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An early presentation of lichen planus pigmentosus: Clinicopathological and dermoscopic study

Gnana Prabha M[1], Bhadrinath N[2]
[1] Post Graduate, Department of Dermatology, KVG Medical College and Hospital, Sullia, Karnataka, India
[2] Professor and HOD, Department of Dermatology, KVG Medical College and Hospital, Sullia, Karnataka, India

Abstract
Lichen planus pigmentosus is a photodistributed dyschromia of unknown etiology described clinically as hyperpigmented gray-blue or brown-black macules or patches in a photodistributed pattern.
Although there has been some debate, lichen planus pigmentosus is considered by many to be a separate diagnostic entity from ashy dermatosis or erythema dyschromicum perstans, which shares similar characteristics. Various treatment strategies have been applied to help resolve or improve the appearance of lichen planus pigmentosus lesions; however, an optimal treatment method is yet to be elucidated.
Lichen Planus Pigmentosus (LPP) is considered a macular variant of lichen planus with an unknown etiology. The condition was first described by Bhutani *et al* as a condition recognized in India, presenting in the third to fourth decade of life and manifesting as dyspigmented macules or patches. Lesions can exhibit a range of pigmentary dyschromia, from brown-blackish to blue or purple-gray macules and patches, which predominantly involve the sun-exposed aspects of upper extremities, face, and neck. Areas of involvement tend to be asymptomatic, but pruritus or burning may be present.
Histology classically demonstrates hyperkeratosis, epidermal thinning, vacuolar degeneration of the basal cell layer, lymphohistiocytic band like infiltrate, variable Civatte or colloid bodies, and melanin incontinence. There are several treatment options that have been reported with variable efficacy for improvement of the appearance of the lesions, but due to the chronicity of the underlying disease process, none of these treatments are curative.

Keywords: lichen planus, lichen planus pigmentosus

Introduction
Lichen planus pigmentosus is a variant of lichen planus which was first described by Bhutani in 1974 [1]. It is a frequently seen is in Indian population [2], with distinctive clinical and histological charecteristics. It presents as persistent and asymptomatic slate-grey pigmentation in darker individuals, and can also rarely present as macular hyperpimentation in lighter individuals involving mainly face, neck, and upper limbs although it may be more widespread. It generally starts from third of fourth decade of life. Here is a case where the lesion started from 3 years of age.

Case Report
Here is a 7 year old, male patient presenting to opd with complaints of asymptomatic lesions initially noticed over neck when he was of 3 year old, and then over back, both upper limb and over face mainly over peri orbital region. Which were initially small in size and gradually progresssed to present size. With no history of similar complaints in family. On examination, they were found to be brown to black colored macules and patches over nape of neck extending towards lateral side of neck, diffusely over upper back, both upper arms, around peri orbital region with dryness of skin noted over patch.

