinning with loss of the normal rete ridge pattern with hyperkeratosis, focal hypergranulosis, and basal vacuolar degeneration. In the dermis, there was sparse subepidermal lymphocyte infiltration and prominent solar elastosis. In addition, perifollicular and periadnexal inflammatory infiltrate was observed, a feature more often visible in cutaneous lupus erythematosus. However, pathological studies by Kamyab et al. have suggested that this finding is not as rare in patients with actinic LP, reporting it in about

25% of cases (6). The variability of histological presentations among reports is explained by the difference in the disease duration and biopsy site (4).

Several treatment strategies have been reported, but there are no clear guidelines in the management. All patients should be advised to use high-SPF sunscreens or sunblocks and avoid sun exposure because there is a clear relationship to UV radiation inducing the lesions and relapses, and a few reports also claim spontaneous remission or regression during the winter months (2). Intralesional and topical corticosteroids are the first-line treatment, resulting in almost complete resolution of lesions, with their only limitation being skin atrophy induced by prolonged use (13, 18). Calcineurin inhibitors act by decreasing the aforementioned T-lymphocyte activation and are safe for long-term use. Topical pimecrolimus and tacrolimus have resulted in major improvement and no recurrence of the lesions, as reported in several articles (15, 22, 23). In refractory and rapidly progressive cases, antimalarials (1), cyclosporine (24), and systemic corticosteroids (23) have been used as effective treatment options. Although acitretin was proved to be a successful treatment for classic LP, its role in actinic LP is not completely clear due to variable results (1, 17, 18).

Residual hyperpigmentation, as seen in our patient, is very common (25) and could be associated with the gray and blue hue seen on dermoscopy. These are signs of deeper deposition of pigment in the dermis, which is typically more persistent and less responsive to treatment (20). After the initial short-term treatment with a high-potency topical corticosteroid, our patient was prescribed pimecrolimus cream due to its action on the inflammatory component of the disease. Topical tretinoin was added to combat the residual hyperpigmentation and signs of chronic actinic damage.

Conclusions

To the best of our knowledge, this is the first reported case of ALP in this part of Europe. Genetic susceptibility could be the reason behind this rare type of photosensitivity in our patient. In addition to strict photoprotection, due to the possible role of T-lymphocytes in the pathogenesis of ALP, we opted for long-term treatment with topical calcineurin inhibitor combined with topical retinoid to address hyperpigmentation and signs of chronic actinic damage.

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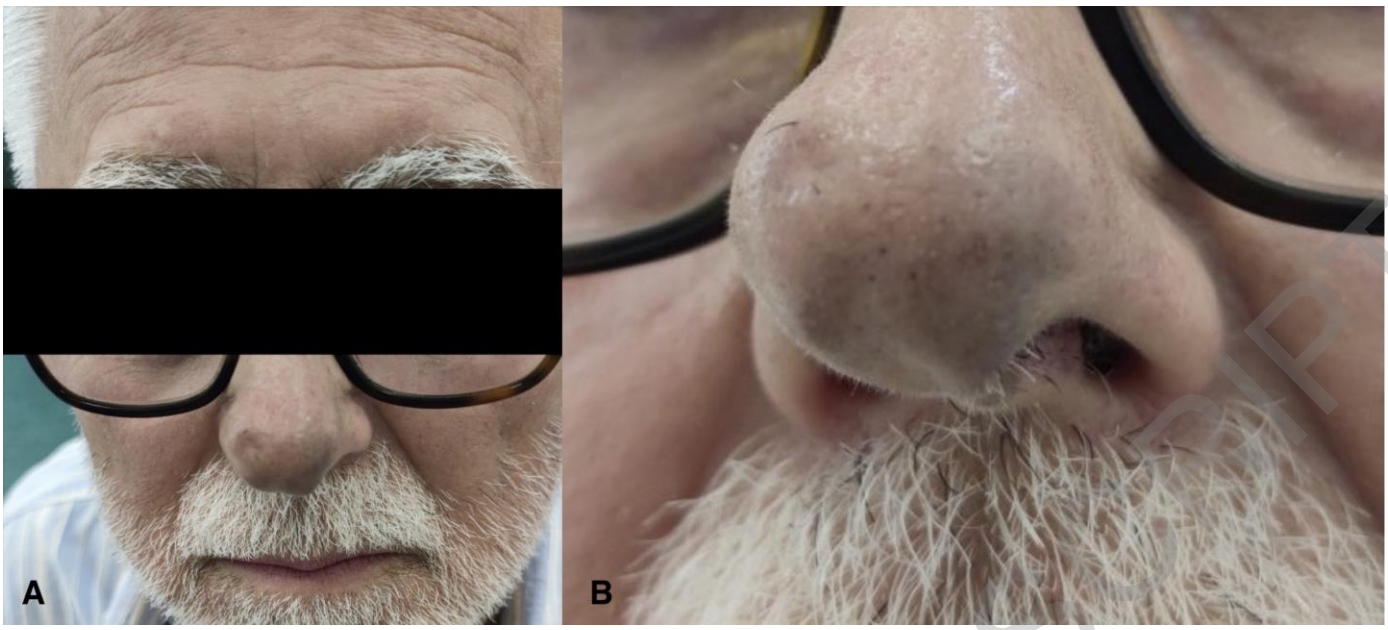

