LICHEN DYSCHROMICUM PERSTANS

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A disorder of hyperpigmentation is described which is seen commonly in middle aged individuals. Face and neck including upper part of the trunk are mostly affected. The lesions are usually bilaterally symmetrical and occur in a localized or generalized distribution. Oral mucosa is infrequently involved. The colour of pigmentation varies from grey, blue and brown to black and it persists for years. None of the patients had any features of lichen planus. Histopathology reveals varying rates of lichenoid tissue reactions. We have tentatively designated this disorder as lichen dyschromicum perstans. Its relationship with other similar disorders is discussed.

Key Words : Hyperpigmentation, Lichenoid tissue reaction

Introduction

Compared to disorders of hypo or depigmentation, the hyperpigmentary disorders have got less attention in literature even though a hyperpigmented skin lesion is no less psychologically traumatizing than a patch of leucoderma. However, their varied clinical presentation and obscure aetiology often poses diagnostic problems. In the present communication, we describe a disorder of hyperpigmentation which we see fairly frequently at the Pigmentation Clinic of the Postgraduate Institute of Medical Education and Research, Chandigarh, India. We have tentatively designated this disorder as lichen dyschromicum perstans (LDP). The present report highlights the clinical and histological features of this entity and its relationship with other clinically similar disorders.

Materials and Methods

The study pertains to fifty-five patients seen during a five-year period from 1988 to 1993. In every patient, a detailed history was sought regarding the site of onset of pigmentation, its rate of progression, associated symptoms and family history of similar pigmentation. Information was also elicited regarding use of cosmetics (soaps, toiletries etc) and drug intake. Any other precipitating or incriminating factors associated with pigmentation were noted. All the above information was recorded in specially devised proforma. During clinical examination, the morphology, distribution, extent of involvement and the exact colour of the pigmentation were recorded. Changes in oral mucosa, hair and nails were also noted.

In every patient, the following investigations were carried out: complete haemogram, routine urine and stool, blood sugar, serum calcium, urea, creatinine, transaminases, alkaline phosphatase, amylase and skiagram of chest.

Histopathological examination of skin was done in 39 patients from diffuse hyperpigmented lesions as well as from perifollicular lesions when present. In histopathology, specific features looked for were: basal cell degeneration, colloid bodies and the nature and site of infiltrate. Special stains were used to detect amyloid and hemosiderin. Direct immunofluorescence (DIF) was performed in 14 patients.

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Results

Incidence : Of 332 patients seen with disorders of hyperpigmentation during the 5-year period of study, 55 had lichen dyschromicum perstans; thus its incidence was 16.5%.

Age and sex distribution : Of 55 patients, 34 were females and 21 were males; the female male ratio being 3:2. Their ages ranged from 10-65 years (mean 30 years). The age range in males (10-40 years; mean 21.9 years) was lower compared to females (14-65 years; mean 34.9 years). Majority of the patients, 47 (85%) were between 15-45 years. Mean age at onset was 30 years in both sexes (Table I).

Table I. Age and sex distribution of patients with lichen dyschromicum perstans

Age group (years)	Males %	Females %	Total %
<15	5 (24)	1 (3)	6 (11)
15-30	9 (43)	10 (29)	19 (35)
30-45	7 (33)	21 (62)	28 (50)
>45	-	2 (6)	2 (4)
Total	21 (100)	34 (100)	55 (100)

Onset of disease and age : Peak age at onset was 20-29 years in males and 30-39 years in females. In 6 (11%) patients, the lesions were preceded by itching. Of 55 patients, 32 (58%) noted only 1-3 patches while 12 (22%) patients had 4-6 patches and rest 11 (30%) had multiple (>6) patches to begin with.

Duration : The duration of the disease ranged from 2 months to 16 years. Males reported earlier (2 months to 8 years; mean 1.37 years) than females (3 months to 16 years; mean 2.06 years). Twelve (22%) patients had a disease of less than 6 months duration; in 27 (49%) patients, duration of the disease varied between 6 months to 3 years. While in 16 (29%) patients, the duration was more than 3 years

Site of onset : Face and neck were the most frequent initial sites of involvement (40 patients; 72%) in both sexes, followed by limbs and trunk. In 5 (9%) patients, lesions started over more than one site. In either sexes, preauricular (Fig.1) and temporal (Fig.2) regions were often the first sites to be involved.

