Lichen planus pigmentosus in two Filipino males: A case report

Roy Lawrence S. Paredes, MD; Ciara Mae H. Dela Cruz, MD; Eugenio Pipo, MD; Johannes F. Dayrit, MD FPDS

INTRODUCTION

Lichen planus is an interesting dermatological condition that affects between 0.5 - 1% of the population and is often characterized by the classic description of pruritic polygonal violaceous flat topped papules.1

Lichen planus pigmentosus is a rare variant of lichen planus and is often found in dark-skinned individuals. Due to its uncommon nature, only a few cases have been generally reported worldwide and the frequency is still unknown.1

CASE REPORT

Case 1

The first case is a 71-year-old male who presented with a five-month history of pruritic, gradually enlarging, gray to black macules and patches on the face, trunk and arms. Lesions initially appeared on the proximal area of the right arm which gradually increased in size and number and have spread to the face, chest, lower abdomen, trunk and interdigital areas (Fig. 1). No other symptoms were associated in the affected areas except for occasional pruritus. Persistence of the lesions prompted consult.

Review of systems was unremarkable and no other co-morbidities were noted. Dermatologic examination showed multiple bluish-gray macules and patches on the central face, right proximal arm, lower abdomen, lower back and intertriginous areas (Fig. 2). Dermoscopic examination revealed blue-gray dots in brown circles (Fig. 3a).

A 4-mm skin punch biopsy was taken from the lower back which revealed thinning of the epidermis with multifocal vacuolar alteration of the basal cell layer. The dermis shows a lichenoid inflammatory infiltrate of lymphocytes. Numerous pigment-laden macrophages were also seen (Fig. 4a).

To further confirm the diagnosis, direct immunofluorescence was done which presented shaggy linear deposits of fibrinogen on the basement membrane zone that is compatible with a lichenoid tissue reaction. The patient was treated with tacrolimus 0.03% ointment bid.
Case 2

The second case is a 19-year-old male who presented with a one-year history of appearance of multiple erythematous, macules and patches that gradually evolved into grayish patches on the bilateral thighs. Lesions were not painful nor associated with any pruritus. No consultation was done and no medications were taken nor applied. Six months prior to consult, lesions increased in size and number now affecting the face, neck and post-surgical site on the abdomen. The patient denies any allergies nor any maintenance medications. Past medical history revealed that patient underwent an exploratory laparotomy secondary to appendicitis two years prior to consult. Physical examination showed multiple well defined, ash-gray, macules and patches on the face, neck, trunk and extremities with no oral lesions nor any nail findings. An ash-gray patch was also observed on the laparotomy scar (Fig. 2). Dermoscopy showed gray homogenous pigmentation with multiple speckled blue-gray dots (Fig. 3b). A 4-mm skin punch biopsy was done on the back which showed mild acanthosis of the epidermis with mild spongiosis, few necrotic keratinocytes, exocytosis of lymphocytes and multifocal vacuolar alteration of the basal cell layer. The dermis revealed mild fibrosis and many pigment-laden macrophages (Figure 4b). The patient was treated with 0.03% tacrolimus ointment with visible lightening of the hyperpigmented patches after 2 months of follow up.

DISCUSSION

Lichen planus pigmentosus is a rare variant of lichen planus predominantly found in the middle age group among those of Indian or Middle Eastern Descent. Clinically lesions present with hyperpigmented macules, and patches on exposed areas. A type of LPP called lichen planus pigmentosus inversus on the other hand, usually presents on skin folds. Patterns of distribution vary as case reports on LPP present as either diffuse, blotchy reticular or linear. Dermoscopic features of LPP usually show blue-gray dots in arcs and circles on speckled blue-gray dots on a homogenous brown background, but wickham striae and vascular patterns are not observed.

LPP can also demonstrate true Koebner phenomenon where skin lesions appear on areas of trauma such as in case 2 in which the patient presented with a gray patch on a previous appendectomy scar. Lesions vary in their symptoms ranging from asymptomatic, pruritic, or even burning sensation on the areas affected. The disease course is variable displaying exacerbations and remission, but most of the time it is chronic in nature. No definite etiology however has been reported. Sunlight has been highlighted as a possible cause for lesions presenting in exposed areas. Retrospective studies cite photosensitizers in mustard oil called allyl thiocyanate as a possible cause as well as Hepatitis C infection.

Lesions of LPP are often confused with erythema dyschromicum perstans, a disease that presents as gray to brown macules and patches on face, trunk and proximal extremities. Histologically, in EDP, the epidermis is either normal or atrophic with presence of pigmenitary incontinence as well as perivascular infiltrate. Direct immunofluorescence reveals no specific staining of the cytid bodies IgG, IgM and C3.
In LPP, lesions display epidermal thinning with basal vacuolization, pigment incontinence and a superficial lichenoid dermal infiltrate. Immunofluorescence helps confirm the diagnosis showing the presence of IgM and IgG and complement to colloid bodies.

EDP differs from LPP as lesions initially present as hyperpigmented macules and patches with a thin ring of erythema at the periphery of the lesions. Also, EDP usually presents as blue gray colored lesions due to the tyndall effect.

Patients affected with LPP often cite that they would want to address the discoloration which for them is cosmetically unacceptable. Several treatment modalities have been used to address the lesions of LPP. Commonly used agents include topical steroids, calcineurin inhibitors and hydroxychloroquine. In a study by Mutairi et al, the use of 0.03% tacrolimus ointment have shown an appreciable lightening effect in 7 out of 13 patients treated. Combination of 0.1% tacrolimus and Nd-YAG laser was also proven to be helpful for a case of linear LPP that was not responsive to topical steroids. Other treatment modalities used were dapsone and hydroxychloroquine.

Despite these reports, no definite treatment still exists. In a study by Vachiiramon et al. sun avoidance and management of Hepatitis C virus showed clinical improvement of lesions. A recent clinical trial, showed that low dose isotretinoin stabilized and decreased pigmentation in early and limited lesions of LPP.

CONCLUSION

LPP is a rarely seen variant of lichen planus that commonly manifests as a brownish gray patches. The lesion mimics hyperpigmented dermatoses such as erythema dyschromicum perstans and ashy dermatosis. Definite diagnosis relies on histopathological characteristic of vacuolar alteration of the basal cell layer, lichenoid infiltrate, and pigment-laden macrophages. Both patients were treated with 0.03% tacrolimus ointment and advised follow up to monitor if there is improvement. This case is definitely an addition to a small database dedicated to cases of LPP in our institution.