Figure 1. A) Slightly infiltrated irregularly shaped violaceous-brown hyperpigmented patch on the tip of the nose with discrete central atrophy; B) close-up view of the lesion.

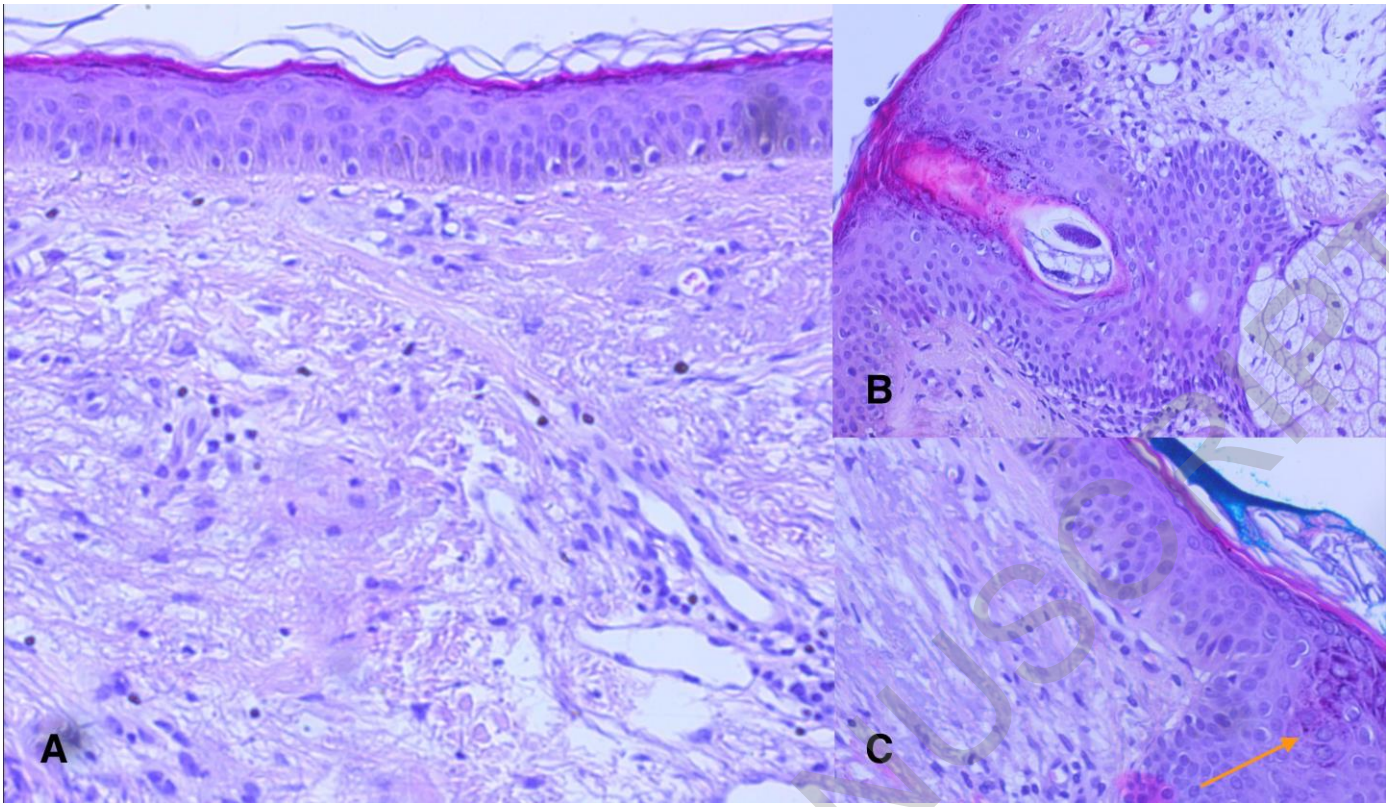

Figure 2. A) Atrophy of the epidermis with basal vacuolar degeneration; in the dermis, small melanocytic cells together with melanophages and papillary and mid-dermal perivascular inflammation; chronic actinic damage (solar