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Case Presentation

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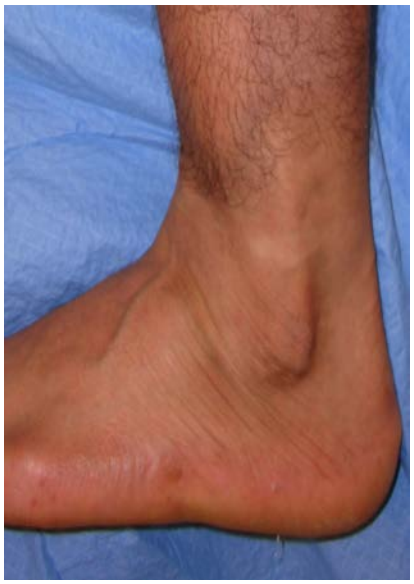
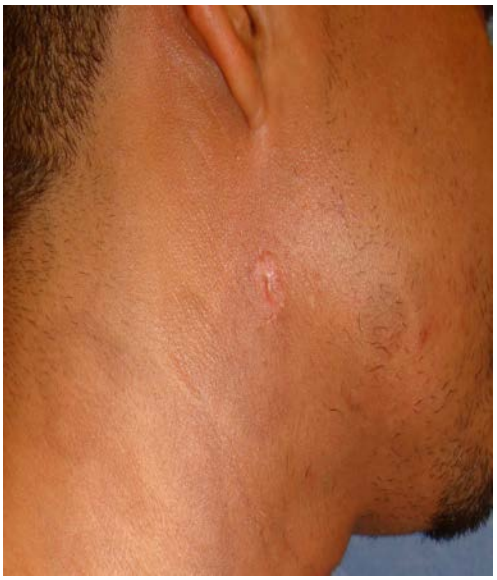
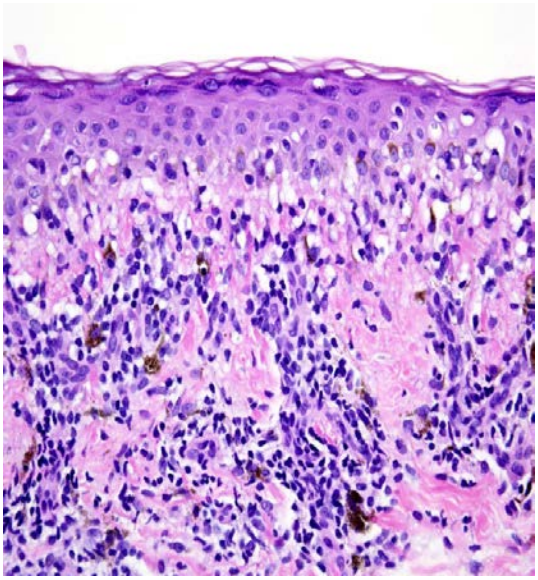
Abstract

Lichen planus pigmentosus (LPP) is an uncommon variant of lichen planus that tends to occur in middle-aged individuals with darker pigmented skin. Clinical findings include hyperpigmented, brown to gray-brown macules and patches in sun-exposed areas, typically on the head and neck. Histopathologic features include epidermal atrophy, vacuolar degeneration of the basal layer of the epidermis, perivascular lymphohistiocystic infiltrate in the upper dermis, and dermal melanophages. We present a unique case of LPP that was characterized by an atypical initial inflammatory phase and subsequent circinate lesions with central clearing.







Case synopsis

A 31-year-old man presented to the Dermatology Clinic in Bellevue Hospital Center in August 2011 with a two-year history of dark areas on the skin of the face, neck, trunk, and extremities. He had no local or systemic symptoms. At the time of examination, he was assured that there were no active lesions and he was subsequently lost to follow-up. In March 2012, he presented with new macules on his face and neck, which had evolved over the course of one week and were asymptomatic without pruritus, burning, or pain. He had used oral fluconazole and topical selenium sulfide without benefit. He denied fevers, chills, diaphoresis, sun sensitivity, exposure to pesticides, contrast dye and ionizing radiation. He denied taking any medications. He specifically denied taking diuretics or nonsteroidal anti-inflammatory drugs. Past medical history included a resolved gastric ulcer.

Physical Examination: Gray-brown, annular, circular and oval, hyperpigmented patches were present on the face, some with a thin, raised, erythematous border. There were reticulated blue-grey patches without erythema on the arms, trunk, and legs. On the buccal mucosa, there was a faint, lacy hypopigmented patch.

Laboratory Data: A complete blood count and basic metabolic panels were normal. Alanine aminotransferase was 37 U/L. Hepatitis B and C serologies were non-reactive. Cholesterol was 207 mg/dL.

Histopathology: There is a superficial, perivascular and lichenoid, lymphocytic infiltrate that extends to the dermoepidermal junction where there are vacuolar changes and necrotic keratinocytes. Numerous melanophages are noted within the papillary dermis.

Diagnosis: Lichen planus pigmentosus.

