

Lichen Planus Pigmentosus: An Overview

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ABSTRACT

Lichen planus pigmentosus (LPP) is considered a rare variant of lichen planus. It is characterized by acquired dark brown to gray macular pigmentation located on sun-exposed areas and commonly found in dark-skinned patients. Inverted, linear, mucosal and follicular types are identified. The exact etiology is not well known however, LPP has been associated with hepatitis C virus, sun exposure, endocrinopathies, autoimmune diseases and contactants such as mustard oil and nickel. Treatment is difficult and consists mainly of avoidance of triggers and topical tacrolimus.

INTRODUCTION

Lichen planus pigmentosus (LPP) is a rare form of lichen planus (LP). It is characterized by oval or irregularly-shaped brown to gray-brown macules and patches on the skin. Areas that are exposed to sun such as the forehead, temples and neck are most commonly affected. However, the macules and patches may also develop on the trunk or in places where two areas of skin touch or rub together (i.e. the armpit, groin, etc).¹ LPP is a chronic, relapsing condition with periods of worsening symptoms separated by periods of remission (decreasing or disappearing symptoms). The cause of LPP is unknown, but studies suggest it may be triggered by UV light, viral infections, or agents applied to the skin such as mustard oil. Treatment for LPP depends on the symptoms in each person.²⁻⁴

HISTORICAL BACKGROUND

It is possible that LPP had been described in 1935 under the name “lichens atypiques ou invisible pigmentogenes” in the French literature⁵ and as

“lichen pigmentosus” in 1956 in the Japanese literature.⁶ In 1957, during the first Central American Congress of Dermatology, Oswaldo Ramirez described a group of patients with a new dermatosis characterized by “ashen macules” with no apparent cause, which he called ashy dermatosis (AD) “dermatitis cenicienta”.⁷ In 1974 Bhutani *et al.*,⁸ described the clinical and histopathological features in a series of 40 patients with similar pigmentation to that described by Ramirez. Exhibited histopathological findings of these pigmented macules were similar to LP, so, he suggested the name LPP. Vega *et al.*, in 1992⁹ presented LPP and AD as two separate entities although they are considered by many authors a single entity. Kanwar *et al.*, in 2003,¹⁰ in their study of 124 Indian patients, described the clinical, epidemiological, and histopathological features of LPP in Indian patients. They confirmed the coexistence of classical LP lesions with LPP lesions.

The inversus type of LPP was first reported in Europe, followed by reports from Japan.¹¹ Fewer

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reports exist from Africa and India, being common in light-skinned.¹² Both classical and inversus patterns of LPP were later considered as a single entity in view of a common clinical picture of mostly asymptomatic pigmentation with lichenoid histology.¹³

EPIDEMIOLOGY

LPP is a disease of the middle-aged, with onset in the third to fourth decades of life and few studies show a slightly greater incidence in females.¹⁴ LPP predominantly affects patients with darker skin phototypes III to VI, with phototype IV being the most frequently affected. It has been commonly observed in India, Latin America, Asia, and Africa. The inversus type is relatively rare, predominantly occurring in Caucasians.¹⁵

PATHOGENESIS

LPP and LP have a similar immunopathogenesis, represented by an altered cellular immune response mediated by T lymphocytes, in which CD8+ T lymphocytes recognize and attack epidermal keratinocytes, causing intense pigmentary incontinence. Pock *et al.* hypothesized that LPP is considered an abortive variant of LP, in which an intensive lichenoid reaction occurs quickly before the compensatory increased proliferation of keratinocytes develops as in typical LP, due to which there is a rapid transformation of papules into brown macules.¹² The rapid regression of the inflammatory infiltrate causes dermal melanin to remain for months to years.^{12,14}

The lichenoid infiltrate is predominantly composed of CD8+ T lymphocytes, which are responsible for the cytotoxic response against keratinocytes. CD4+ T cells and Langerhans CD1a+ cells can also be found. Keratinocytes

in LPP can focally express HLA DR, similar to those in LP.¹⁶⁻¹⁸

Direct immunofluorescence is positive in few cases. IgM and C3 within the dermoepidermal junction,¹⁰ globular IgM in the papillary dermis,¹⁹ and IgG and IgA in keratinocytes²⁰ have all been documented. In addition, IgM, IgG, IgA, and C3, have been described in cytooid bodies.²¹ These findings support that LPP and LP share similar immunopathogenesis, although this immunopathogenic mechanism might be minor in LPP.¹⁹

The main predisposing factors probably include sun exposure and photosensitizing topical agents specially in the actinic variant. Topical application of mustard oil, which contains allyl isothiocyanate, a potential photosensitizer, and amla oil, has been proposed as possible inciting agents.^{14,22} Koebnerization due to friction in the flexures or due to tight clothing is thought to contribute to the inversus type of LPP. Patch tests have proved positive mainly for nickel.²³ Majima *et al.* reported the disappearance of lesions after the discontinuation of wearing tight underclothes among two Japanese patients.²⁴

There are a few reports of association of hepatitis C virus infection with LPP.^{14,19} In a series of patients in Kuwait, 20 out of 33 patients had a positive hepatitis C virus serology.¹⁹ LPP often presents in women around the time of menopause, suggesting a possible influence of hormonal factors on pathogenesis.¹⁵

CLINICAL PRESENTATION

LPP manifests as pigmentation of insidious onset without any features of inflammation or preceding raised lesions. It is typically asymptomatic although occasionally can have mild pruritus

which is considered a marker of activity and progression. Pruritus is present in the initial and active phase and disappears quickly with or without treatment. Rarely, a burning sensation might be present.¹⁵