Sites involved : Lesions were generalised in 43 (78%) patients and in rest 12 (22%) were localized. Forty nine (89%)

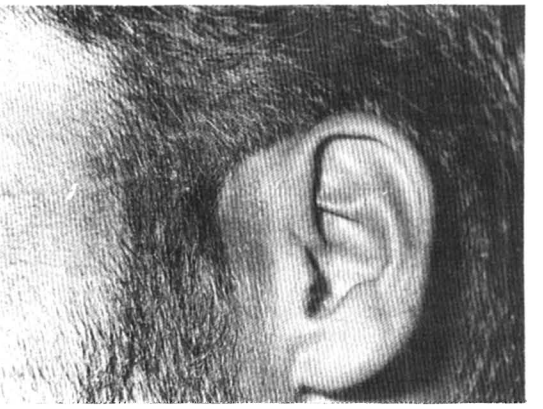

Fig. 1. Preauricular macular pigmentation, the commonest site of initial involvement.

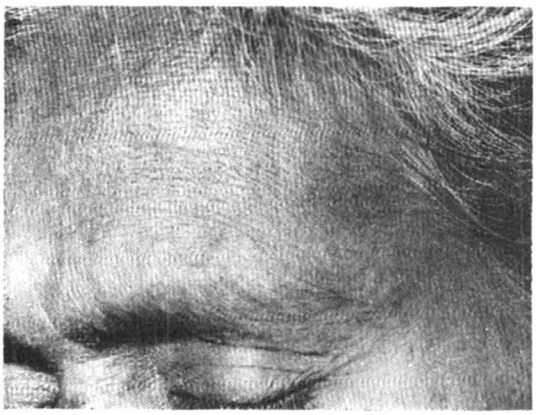

Fig. 2. Macular pigmentation of temple, often the first site to be affected.

patients, had bilateral symmetrical involvement. Face and neck were the commonest affected sites found in 51 (93%) patients either alone or in combination with other sites eg trunk (Fig. 3), limbs etc. Face

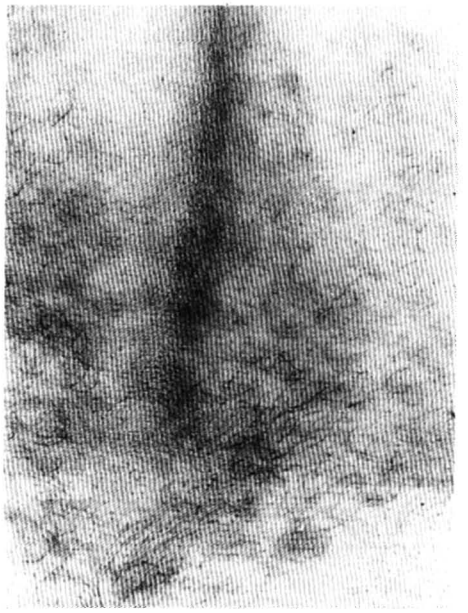

Fig. 3. Patchy pigmentation on lower part of back of trunk.

was completely spared only in 4 patients. Flexures like axillae, groins and inframammary folds were also involved besides neck. The cubital and popliteal fossae were, however, relatively spared. Only 1 patient had lesions below the knees. Palms and soles were spared in all the 55 patients (Table II). Perifollicular hyperpigmented tiny macules were seen in 6 patients (Fig.4).