elastosis) was also found (hematoxylin and eosin, original magnification ×40); B) inflammatory infiltrate, composed of lymphocytes and histiocytes in perifollicular areas (hematoxylin and eosin, original magnification ×40); C) focal hypergranulosis (arrow; hematoxylin and eosin, original magnification ×40).

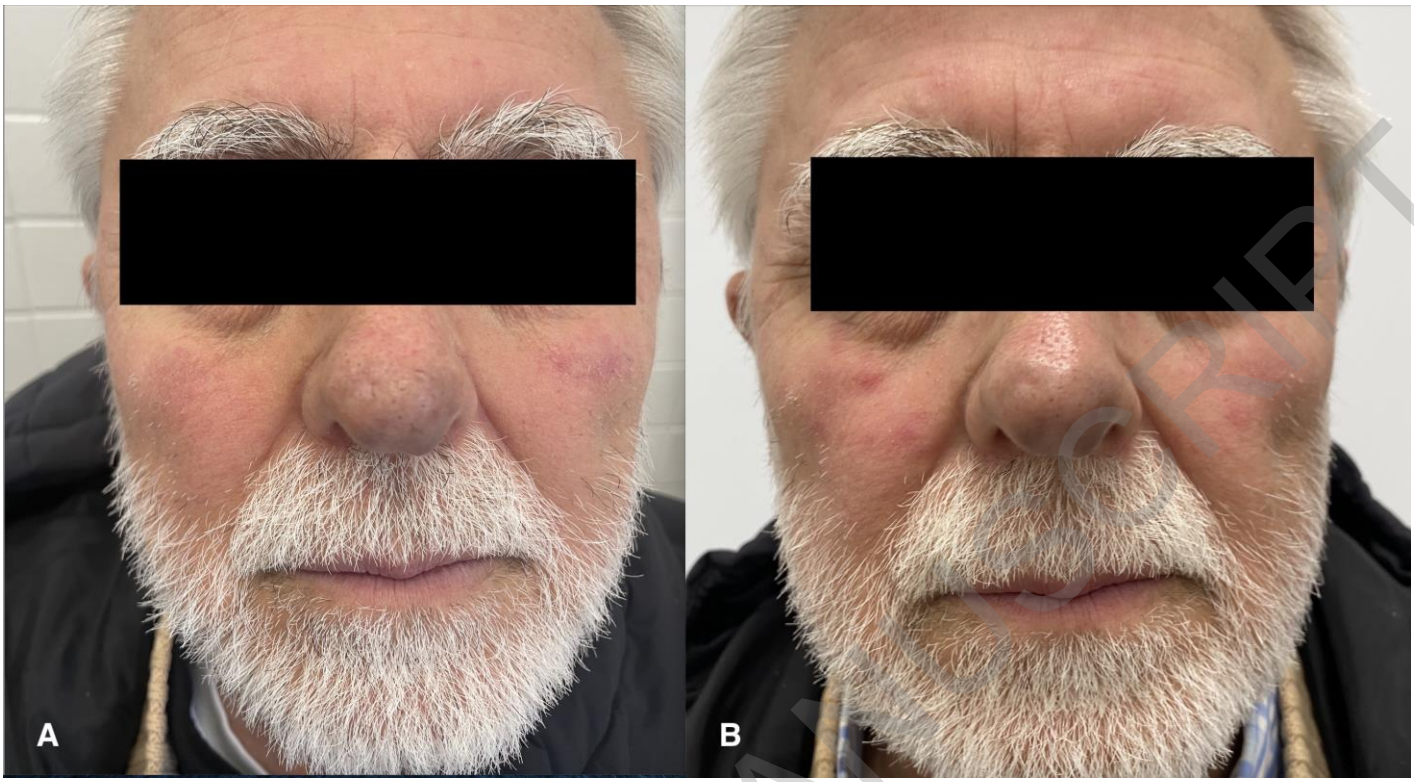

Figure 3. A) Mild residual pigmentation after 3 months of therapy; B) after 6 months of therapy.