LPP is characterized by a symmetrical distribution of dark brown to gray or gray-blue, round or oval macules with irregular and poorly-defined borders, which eventually enlarge and coalesce. Rarely, incipient LPP manifests as entirely erythematous macules which represent an active phase that rapidly resolves.¹

LPP is mainly located on the face and neck. On the face, it has a predilection for the temporal and preauricular areas, whereas on the neck it affects all sides. The arms are more frequently involved than the legs and trunk. Flexural areas are affected in few patients, mainly the axillae, inframammary folds and the inguinal creases. Surprisingly, there is no involvement of scalp, mucosa, or nails.¹⁵

Although the pigmentation is usually diffuse,^{10,14} reticular, blotchy, perifollicular, annular, and gyrate patterns are also encountered. There are isolated case reports of linear unilateral hyperpigmentation in the extremities (Blaschkoid) and segmental patterns on the trunk, all of which may coexist in the same patient. Occasionally, LPP can present concomitantly with LP; therefore, it is not surprising that active or resolving LP lesions be found in patients with LPP.¹⁵ Evolution is chronic and progressive with remissions and exacerbations, but the course and duration are unpredictable.²⁵

CLINICAL VARIANTS

Lichen planus pigmentosus inversus

The term lichen planus pigmentosus inversus

(LPPI) was proposed by Pock *et al* in 2001, after the report of 7 cases of LPP located predominantly on intertriginous areas.¹² In their study, Gaertner and Elstein, in 2012, considered the clinical and histological manifestations of LPPI as distinct from other similar entities, such as LPP and erythema dyschromicum perstans (dermatosis cicatricata or ashy dermatosis).²⁶

It predominantly affects women, mostly after 40 years, but has been described in individuals 15–82 years of age. Few cases were detected in children.^{11,27} LPPI affects fair and dark skin, with a worldwide distribution.^{11,28–30} The characteristic manifestations of LPPI are hyperchromic macules, usually small, lenticular, with mild to absent pruritus, affecting intertriginous areas, mainly axillae. Cervical, inguinal and popliteal areas could be affected. Larger lesions tend to have a linear or angular configuration, with the longitudinal axis following the cleavage lines.^{26,30,31} Neck involvement is problematic in making the diagnosis of LPPI; if extensive with face involvement, LPP should be considered to be the diagnosis, but if neck and face involvement are mild and folds are predominantly affected, the diagnosis is likely LPPI. Although LPPI is usually bilateral in distribution, unilateral distribution is not rare.^{32–34} The hypothesis that external stimuli such as friction may have a role in the etiology does not explain unilateral presentation. In a minority of patients, additional lesions with features of LP or LPP may be found outside of the flexor surfaces. The association between LPPI and malignant neoplasms was suggested.^{12,31} The course of LPPI is generally rapid and some cases resolve in few weeks without therapy, while others may persist for years.³⁵ The pathogenesis of LPPI is similar to LPP, but

in LPPI a transition from papular LP to macular lesions of LPP has been observed. No causal relation could be detected with medications, sun exposure, internal malignancy, liver diseases, impaired carbohydrate metabolism, and race which were all proposed in the pathogenesis of LPP.²⁹ Ohshima *et al.* proposed that stimulation of sweat, moist environment or other external stimuli such as friction by way of Koebner phenomenon, may be involved in the etiology of LPPI.¹⁸ Majima *et al.* suggested that tightly fitting underclothes may have a possible causative role.²⁴ The pathogenesis is suggested to be related to a T lymphocyte mediated, cytotoxic activity against basal keratinocytes similar to classic LP.¹⁷ However, Pock *et al.* proposed that it is distinct from classic LP where the lichenoid reaction occurs with dramatically intensive hydropic degeneration of basal keratinocytes within such a short period that the compensatory increased proliferation of keratinocytes cannot develop.¹² Histopathological findings are similar to those found in classic LPP, although in LPPI, a band-like lichenoid infiltrate is more common.³⁶

Lichen planus pigmentosus along Blaschko's lines (linear, zosteriform)

LPP along Blaschko's lines is also known as linear LPP if it affects the limbs or the face, or zosteriform LPP if the trunk is affected.³⁷ It may be unilateral or bilateral, distributed along one or many Blaschko's lines.³⁸ It affects predominantly men, mostly in Asia.³⁹⁻⁴¹ In 2004, Hong *et al.* were the first to report two cases of LPP that presented as unilateral lesions with a linear pattern: one was localized on the leg and the other on the arm.⁴⁰ Seo *et al.* later reported another case of linear LPP in a male patient with a history of gastric cancer.⁴¹ Akarsu *et al.* were the first to

report a case of LPP with unilateral Blaschkoid involvement of the trunk.⁴²

The linearity of the lesion is probably related to Blaschko's lines, which suggests that the predisposition to develop LPP might be determined during embryogenesis through mosaicism, which determine cell populations with different immunological properties.³⁷

Blaschko's lines are thought to reflect T-lymphocytic migration and the clonal expression during embryogenesis of the skin. The genetic mosaicism may induce a mosaic T-cell response according to the trigger by viral infection or drugs. Some apoptotic changes may be also responsible for the response of the mosaic T cells that are present along Blaschko's lines.⁴¹ It has also been associated with hepatitis C virus.³⁸ It should be differentiated from other diseases with linear configurations such as linear and whorled nevoid melanosis, lichen striatus, and incontinentia pigmenti.¹⁵

Follicular LPP

Follicular LPP presents with pinpoint slate-gray macules affecting the hair follicle openings without any perilesional erythema or exaggerated skin markings.²⁵ It was initially described by Bhutani *et al.*,⁸ as perifollicular LPP. However, with the use of dermoscopy, the disease has been identified to involve hair follicle openings, unlike sparing of hair follicle in classical LPP.⁴³ Follicular LPP presents in younger patients during periods of disease instability and predominantly involves the upper limbs and trunk. Coexisting LP or lichen planopilaris are often associated.⁴³

