Volume 21 Number
September 2015

Case presentation

Lichen planus pigmentosus in linear and zosteriform pattern along the lines of Blaschko

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Dermatology Online Journal 21 (10): 11

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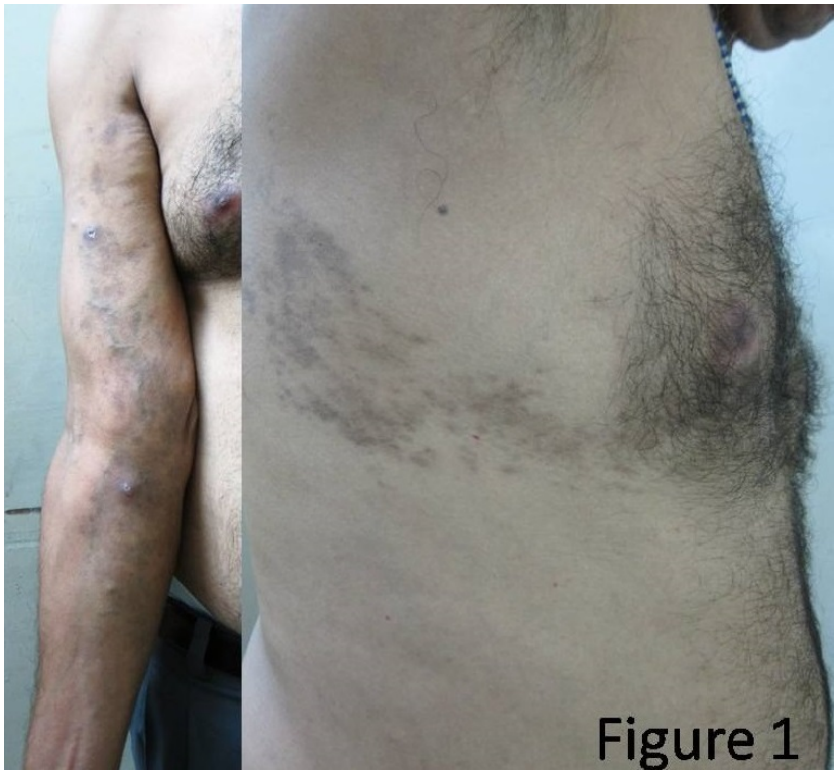
Abstract

Lichen planus pigmentosus (LPP) is a variant of lichen planus (LP) reported in various ethnic groups. It occurs predominantly in the third or fourth decade of life and is characterized by the insidious onset of dark-brown macules in sun exposed areas and flexural folds. Rarely, has it been described in a linear or segmental distribution. Herein we describe a case of LPP with lesions lateralized to right side of body along the lines of Blaschko, in a linear and zosteriform pattern.

Case synopsis

A 48-year-old man presented to us with asymptomatic patches of hyper-pigmentation on the body for 1 year. The pigmentation initially appeared on the volar aspect of his right forearm and gradually these lesions increased in number to involve the right arm, chest, and right lower limb in a discontinuous distribution. The pigmentation was confined to the right side of body and and progressed to its present extent over a period of two months with no change in number or color ever since. At no time during the clinical evolution was the rash palpable. There was no history of any drug intake, topical application, significant sun exposure, or trauma prior to eruption. The patient never had herpes zoster and family history was not contributory.

Hematological and biochemical investigations were within normal limits. Serology for hepatitis B and C virus was negative. On cutaneous examination, uniformly pigmented dark brown-black macules were present in a linear non-contiguous pattern on flexural surfaces of the right forearm and arm and in a zosteriform pattern on the right chest wall. Multiple linear swirls involved the right buttock, thigh, and leg (Figure 1, 2). These macules were 0.2-2 cm in size, irregular in shape, and well circumscribed. His face, mucous membranes, scalp, and nails were spared. Histopathology examination of a hyperpigmented macule on the forearm revealed an unremarkable epidermis, pigment incontinence and melanophages, and scanty perivascular lymphocytic infiltrate in the upper dermis, thus compatible with the diagnosis of LPP (Figure 3). He was started on topical tacrolimus 0.1% to be applied twice daily.