Discussion: Lichen planus pigmentosus (LPP) is a rare variant of lichen planus that tends to occur in middle-aged individuals with darker pigmented skin. LPP was first reported in a series of Indian patients in 1974 and has most commonly been described in patients from India, the Middle East, and Latin America [1-4]. Clinical findings include brown to gray-brown macules and patches on sun-exposed areas and, in the rare inversus variant, on the flexural areas and intertriginous zones [5]. Lesions are commonly observed on the forehead, temples, and neck; they are largely distributed symmetrically. The pattern of pigmentation is generally diffuse, although follicular, reticular, and unilateral linear variants have been described. Cases of the linear variant have been reported mainly from Japan and Korea and may occur within Blaschko lines [1, 6-9]. Although there are few reports of annular LPP, annular lesions have been present in a number of published photographs and are common in both the inversus and actinic forms of the disease [10]. Some cases of LPP also have shown gyrate patterns [11]. In contrast to classic LP, the lesions of LPP are flat or minimally raised, are never hyperkeratotic, and are rarely symptomatic. Although LPP may occur concurrently with other forms of lichen planus, few studies have examined their co-occurrence. One retrospective series reported that 19 of 124 patient with LPP also had typical lesions of lichen planus [2]. Other conditions associated with LPP have included hepatitis C infection.

In contrast to the classic lesions of lichen planus, the lesions of LPP usually are asymptomatic, although some lesions may be associated with pruritus or a burning sensation [2]. The disease course is chronic, with exacerbations and remissions. Histopathologic features of LPP include epidermal atrophy, vacuolar degeneration of the basal layer of the epidermis, a perivascular lymphohistiocystic infiltrate in the upper dermis, and dermal melanophages [12]. The etiology is unknown although ultraviolet light has been suggested to play a role in patients with photodistributed eruptions. Several retrospective studies report associations between LPP and topical applications of mustard and amla oils [2, 3]. These oils are used for body massage, hair dressing, cooking, and the preparation of local medicines in India. Mustard oil contains allyl thiocynate, which is a potential photosensitizer and a potential pathogenetic agent in LPP [2].

Although LPP is considered to be a variant of lichen planus, the main entity in the differential diagnosis is erythema dyschromium perstans (EDP) or ashy dermatosis. LPP and EDP share many clinical similarities. Both conditions are characterized by gray-to-brown macules and patches and were only described as distinct clinical and histopathologic entities in 1992 [12]. Clinically, early EDP lesions often have an inflammatory phase, in which a thin ring of erythema surrounds the hyperpigmented macules and patches. Such an inflammatory phase is not typically observed in LPP [4]. Although erythema in the new lesions of our patient was most prominent around the border, the lesions did not have the characteristic, evanescent rim associated with EDP. Our patient's lesions evolved into circinate, confluent , hyperpigmented patches with central areas of clearing. Such findings are atypical for EDP.

A histopathologic feature that distinguishes LPP from EDP is the location of melanin deposition. In EDP, melanin deposition occurs in the deeper dermis, thus producing blue-grey colored lesions from the Tyndall effect; melanin deposits in LLP are located in the superficial dermis [10]. Our patient's histopathologic features showed melanophages in the superficial dermis as well as clinical lesions in sun-exposed areas. Other entitities in the differential diagnosis of LPP include lichen planus actinicus, lichen planus inversus, and lichenoid drug eruption. Although our patient's lesions did feature central clearing, they were without the annular plaque morphology that typically is associated with lichen planus actinicus. Inverse lichen planus is made less likely because of the sun-exposed location of our patient's lesions. A lichenoid drug eruption is unlikely owing to our patient's lack of drug exposures.

Treatment of LPP can be challenging because the clinical course is inherently variable. Few effective therapies have been reported. Some cases may spontaneously resolve within weeks; other cases may persist for years. Treatment strategies should always include the use of sunscreen and sun avoidance. One open-label, uncontrolled study reported lightening of pigmentation in seven of 13 patients that were treated with 0.1% topical tacrolimus for 12 to16 weeks [1]. However, the remaining six patients received no benefit. It is unknown whether the partially treated subjects ever fully cleared. Another group reported that the combination of 0.1% topical tacrolimus and neodymium:yttrium-aluminum-garnet laser was helpful in a single dark-skinned individual with linear LPP who had not responded to topical glucocorticoids [9]. Another case reported the ineffectiveness of four weeks of twice daily topical tacrolimus 0.2% then two weeks of clobetasol in the treatment of LPP inversus [13].

Owing to the dearth of evidence for the effective treatment of LPP and the evidence for topical glucocorticoids and oral retinoids in the treatment of lichen planus, our patient received acitretin 25 mg daily along with topical mometasone twice daily [14-16]. Acitretin was discontinued after two months owing to transaminitis. Of note, after two months of treatment, the erythema-rimmed

macules and patches that originally had presented on our patient's neck had evolved into grouped circinate patches with central clearing but no erythema. It is unclear whether the evolution of patches reflects effectiveness of treatment or the natural course of disease.

Our patient adds to a very small literature base that describes the presentation and treatment of LPP. This case is notable for the rarely reported presentation of an initial inflammatory phase and evolution into annular lesions. We believe that acitretin is a medication that deserves more investigation in this small and challenging patient population.

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