LPP with frontal fibrosing alopecia (FFA)

Initially described from Australia in 1994, FFA is a variant of lichen planopilaris, characterized by progressive scarring alopecia affecting fronto-

temporal areas and eyebrows in female patients, especially after menopause. LPP has been studied as a condition that can precede or accompany FFA.^{19,44-46}

FFA-associated LP pigmentosus was first described by Dlova⁴⁵ in 2013 in more than 50% of 44 FFA patients, mostly African women. In contrast, a large FFA series published in 2014 reporting 355 Caucasian patients with FFA did not mention LP pigmentosus in association with their findings.

Scalp dermoscopy usually reveals loss of follicular openings, perifollicular erythema and perifollicular scales, adhered to the base of the hair shaft. The diagnosis must be confirmed with histopathology, with findings that include marked perifollicular fibrosis with reduction in the number of follicles and substitution by fibrous tracts. Lichenoid lymphocytic infiltrate around the infundibulum, isthmus and follicle bulge could also be found.^{47,48}

Mucosal LPP

It is a rare clinical variety of LPP. It was described by Laskaris *et al* as LP of oral mucosa with intense melanosis in a 54-year-old male. Based on the typical clinical reticular appearance and the histologic picture of oral lesions, the diagnosis of mucosal LPP was made.⁴⁹

HISTOPATHOLOGY

The histopathology of LPP forms a continuous spectrum with the earliest lesions showing marked inflammation at the interface, which later subsides, leaving behind the characteristic dermal pigmentation in older lesions.^{14,19} Characteristically, vacuolar degeneration of the epidermal basal cell layer, band-like lichenoid or perivascular lymphocytic infiltrates in the pap-

illary dermis, as well as superficial pigmentary incontinence and melanophages are seen in LPP. A mild epidermal atrophy with a basket-weave pattern of hyperkeratosis may be present in some cases. In older lesions, marked pigmentary incontinence, melanophages, decrease in vacuolar degeneration, and slight perivascular lymphocytic infiltrate are observed. Kanwar *et al.*, in 2003, reported a mild perivascular infiltrate in contrast to a band-like infiltrate.¹⁰

According to Al-Mutairi and El-Khalawany, the histological pattern varies according to the age of the lesions. The newer lesions exhibit a band-like infiltrate, the older lesions are characterized by a perivascular infiltrate.¹⁹ The direct immunofluorescence pattern also differed in the two studies. Kanwar *et al.* found that in 14.3% of cases there was a linear deposition of IgM or C3 in the basement membrane zone.¹⁰ Al-Mutairi and El-Khalawany, however, reported that in 16.7% of cases in the papillary dermis there was a globular deposition of IgM.¹⁹ Based on the similarity of DIF studies in LP and LPP, a correlation in the immunopathogenesis of the two diseases has been postulated.²⁰ There may also be CD1a expression in epidermis and upper dermis along with the predominance of CD8+ T cells.¹⁴ Some observers have also noted the presence of IgA and IgG in epidermal cells, the presence of C3 and fibrinogen in blood vessels and clusters of fluorescent bodies in the upper dermis.

ASSOCIATIONS

LPP can coexist with other variants of LP such as oral LP and classic LP.³²

LPP has been associated with endocrinopathies (diabetes, thyroid disease, and dyslipidemia). These associations may be due to the chronic

inflammatory state in patients with LPP, which may produce insulin resistance.⁵⁰ LPP has also been linked to autoimmune diseases such as vitiligo, lupus erythematosus,⁵¹ and circulating antinuclear antibodies,²⁰ acrokeratosis of Bazex,⁵² atopic dermatitis, and nephrotic syndrome.⁵³ head and neck carcinoma (paraneoplastic LPP),⁵² hepatitis C infection.³⁸

DERMOSCOPIIC FEATURES

Dermoscopy of LPP lesions has revealed pigmentation in different nonspecific patterns. Friedman *et al.*⁵⁴ described pigmentation in lesions of LPPI as diffuse, dotted, and mixed patterns. Diffuse pattern referred to structureless areas of brown pigmentation, probably epidermal pigmentation. Dotted pattern described fine or coarse blue-gray dots indicating dermal melanophages, and mixed pattern referred to lesions showing both epidermal and dermal components. The presence of dotted pattern correlated with a tendency for the pigmentation to persist.³⁴

In LPPI, brown homogeneous areas that represent epidermal pigmentation have been described on dermoscopy in addition to gray–brown or gray–blue dots and globules that represent pigmentary incontinence and melanophages in the papillary dermis. These gray dots are initially grouped in a diffuse black pepper-like pattern, but with time, they converge to form reticular, linear, and cobblestone patterns. White dots are secondary to lack of pigmentation in the follicular openings, and the absence of pigment in skin furrows may be due to lack of exposure to friction.⁵⁵

In their study on facial lesions of LPP, Sharma *et al* 2017,⁵⁶ showed that dots and/or globules are the most common dermoscopic findings. Various distribution patterns of dots and globules like

“hem-like,” “arcuate,” “incomplete reticular,” and “complete reticular,” were noted resembling different stages of the pigmentary network of skin. It is possible that these different patterns represent varying degrees of pigment incontinence, i.e., mild cases show hem-like and arcuate pattern. Whereas, more severe pigmentation demonstrates incomplete and complete reticular network. The finding of a central dot surrounded by a hypopigmented halo, resembling a targetoid lesion, corresponded with follicular plugging on histopathology. However, this may not necessarily be a clue to the diagnosis of LPP, and has been described in other unrelated diseases on the face as well.⁵⁷⁻⁵⁹ Gray-brown granular dots were reported in a case of axillary LPPI.⁵⁵