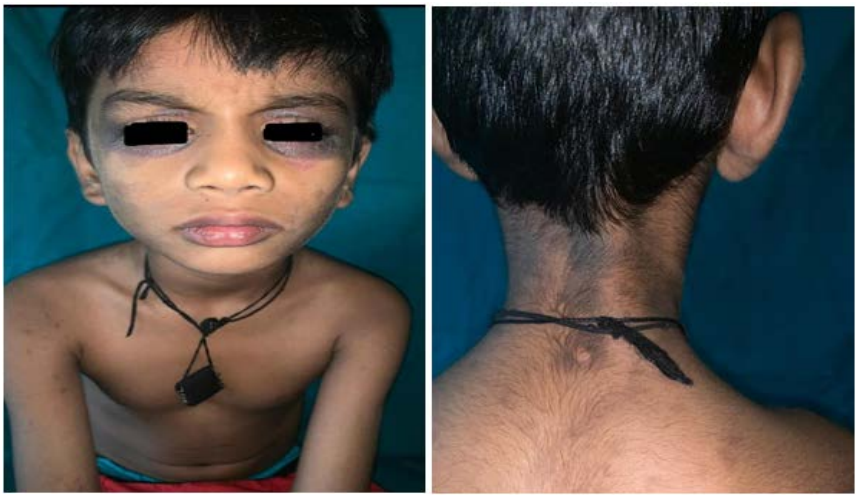

Fig 1

For which incisional biopsy was done from lesion over neck. And dermoscopy was done under non polarized light. Histopathology report showed as epidermis with basal pigmentation. Dermis shows melanophages with pigment. No basal vacuolar or band like dermal infilterate of lymphocytes. No perivascular infilterate.

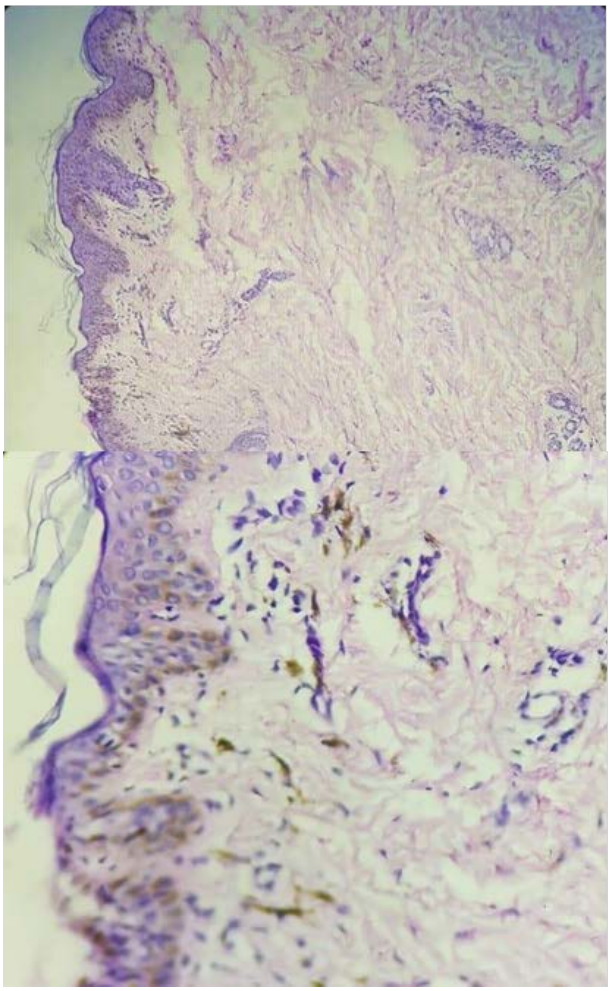

Fig 2

Dermoscopy showed diffuse dark brown globules indicating epidermal pigmentation. And grayish dotts indicating dermal melanophages [3]. Investigation was negative for hepatitis c. And patient was given vitamin A supplements along with topical tacrolimus 0.03% ointment with not much improvement.

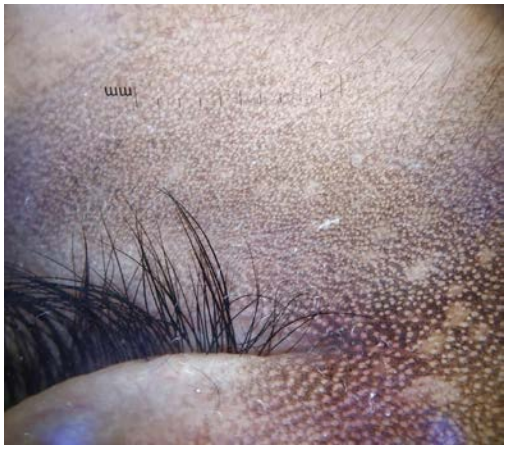

Fig 3

Fig 4

Discussion

LPP is a chronic hyperpigmentary disorder runs a prolonged and insidious course and has unknown etiology. Cases from are reported mainly from India, Japan, Korea, Middle east and Latin America, although initially described in Indians, subsequently seen in other racial and ethnic group [4]. It usually starts from third and fourth decade of life, and has slight female predilection.