Oral mucosae were involved in only 3 patients. Two had diffuse dark blue hyperpigmentation on buccal mucosae of both sides and lateral border of tongue and in the other patient, bluish pigmentation was

Table II. Sites of involvement in lichen dyschromicum perstans

Sites	No. of patients	%
Face	51	93
Forehead (including temple)	41	74
Cheeks	20	36
Chin	12	22
Nose	9	16
Periorbital	9	16
Neck	40	72
Trunk	19	34
Upper limbs	26	48
Lower limbs	6	11
Genitalia (males)	2	4
Flexure folds		
Axillae	10	18
Inguinal	3	5
Submammary (females)	6	11

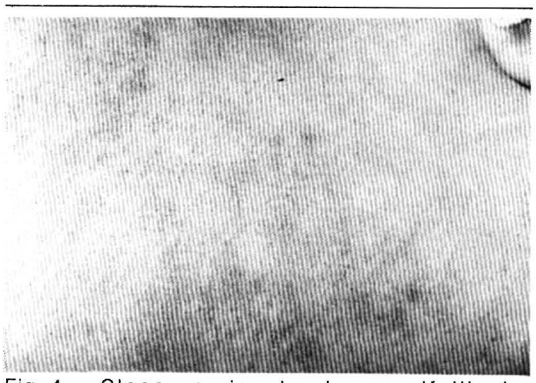

Fig. 4. Close up view to show perifollicular pigmentation.

speckled in nature and the sites affected were similar.

Nails were spared in all patients. Involvement of scalp was observed only in one patient in the form of diffuse hyperpigmentation over bald areas. Involvement of body surface area (BSA) ranged from less than 1% to 90%. Thirty (54%) patients had <10% BSA involvement; 17 (31%) patients had involvement between 10%-50%, while 6 (11%) patients had >50% BSA involvement. Only 2 (4%) patients had less than 1% involvement of body surface. Patients with shorter duration of the disease

had a lesser body surface involvement.

Colour and type of pigmentation : Shades of colours ranging from dark brown to black in varying proportions were observed in patients. No elevated red border suggestive of erythema dyschromicum perstans (EDP) around the pigmented patches was noticed in any patient. The various colour shades which were encountered were: bluish-black pigmentation in 26 (47%) patients; slate gray in 15 (27%), purplish or violaceous in 8 (15%), dark brown 4 (7%) and blackish in 2 (4%) patients. Lighter colours were seen in patients with a disease of shorter duration. In every patient only one pattern of colour was observed.

The pattern of pigmentation was diffuse in 37 (67%) patients, reticular in 13 (24%) and blotchy in 5 (9%) patients. Seven (13%) patients had perifollicular pigmentation mostly on arms and forearms.

Symptoms : Fourteen (25%) patients complained of mild to moderate itching and 12 (22%) observed darkening of pigmentation on exposure to sunlight. In 29 (53%) patients, the lesions were asymptomatic.

Occupation : Various occupations observed were: housewives-24, students-13, mechanical work-7, clerical job-5, teachers-4 and business-2. Occupation seemed to have no impact on the disease.

Precipitating factors : Psychological stress in 4 patients and physical stress in 2 patients were incriminated in the precipitation of the disease. History of use of cold cream and toiletries mostly pertained to females. However, these did not have any bearing on the initiation of the disease and neither was there any temporal correlation. History of use of mustard oil and other oils for body massage was available in 15 patients and in one, there was occupational exposure to cutting oils. However, there was no apparent relation of development or aggravation of pigmentation with the use of oils or toiletries in any patient. None of our patients had history of usage of hair dye.

Associated cutaneous and systemic diseases : Various other cutaneous diseases were observed in 18 patients. The naevoid conditions were seen in 5 patitents, vitiligo, acne vulgaris and dermatophytic infections in 4 patients each and scalp psoriasis in one patient.

Six patients had various systemic diseases in association. These were hypertension and diabetes mellitus in 2 patients each and hypothyroidism and pulmonary tuberculosis in one patient each.

No family history of similar skin pigmentation or lichen planus was obtained in any patient. There was no history of any metallotherapy or intake of known photosensitizing drugs in any of the patients. None of the female patients were on oral contraceptives.