In a study of LPP associated with FFA, a dotted pattern (dotted/speckled blue-gray dots/blue-gray dots in circles) was also found. A pseudo-network pattern and telangiectasias were also found, particularly with facial involvement of LPP. Loss of facial vellus hairs, pigmentation of follicular openings and rhomboidal structures were described, likely because of greater follicular involvement in cases of combined LPP and FFA.⁴⁸ Dermoscopy may prove beneficial in differentiating among the various heterogeneous entities which can present as facial melanosis.^{60,61}

DIFFERENTIAL DIAGNOSIS

A number of diseases clinically and histologically similar to LPP have been reported in the literature during the last eight decades. These include ashy dermatosis, pigmented contact dermatitis, macular amyloidosis, and others.

Erythema dyschromicum perstans/ashy dermatoses

The first description of ashy dermatosis is credited

to Ramirez of El Salvador in 1957 who termed these patients Los cenicientos, meaning the ashens. ⁶² It was later called “erythema dyschromicum perstans” to highlight the erythematous halo around pigmented macules. ⁶³ In the 1980s, Bhutani *et al.*, ⁶³ gave a detailed account of a similar disease presenting as brown-to-slate-gray macules on the face, trunk and flexures of Indian patients. EDP is characterized by the appearance of discrete ash colored macules with slightly elevated erythematous margins. The lesions are asymptomatic and later coalesce, losing the erythematous borders and finally become gray-blue. The face, neck, arms, and trunk are involved. There are no associated systemic symptoms. The occurrence of classic LP in patients with ashy dermatosis further suggests that the two disorders may be related. ¹²

Clinically, it can be distinguished from LPP by a more widespread distribution beyond the sun-exposed areas. Extensive involvement over covered areas such as the trunk is common. Generalized hyperpigmentation could develop in the late stage of EDP. Pruritus, pigment hue, pattern, symmetry and too subtle (erythematous halo) are regarded as discriminating criteria between LPP and EDP. ²²

Histologically, both diseases are very similar and show an interface dermatitis characterized by an inflammatory infiltrate that abuts or obscures the dermoepidermal junction. ¹⁴ However, the infiltrate in EDP is not lichenoid but periadnexal. Pigmentation is in deeper dermis compared to the dermal pigmentation in LPP. Colloid bodies are typical of LPP but occasional in EDP. Dermoscopy may prove beneficial in differentiating between both entities. ²²

There is still a controversy regarding the rela-

tionship of EDP and LPP. Pittelkow and Daoud in 2008 suggested that LPP bears significant similarity to EDP and may represent an overlap disease. ⁶⁴ Zaynoun *et al.* in 2008 ⁶⁵ attempted to resolve the controversy by classifying ashy dermatosis clinically. According to their classification, ashy dermatosis may be classified into three categories. The first category is ashy dermatosis in which patients have idiopathic eruptive hyperpigmented macules, irrespective of the presence or absence of interface dermatitis histologically at the time of examination. Second category includes patients with EDP with lesions similar to those of ashy dermatosis, but who have or have had lesions with erythematous borders. The last category includes simulators as LPP, actinic LP postinflammatory hyperpigmentation, drug-induced melanoderma, mastocytosis. ⁶⁵

Pigmented contact dermatitis/Riehl's melanosis

Pigmented contact dermatitis (PCD) is a non-eczematous variant of contact dermatitis, characterized clinically by diffuse or patchy brown pigmentation with little or no signs of dermatitis, with predilection for the face and forehead. ²² The term “pigmented contact dermatitis” was coined by Osmundsen, a Danish dermatologist who described an epidemic of melanosis in Copenhagen. ⁶⁶ In a series of excellent observations and investigations, he proved that the melanosis was in fact due to contact dermatitis caused by an “optical whitener” present in a washing powder. Clinical characteristics, histopathology, and DIF features do help to differentiate cases with pigmented contact allergy. ²² But, a definitive diagnosis of PCD can only be established with a positive patch identifying the allergen. ⁶⁷

Melasma

Melasma is characterized by symmetrical pig-

mentation in sun-exposed areas; most commonly, face, with rare involvement of the “V” of neck and forearms.⁶⁸ The pigmentation can be epidermal, dermal, or mixed and based on areas affected, it may be centrofacial, mandibular, or maxillary. Pregnancy (chloasma) and hormonal therapy are common etiological factors in this condition, as such melasma has a strong female predilection. Both melasma and LPP share many epidemiological, clinical and histopathological features as both diseases are common in the dark skinned people. The location and distribution on the face are similar and both share the same melanin depositions in the epidermis and dermis.⁶⁹

Due to their clinical and histopathological similarities, Sharquie *et al* in 2015, suggested that there is a spectrum of diseases where melasma is at one pole, and butterfly lichen planus actinicus (LPA) at the other pole, with some cases lying in the middle of spectrum that have features of both diseases, and deserve the name melasma-like lichen planus actinicus (MLPA).⁶⁹

Postchikungunya pigmentation

An important differential to be considered, particularly in the Indian subcontinent, is post-chikungunya pigmentation. This pigmentation occurs after the infection with chikungunya virus and consists of freckle-like macules or a diffuse slaty-gray pigmentation in the malar area and root of the nose. It can persist up to 6 months.⁶⁸ It can have similar clinical manifestations as LPP; however, preceding history of fever and joint pain and characteristic involvement of the tip of the nose (Chik sign) helps in differentiation. Histology in post-chikungunya pigmentation shows increased pigmentation in basal layer, pigmentary incontinence, and melanophages with sparse dermal lymphohistiocytic infiltrate. Pigmenta-

tion usually resolves in weeks, but can persist for months after the remission of fever.⁷⁰