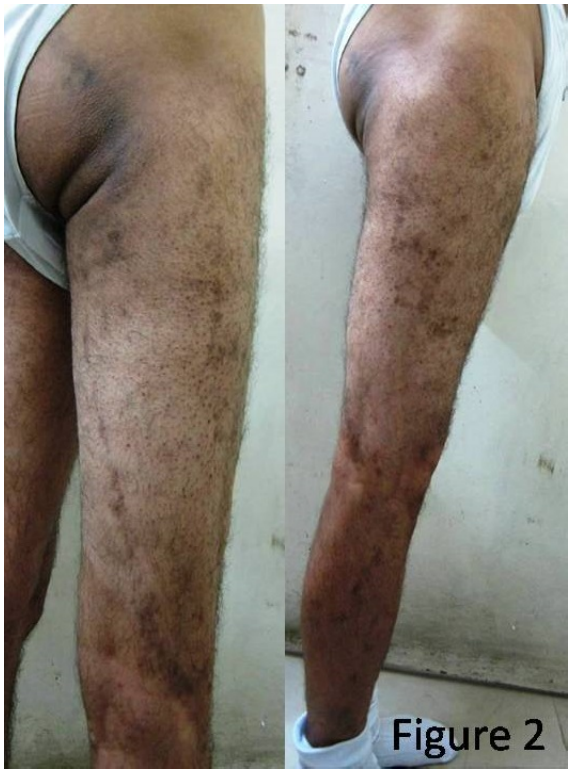


Figure 1 and 2. Caption goes here

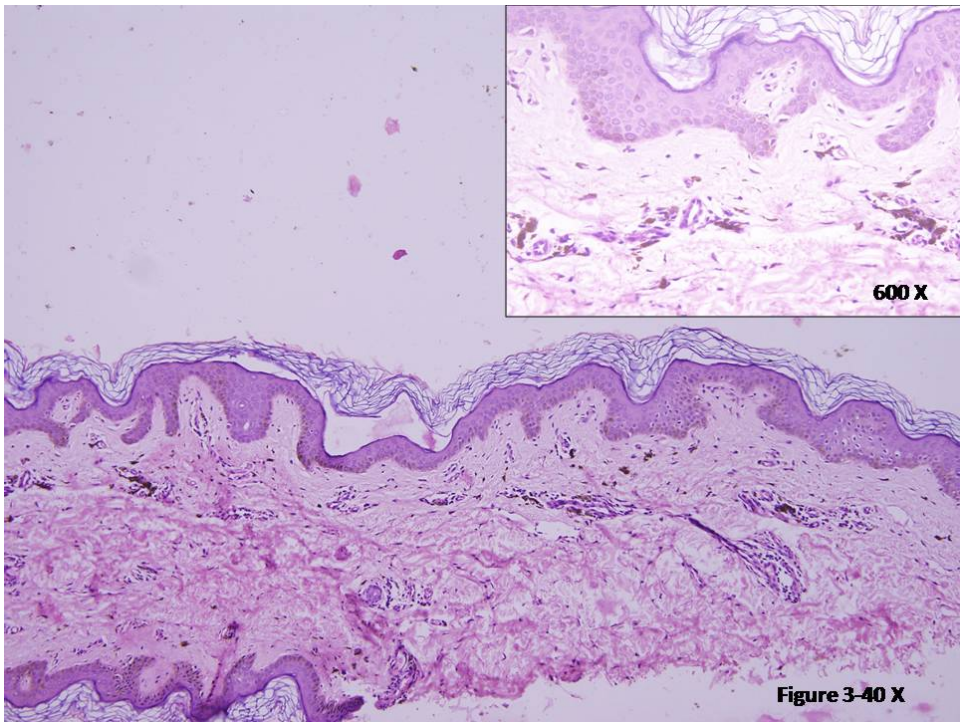


Figure 3. Caption goes here

Discussion

Lichen planus pigmentosus (LPP) is a pigmentary disorder characterized by the insidious onset of bilateral, symmetrically distributed slate grey to brownish black macules in sun-exposed areas and flexural folds, with or without slight pruritus. Diffuse, reticular, blotchy, and perifollicular forms have been reported [1, 2]. The preauricular region, including the temples and forehead, is the initial site of onset and is involved in almost all patients [2]. Although the exact etiopathogenesis is still elusive, mustard oil, amla oil, henna, and hair dye have been postulated to be the precipitating factors in predisposed individuals [2]. Microscopic findings include atrophy of the epidermis, vacuolar degeneration of the basal cell layer, and scarce dermal perivascular or lichenoid infiltrate. The presence of dermal melanophages and pigment incontinence are the two constant features seen on histopathology [3].

LPP in a linear, zosteriform, and segmental arrangement has been previously reported in the literature [4, 5, 6, 7, 8]. Another potential condition in the differential diagnosis for such a presentation in an adult is ‘progressive cribriform and zosteriform hyperpigmentation’ (PCZH). This entity was first described by *Rower et al*, as the presence of uniformly tan cribriform macular pigmentation in a zosteriform distribution, with no evidence of preceding inflammation. It is histologically characterized by basal layer hypermelanosis [9]. Pigmentation in our patient was linear along the limbs and zosteriform on the trunk and histopathology revealed melanophages with pigment incontinence in dermis, thus ruling out PCZH.

Patterns of LPP reported as ‘linear’, ‘zosteriform’ and ‘segmental’ [4, 5, 6] actually reflected the distribution of lesions along Blaschko lines and these terms should not be used in that context. These lines have frequently been confused with dermatomes, as both the distribution patterns are characterized by a striking demarcation of cutaneous lesions at the midline [6]. Blaschko lines do not relate to any vascular, neural, or lymphatic structures in the skin and differ from dermatomes in being more numerous, whereas dermatomal division refers to the metameric distribution of sensory nerves. Cutaneous areas representing mosaicism, are populated by cells with distinct antigenic and/or immunologic properties and can differ from the rest of the body in susceptibility (locus minoris resistentiae) or resistance (locus maioris resistentiae) to skin disorders. Although inflammatory response in these mosaic conditions is localized to these lines, this phenomenon does not reflect T cell migration during embryogenesis, as has been misunderstood by some authors in the past [5, 6].

Our patient was unique in that he manifested LPP in multiple patterns of cutaneous mosaicism. He had lesions lateralized to the right side of body along the lines of Blaschko, in classic narrow bands along the limbs (type Ia) and broader bands (type Ib) on the trunk [10]. Unilateral, concurrent linear, and zosteriform distribution of lesions has never been described in a case of LPP, to the best of our knowledge. The role of tacrolimus in the treatment of LPP has been explored only recently [3] and since tacrolimus 0.1% is known to be more effective than 0.03%, it is likely that it would be more effective. Our case had an extensive distribution of lesions, in multiple patterns of cutaneous mosaicism. LPP along Blaschko lines is very uncommon and simultaneous incidence of a linear and zosteriform pattern is owing to mosaicism and its impact on phenotype, related to the proportion of cells that harbor the mutation and their tissue distribution.

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