Histopathology : Skin specimens available from 39 patients (including mucosal biopsies in 3 patients) showed certain epidermal and dermal changes common to all samples. The findings are highlighted in Table III (Figs 4-7). Section from 6 patients with perifollicular hyperpigmented lesions showed moderate to dense collection of perifollicular lymphomononuclear infiltrate in all 6 patients and basal cell degeneration of the hair follicle in 3 patients. Mucosal biopsies from 3 patients showed increased number of mealanophage and free melanin granules in lamina propria. Dermal collagen was normal and there was no vascular change. Amyloid material and iron pigment was absent in all the specimens. In most of the specimens (27/39) changes were mild to moderate in nature

Table III. Histologic features in lichen dyschromicum perstans (LDP) and in other related disorders

Histologic feature	Bhutani et al[4] LPP (n=40)	Vega et al[9] LPP (n=11)	Vega et al[9] AD (n=20)	Present study LDP (n=39)
Hyperkeratosis	-	10 (97)	16 (80)	6 (15)
Thinned epidermis	-	9 (81)	13 (65)	3 (8)
Basal cell vacuolization/ degeneration	*	10 (91)	17 (85)	30 (77)
Perivascular infiltrate	*	10 (91)	19 (95)	32 (82)
Band like infiltrate in the upper dermis	6 (15)	11 (100)	*	0 (0)
Melanophage	*	11 (100)	20 (100)	24 (62)
Colloid bodies	2 (5)	*	*	0 (0)

Figures in parentheses indicate percentage
* Number not mentioned, - Absent

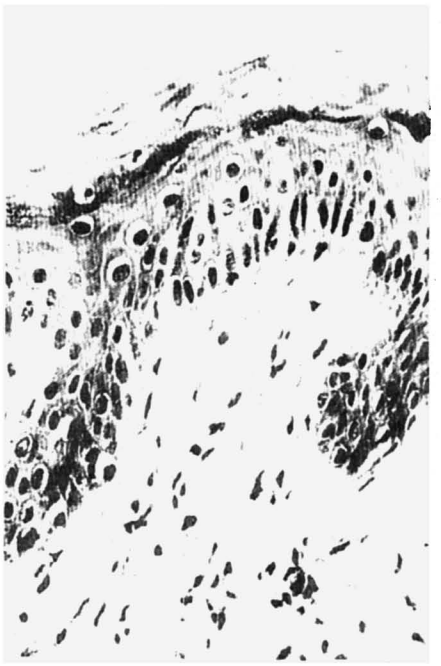

Fig. 5. Hyperpigmented basal keratinocytes (H&Ex550).

whereas in 12 specimens the changes were quite significant. Patients who showed significant histopathological changes mostly had darker pigmentation.

Direct immunofluorescence (DIF) : DIF was carried out in 14 (25%) patients. Of these

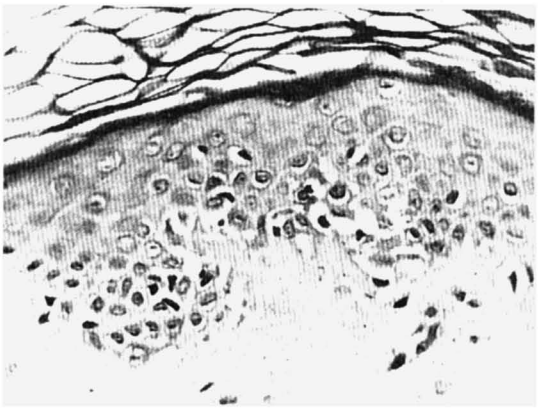

Fig. 6. Focal basal cell degeneration (H&Ex550).

only one patient showed deposition of IgM at dermoepidermal junction and one showed deposition of complement (C3) at the same site.

Discussion

A number of pigmentary disorders with clinical and histological features almost similar to the one described by us have been reported in the literature under various names such as 'dermatosis ceincienta' (ashy dermatosis)[1] or erythema dyschromicum perstans,[2] lichen pigmentosus[3] and lichen planus pigmentosus.[4] However, there are subtle differences and the exact relationship between all these entities is not clear. Ramirez in 1957[1] first described ashy dermatosis.