Acanthosis nigricans

It is characterized by hyperpigmented velvety plaques on the flexures; commonly the neck, axilla, and sometimes the groin with occasional involvement of the face in hereditary form. It may be familial or associated with diabetes, ingestion of drugs (nicotinic acid, insulin, corticosteroids) or malignancy.⁷¹

Terra-firma forme dermatosis

Terra-firma forme dermatosis is a benign condition with a characteristic clinical appearance of asymptomatic brown or black hyperkeratotic plaques or papules. Lesions are typically located on the neck, face, trunk, and ankles, although they may involve any area of the body. The application of 70% isopropyl alcohol is both diagnostic and therapeutic for this disorder.⁷² It has also been referred to as Duncan’s dirty dermatosis since the condition was first described by Duncan in 1987. This condition affects people of all ages, with some reports of a higher incidence in children and young adults.⁷² Biopsies are not frequently performed. If tissue is obtained, they show lamellar hyperkeratosis with compact orthokeratotic whorls and an absence of parakeratosis, acanthosis, papillomatosis, keratotic material between the papillae, and melanin deposition not only within the basal layer but also in the hyperkeratotic areas. The etiology of terra firma-forme dermatosis is unclear. Many believe this condition is caused by delayed keratinocyte maturation, which leads to the retention of keratinocytes and melanin within the epidermis.⁷³

Drug-induced pigmentation

Grayish pigmentation of the skin may occur due to deposition of drugs which include topical

hydroquinone (exogenous ochronosis), argyria, hydroxychloroquine, chloroquine, amiodarone, and minocycline. Biopsy shows banana-shaped yellow-brown granules in and around collagen bundles along with foreign body type granulomatous reaction.⁶⁷

Post-inflammatory pigmentation

Inflammatory skin reactions have the ability to disturb the dermoepidermal junction, causing melanin to pass into the dermis and producing persistent hyperpigmentation.

There is history suggestive of preexisting inflammation. In addition, the pattern of pigmentation may provide clues to underlying condition. The lesions are ill-defined and may show areas with patchy hypopigmentation.⁶⁷

Macular amyloidosis

Macular amyloidosis is a chronic disease thought to result from a combination of genetic and environmental causes. It occurs as a result of chronic friction such as scratching or using abrasive materials for scrubbing the skin.⁶⁷ However, the precise molecular mechanisms underlying its pathogenesis are not known. Macular amyloidosis has been reported in association with multiple endocrine neoplasia type 2A.⁷⁴

Cosmetic disfigurement and severe pruritus significantly impair quality of life. Macular amyloidosis is usually diagnosed clinically and can be confirmed by histology which shows amyloid deposition in the dermal papillae. The most common dermoscopic finding of macular amyloidosis is a central hub of either white or brown surrounded by various configurations of brownish pigmentation, including fine radiating streaks, dots, leaf-like projections, and bulbous projections.⁷⁵

Nevus of Ota and Hori, acquired bilateral nevus of Ota-like macules

Nevus of Ota manifests around puberty when the dermal melanocytes pigment under the influence of the hormonal spurt. These dermal melanocytes represent a defect in migration of melanocytes from the neural crest to the epidermis. It is almost always unilateral with blue-gray pigmentation in the ophthalmic and maxillary distribution of the trigeminal nerve, including the skin, eye, and oral mucosa. Hori nevus or acquired bilateral nevus of Ota-like macules manifests in adults with a speckled or confluent brownish-blue or blue-gray pigmentation of temples, malar area (where it mimics melasma), root of the forehead, eyelids, and alae nasi. Biopsy shows spindle-shaped dendritic melanocytes in the dermis.⁶⁷

Actinic lichen planus

Lichen planus actinicus is a photo-distributed variant of LP that most often occurs in dark-complexioned young adults of Middle Eastern descent.⁷⁶ Other names for this disorder include: summertime actinic lichenoid eruption, LP atrophicus annularis, LP in subtropical countries, LP subtropicus, LP tropicus, and lichenoid melanodermitis. Although the etiology of this disease has not been determined, sunlight appears to be the triggering factor in most cases.⁷⁷

The lesions of lichen planus actinicus usually occur on the forehead, face, neck, and the extensor surfaces of the arms and hands. Several morphologic patterns have been reported including annular hyperpigmented plaques, melasma-like patches, dyschromic papules, and classic lichenoid papules/plaques. Rarely, the non-exposed skin of the legs, trunk, genitals, and oral mucosa may be affected. Pruritus, scaling, nail

involvement, and the Köbner reaction frequently are absent.⁶⁷ Treatment is not always necessary as some cases of actinic LP resolve on their own. Mild cases can often be managed with topical steroids, while more intensive therapies may be required for severe cases.⁷⁸

TREATMENT

LPP is a disease which is basically recalcitrant to treatment and therapies attempted in this disorder are quite ineffective. Management includes avoidance of exacerbating factors which includes photoprotection in the actinic variant. In the inversus variant, measures to reduce friction in the body folds such as weight reduction and avoidance of tight clothes are advisable.