Etiology is not known in our case. Sun exposure has been postulated as one the causes. But as the age of the patient is very young, sun exposure may not be the cause in this case and other postulated etiological agents Abnormalities in the T-lymphocyte functions, hepatitis C virus, cosmetic agents such as kumkum and hair dyes mustard oil (contains the potential photosensitizer allyl thiocyanate, amla oil where photosensitivity may be precipitated by fragrances). So the probable cause could be abnormal T-lymphocyte function and genetic predisposition, further studies are required to confirm in this respect [5].

LPP mainly occurs on sun exposed sites like face, neck, and intertriginous areas such as axillary, inguinal, and sub mammary folds. Palms, soles, nails are not affected, rarely it can be generalized and oral mucosa can be affected. The disease is insidious in onset and progressive course so that old lesions enlarge in size and deepening of colour can also be seen [4]. Lesions may initially state as small, ill to well defined, oval to round macules, color may vary from slate gray to brownish black. And later become confluent to larger areas of pigmentation. Pigmentation is mostly diffuse or reticular pattern. Although disease is asymptomatic it may also present with pruritis and burning sensation in one-third of cases [4].

Differential diagnosis of the condition being erythema dyschromica perstans, drug induced hyperpigmentation,(a case of facial dyspigmentation), macular amyloidosis, urticaria pigmentosa, berloque melanosis, riehl's melanosis (pigmented cosmetic melanosis), heavy metal and tar induced pigmentation, frictional melanosis [5].

The histopathological changes consist of vacuolar degeneration of basal layer of epidermis, hence thinning of epidermis. A band of infilterate of lymphocytes are usually present at the dermo-epidermal junction. In the dermis, a

perivascular lymphohistiocytic infilterate and presence of melanophages or with melanin incontinence are the important features present [4]. It has to be differentiated from ashy dermatoses as both have similar histopathology findings.

In dermoscopy, the colour of dots, globules and background are the important clue to diagnosis in case if we want clarity in diagnosis. The colour appreciated here ranged from dark brown to light brown. lichen planus pigemtosus shows larger dots and globules that are brownish in colour (due to pigment incontinence) this is due to melanin deposit which is relatively more superficial which is a result of lichenoid interface dermatitis occurring just at dermoepidermal junction. Where as in case of ashy dermatosis presence of small dots which bluish gray over a bluish background, which is due to tyndall effect due to presence melanin in deep dermis [6]. And normal white dots which are seen due to eccrine openings.

Siddharth sonathalia *et al* found diffuse light to dark brown as background colour which is characteristics, along with which they noticed a faint erythema along exaggerated pigmented network. And they have also described targetoid lesions which showed a central dot surrounded by white halo, correlating it to follicular plugging of histopathology [6].

Reported treatment modalities include topical immunomodulators which has improvement by lightening of pigmentation in 12 weeks [7], topical glucocorticoids,

Topical keratolytics, oral vitamin A 100000IU for 15 days and 15 days without treatment for 10 or more treatments reported improvement [1], superficial chemical peels, oral steroids, Nd-YAG laser therapy for 28 sessions shows improvement and lightnening of pigmentation [8].

Amanjot K arora reported a case of lichen planus pigmentosus which presented as racoon eyes in a 44 year old female. There was mainly periocular pigmentation. But in our case lesions are mainly seen as around eyes along with neck, upper back and upper limbs. Periorbital pigmentation may be due to idiopathic or secondary cause, which may be due to excessive pigmentation akin to dermal melanocytosis, post inflammatory hyperpigmentation secondary to atopic allergic, cosmetic, genetic [9].

Kanwar *et al* reported that lichen planus pigmentosus usually affects female and rare in children, in contrast here our patient seven year old [10].

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