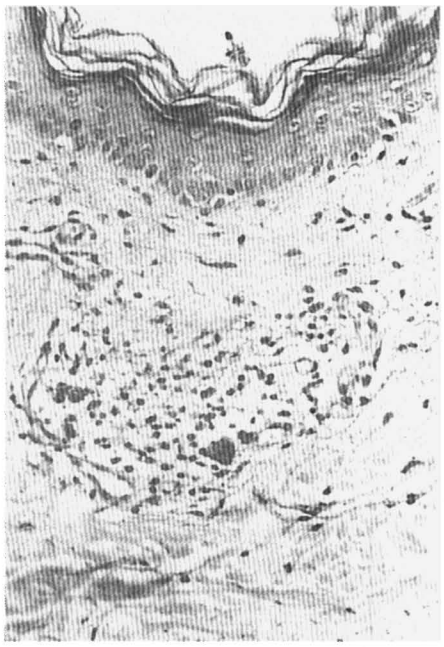

Fig. 7. Perivascular lymphomononuclear infiltrate in the upper dermis (H&E x 280).

Later Convit et al[2] observed a condition very similar to lesions of pinta in late phase; they called it erythema dyschromicum perstans (EDP). Ashy dermatosis (AD) or EDP is clinically characterized by blue-grey hyperpigmented macules of variable shapes and sizes. The lesions occasionally have an elevated active red border which leaves a residual hypopigmented halo. The distribution tends to be symmetrical and face, neck, trunk and upper extremities are commonly affected. The condition is chronic and occasionally pruritic. It is seen mainly in dark skinned individuals and no age or sexual predilection exists. A clinically and histologically similar condition was described by Shima et al in 1956 from Japan and was labelled as lichen pigmentosus.[3] It differed from EDP in that the lesions were smaller macules rather than ill-defined areas. The histological features also resembled atrophic lichen planus especially those forms reported in the French literature as 'lichen invisible pigmentogene'.[5] Bhutani et al 1974,[5] introduced a new entity which they labelled as lichen planus pigmentosus (LPP). It is characterized by hyperpigmented dark brown or slate grey macules distributed mostly over exposed areas and flexures. There are exacerbations and remissions occasionally accompanied by pruritus. The lesions lack the erythematous border seen in EDP. The clinical association of this disorder with lesions of classical lichen planus in about one third of the patients and demonstration of colloid bodies on histopathology prompted Bhutani et al to coin the term lichen planus pigmentosus. They suggested that ashy dermatosis is similar to LPP and it was further emphasized that AD is a macular variant of lichen planus.[6] Naidorf and Cohen[7] while agreeing with Bhutani et al suggested that the disorder should be called the erythema dyschromicum variant of lichen planus even though no erythema is seen and dyschromia is pigmentation. However, confusion still prevails and it is still controversial whether EDP and LPP are same. Vega et al in 1992[8,9] for the first time referred to the existing confusion and presented emphatically the clinical and histological differences between EDP and LPP. We also feel that EDP and LPP are distinct clinical entities.

Though several of the clinical and histologic features of the above hyperpigmentary disorder described by us fit into LPP, a notable exception is that none of our 55 patients had any preceding or concomitant lesions of lichen planus. As highlighted earlier, the term LPP suggests that the disorder has some relation to LP and it has been referred to as a variant of lichen planus peculiar to Indians.[10] The lack of lichen planus lesions in the patients presented

prompts us to name this disorder as lichen dyschromicum perstans (LDP). We feel that the term LDP is quite apt as it encompasses the clinical and histological features of the entity and clearly indicates that the disorder is unrelated to lichen planus. Moreover, retaining of the words, dyschromicum perstans reminds one that the disorder has some features of EDP viz, a varied pigmentation and persistent course. The term LPP should be reserved for those cases where in addition to hyperpigmented macular patches, the patient has either some lesions or history of lesions in past suggestive of LP.

Thus in conclusion, LDP is a distinct clinical entity characterized by pigmented lesions which are initially small, discrete, ill-defined oval to round macules which later become confluent to form large areas of pigmentation. The pigmentation varies from bluish-black to slate-grey or purplish. It may be diffuse or reticular. The lesions are only mildly pruritic and face and neck are the most frequent initial sites of involvement. Later the trunk and extremities may be involved. The palms and soles are spared and involvement of oral mucosae is infrequent. Both the sexes are affected. The course is chronic and persistent. No definite precipitating factors could be identified in any of the patients. None of the patients had any preceding or associated erythema around the lesions. No lesions of lichen planus were observed at any stage. The histopathological findings are same as observed in other disorders exhibiting a lichenoid tissue reaction.

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