Topical agents include hydroquinone, which is the most commonly used agent, often in combination with retinoic acid, corticosteroids, azelaic acid, kojic acid, and glycolic acid.^{10,19} Topical steroids have been commonly used but are of doubtful efficacy. Topical tacrolimus, specific suppressor of T-cell mediated inflammation, is considered a reasonable treatment option.¹⁴ Al-Mutairi and El-Khalawany *et al.* found tacrolimus ointment (0.1%) to be effective in 53.8% of patients.¹⁹

Systemic treatment modalities have also been tried including systemic corticosteroids and Vitamin A which was recommended by Bhutani *et al.*⁷⁹ The use of dapsone may halt the progression of pigmentation according to some authors. Sehgal *et al.* suggested a combination of oral diamino-diphenyl-sulfone (dapsone) along with oral immunomodulator, topical tacrolimus along with photoprotection for the treatment of LPP.¹⁴ Since the clearance of the lesion is seldom complete, the use of lasers or their combination with

topical agents can be considered. A report on use of low-fluence Q-switched Nd-YAG (1064 nm) in 3 weekly sessions, combined with 0.1% tacrolimus, in linear LPP of the forehead, for a total duration of 4 months showed complete clearance of pigmentation, with no recurrence at 6 months of follow-up.⁸⁰

PROGNOSIS

Course is highly variable. Pigmentation may last only a few weeks and then resolve spontaneously. Sometimes, it may last months to years and be refractory to treatment. This unpredictable course, added to the rarity of the disease and marked overlap with conditions such as EDP, has prevented proper comparative studies between different treatment options.¹⁵

CONCLUSIONS

LPP is a pigmentary disorder associated with lesions of LP. Several clinical variants of LPP have been identified. A controversy has erupted as to whether LPP and EDP are the same disorder or separate entities. LPP is recalcitrant to treatment and therapies attempted in this disorder are quite ineffective, however the most reasonable treatment is topical tacrolimus.

REFERENCES

1. Rieder E, Kaplan J, Kamino H, et al. Lichen planus pigmentosus. *Dermatol Online J* 2013; 19:20713.
2. Lichen planus pigmentosus. Orphanet. May 2011.
3. Lichen planus. *DermNet NZ*. March 2015.
4. Vashi N, Kundu R. Approach to the patient with hyperpigmentation disorders. *Up To Date*. January 2015.
5. Gougerot M. Lichens atypiques ou invisibles pigmentogenes reveles par des pigmentations. *Bull Soc Fr Dermatol Syphiligr* 1935; 42:792-94.
6. Shima T. Supplementative study of lichen pigmentosus. *Jpn J Dermatol* 1956; 66:353-56.

7. Ramirez CO. Dermatitis cenicenta: Estudio clinico de 32 casos. *Derm Mex* 1963; 7:232-44.
8. Bhutani LK, Bedi TR, Pandhi RK, et al. Lichen planus pigmentosus. *Dermatologica* 1974; 149:43-50.
9. Vega ME, Waxtein L, Arenas R, et al. Ashy dermatosis versus lichen planus pigmentosus: A controversial matter. *Int J Dermatol* 1992; 31:87-88.
10. Kanwar AJ, Dogra S, Handa S, et al. A study of 124 Indian patients with lichen planus pigmentosus. *Clin Exp Dermatol* 2003; 28:481-85.
11. Chen S, Sun W, Zhou G, et al. Lichen planus pigmentosus-inversus: Report of three Chinese cases and review of the published work. *J Dermatol* 2015; 42:77-80.
12. Pock L, Jelínková L, Drlík L, et al. Lichen planus pigmentosus-inversus. *J Eur Acad Dermatol Venereol* 2001; 15:452-54.
13. Ghorbel HH, Badri T, Ben Brahim E, et al. Lichen planus pigmentosus inversus. *Indian J Dermatol Venereol Leprol* 2014; 80:580-85.
14. Sehgal VN, Verma P, Bhattacharya SN, et al. Lichen planus pigmentosus. *Skin med* 2013; 11:96-103.
15. Robles-Mendez J, Rizo-Fr P, Herz-Ruelas M, et al. Lichen planus pigmentosus and its variants: review and update. *Int J Dermatol* 2018; 57:505-09.
16. Dizen Namdar N, Kural E, Pulat O, et al. Lichen planus pigmentosus-inversus: 5 Turkish cases. *J Eur Acad Dermatol Venereol* 2016; 30:450-52.
17. Kashima A, Tajiri A, Yamashita A, et al. Two Japanese cases of lichen planus pigmentosus-inversus. *Int J Dermatol* 2007; 46:740-42.
18. Ohshima N, Shirai A, Saito I, et al. Lichen planus pigmentosus-inversus occurring extensively in multiple intertriginous areas. *J Dermatol* 2012; 39:412-14.
19. Al-Mutairi N, El-Khalawany M. Clinicopathological characteristics of lichen planus pigmentosus and its response to tacrolimus ointment: an open label, non-randomized, prospective study. *J Eur Acad Dermatol Venereol* 2010; 24:535-40.
20. Parodi G, Parodi A, Guarrera M, et al. Lichen pigmentosus with scarring alopecia and circulating antinuclear antibodies. *Int J Dermatol* 1990; 29:227-28.
21. Kulthanan K, Jiamton S, Varothai S, et al. Direct immunofluorescence study in patients with lichen planus. *Int J Dermatol* 2007; 46:1237-41.
22. Tienthavorn T, Tresukosol P, Sudtikoonaseth P. Patch testing and histopathology in Thai patients with hyperpigmentation due to erythema dyschromicum perstans, lichen planus pigmentosus, and pigmented contact dermatitis. *Asian Pac J Allergy Immunol* 2014; 32:185-92.
23. Verma P, Pandhi D. Topical tacrolimus and oral dapsone combination regimen in lichen planus pigmentosus. *Skinmed* 2015; 13:351-54.
24. Majima Y, Yagi H, Ikeda Y, et al. Two cases of lichen planus pigmentosus inversus: Possible causative role of tightly fitting underclothes. *Eur J Dermatol* 2013; 23:904-05.
25. Kumaran M, Dabas G, Parsad D. Lichen planus pigmentosus – An appraisal. *Inter J Dermatol* 2018; 57:742-54.
26. Gaertner E, Elstein W. Lichen planus pigmentosus-inversus: Case report and review of an unusual entity. *Dermatol Online J* 2012; 18:11-15.
27. Nurmohamed S, Hardin J, Haber RM. Lichen planus pigmentosus inversus in children: Case report and updated review of the literature. *Pediatr Dermatol* 2018; 35:e49-e51.
28. Mohamed M, Korbi M, Hammedi F, et al. Lichen planus pigmentosus inversus: a series of 10 Tunisian patients. *Int J Dermatol* 2016; 55:1088-91.
29. Bennisar A, Mas A, Julia M, et al. Annular plaques in the skin folds: 4 cases of lichen planus pigmentosus-inversus. *Actas Dermosifiliogr* 2009; 100:602-05.
30. Barros HR, Almeida JR, Mattos- Dinato SL, et al. Lichen planus pigmentosus inversus. *An Bras Dermatol* 2013; 88(6 Suppl 1):146-49.
31. Jung YJ, Lee YH, Lee SY, et al. A case of lichen planus pigmentosus-inversus in a Korean patient. *Ann Dermatol* 2011; 23:61-63.
32. Kim BS, Aum JA, Kim HS, et al. Coexistence of classic lichen planus and lichen planus pigmentosus-inversus: resistant to both tacrolimus and clobetasol propionate ointments. *J Eur Acad Dermatol Venereol* 2008; 22(1):106-07.
33. Kim BS, Park KD, Chi SG, et al. Two cases of lichen planus pigmentosus-inversus arising from long-standing lichen planus-inversus. *Ann Dermatol* 2008; 20(4):254-56
34. Murzaku EC, Bronsnick T, Rao BK. Axillary lichen planus pigmentosus-inversus: dermoscopic clues of

- a rare entity. Diagnosis: lichen planus pigmentosus (LPP). *J Am Acad Dermatol* 2014; 71(4):e119-20.
35. Villaverde RR, Gutierrez MG, Cano DS, et al. Lichen plano pigmentoso inverso: fácil de diagnosticar, difícil de tratar (Lichen planus pigmentosus inversus: easy to diagnose, difficult to treat). *Piel* 2011; 26:538-40.
 36. Tabanluglo- Duncan D, Ince-Uysal P, Yayla D et al. Lichen planus pigmentosus-inversus: A peculiar variant of lichen planus. *Dermatol Sinica* 2017;35:210-12.
 37. Vineet R, Sumit S, K GV, et al. Lichen planus pigmentosus in linear and zosteriform pattern along the lines of Blaschko. *Dermatol Online J* 2015; 21:11-15.
 38. Vachiramon V, Suchonwanit P, Thadanipon K. Bilateral linear lichen planus pigmentosus associated with hepatitis C virus infection. *Case Rep Dermatol* 2010; 2:169-72.
 39. Sonthalia S, Das A, Sharma S. Co-localization of linear lichen planus pigmentosus and milia in a child. *Indian J Dermatol* 2016; 61:237-40.
 40. Hong S, Shin JH, Kang HY. Two cases of lichen planus pigmentosus presenting with a linear pattern. *J Korean Med Sci* 2004; 19:152-54.
 41. Seo JK, Lee HJ, Lee D, et al. A case of linear lichen planus pigmentosus. *Ann Dermatol* 2010; 22:323-25.
 42. Akarsu S, Ilknur T, Özer E, et al. Lichen planus pigmentosus distributed along the lines of Blaschko. *Int J Dermatol* 2013; 52:253-54.
 43. Sindhura KBN, Vinay K, Kumaran MS, et al. Lichen planus pigmentosus: a retrospective clinico-epidemiologic study with emphasis on the rare follicular variant. *J Eur Acad Dermatology Venereol* 2016; 30:e142-e44.
 44. Rao R, Sarda A, Khanna R, et al. Coexistence of frontal fibrosing alopecia with lichen planus pigmentosus. *Int J Dermatol* 2014; 53:622-24.
 45. Dlova NC. Frontal fibrosing alopecia and lichen planus pigmentosus: Is there a link? *Br J Dermatol* 2013; 168:439-42.
 46. Berliner JG, Mc Calmont TH, Price VH, et al. Frontal fibrosing alopecia and lichen planus pigmentosus. *J Am Acad Dermatol* 2014;71:e26-27.
 47. Rubegni F, Fimiani M. Frontal fibrosing alopecia:role of dermoscopy in differential diagnosis. *Case Rep Dermatol* 2010; 2:40-45.
 48. Pirmez R, Duque-Estrada B, Donati A, et al. Clinical and dermoscopic features of lichen planus pigmentosus in 37 patients with frontal fibrosing alopecia. *Br J Dermatol* 2016; 175:1387-90.
 49. Laskaris GC, Papavasiliou SS, Bovopoulou OD, et al. Lichen planus pigmentosus of the oral mucosa: a rare clinical variety. *Dermatologica* 1981; 162:61-63.
 50. Torres J, Pena Romero AG, Reyes E, et al. Lichen planus pigmentosus in patients with endocrinopathies and hepatitis C. *J Am Acad Dermatol* 2013; 68:139-43.
 51. Seidel A. Lichen planus pigmentosus (LPP) and lichen planus pigmentosus: 35 cases in Armenia, Colombia. *J Am Acad Dermatol*. 2015; 72:114-17.
 52. Sassolas B, Zagnoli A, Leroy JP, et al. Lichen planus pigmentosus associated with acrokeratosis of Bazex. *Clin Exp Dermatol* 1994; 19:70-73.
 53. Mancuso G, Berdondini RM. Coexistence of lichen planus pigmentosus and minimal change nephrotic syndrome. *Eur J Dermatol* 2009; 19:389-90.
 54. Friedman P, Sabban EC, Marcucci C, et al. Dermoscopic findings in different clinical variants of lichen planus. Is dermoscopy useful? *Dermatol Pract Concept* 2015; 5:51-55.
 55. Guengoer S", Topal IO, Goencue EK. Dermoscopic patterns in active and regressive lichen planus and lichen planus variants: a morphological study. *Dermatol Pract Concept*. 2015; 5:45-53.
 56. Sharma VK, Gupta V, VEDI KK, et al. Dermoscopy and patch testing in patients with lichen planus pigmentosus on face: Across-sectional observational study in fifty Indian patients. *Indian J Dermatol Venereol Leprol* 2017; 83:656-62.
 57. Zalaudek I, Giacomel J, Argenziano G, et al. Dermoscopy of facial nonpigmented actinic keratosis. *Br J Dermatol* 2006; 155:951-56.
 58. Zalaudek I, Giacomel J, Schmid K, et al. Dermoscopy of facial actinic keratosis, intraepidermal carcinoma, and invasive squamous cell carcinoma: A progression model. *J Am Acad Dermatol* 2012; 66:589-97.
 59. Lallas A, Argenziano G, Apalla Z, et al. Dermoscopic patterns of common facial inflammatory skin diseases. *J Eur Acad Dermatol Venereol* 2014; 28:609-14.
 60. Gil I, Segura S, Martinez-Escala E et al. Dermoscopic and reflectance ochronosis. *Arch Dermatol* 2010; 146:1021-25.
 61. Wang L, Xu AE. Four views of Riehl's melanosis: Clinical appearance, dermoscopy, confocal microscop-

- py and histopathology. *J Eur Acad Dermatol Venereol* 2014; 28:1199-206.
62. Ramirez CO. Los Cenicientos: Problema Clinico. In: Report of the First Central American Congress of Dermatology, San Salvador, 5-8 December, 1957. San Salvador: Central American Dermatological Society, 1957; 122-30.
 63. Bhutani LK. Ashy dermatosis or lichen planus pigmentosus: What is in a name? *Arch Dermatol* 1986; 122:133-36.
 64. Convit J, Kerdel-Vegas F. Erythema dyschromicum perstans a hitherto undescribed skin disease. *J Invest Dermatol* 1961; 36:457-62.
 65. Zaynoun S, Rubeiz N, Kibbi AG. Ashy dermatoses - A critical review of the literature and a proposed simplified clinical classification. *Int J Dermatol*. 2008; 47:542-44.
 66. Osmundsen PE. Pigmented contact dermatitis. *Br J Dermatol* 1970; 83:296-301.
 67. Mathews L, Thappa DM, Singh N et al. Lichen planus pigmentosus: A short review. *Pigment Int* 2016;3:5-10.
 68. Khanna N, Rasool S. Facial melanoses: Indian perspective. *Indian J Dermatol Venereol Leprol* 2011; 77:552-63.
 69. Sharquie KE, Noaimi AA, and Al-Shukri MA. Melasma, Melasma-Like Lichen Planus Actinicus, and Butterfly Lichen Planus Actinicus Build up One Spectrum (Clinico-Histopathological Study). *J Cosmetic Dermatol Sci Appls* 2015; 5:212-22.
 70. Riyaz N, Riyaz A, Rahima, et al. Cutaneous manifestations of chikungunya during a recent epidemic in Calicut, north-Kerala, south India. *Indian J Dermatol Venereol Leprol* 2010; 76:671-75.
 71. Gupta V, Sharma V. Ashy dermatosis, lichen planus pigmentosus and pigmented cosmetic dermatitis: Are we splitting the hair? *Indian J Dermatol Venereol Leprol* 2018; 84(4):510-14.
 72. Akkash L, Badran D, Al-Omari AQ. Terra firma-forme dermatosi. Case series and review of the literature. *J Dtsch Dermatol Ges* 2009; 7:102-07.
 73. Erkek E, Sahin S, Cetin ED, et al. Terra firma-forme dermatosis. *Indian J Dermatol Venereol Leprol* 2012; 78:358-60.
 74. de Argila D, Ortiz-Romero PL, Ortiz-Frutos J, et al. Cutaneous macular amyloidosis associated with multiple endocrine neoplasia 2A. *Clin Exp Dermatol* 1996; 21:313-14.
 75. Errichetti E, Stinco G. Dermoscopy in General Dermatology: A Practical Overview. *Dermatol Ther (Heidelb)*. 2016; 6(4):471-507.
 76. Salman SM, Kibbi AG, Zaynoun S. Actinic lichen planus. *J Am Acad Dermatol* 1989; 20:226-31.
 77. Meads S, Kunishige J, Ramos-Caro F, et al. Lichen Planus Actinicus *Cutis* 2003; 72:377-81.
 78. Chuang T. Lichen Planus. *Medscape Reference*. May 2015.
 79. Bhutani LK, George M, Bhate SM. Vitamin A in the treatment of lichen planus pigmentosus. *Br J Dermatol* 1979; 100:473-74.
 80. Kim JE, Won CH, Chang S, et al. Linear lichen planus pigmentosus of the forehead treated by neodymium: Yttrium-aluminum-garnet laser and topical tacrolimus. *J Dermatol* 2012; 39:189-91.