Lichen planus pigmentosus, ashy dermatosis, erthyema dyschromicum perstans or acquired spontaneous macular hyperpigmentation?

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Abstarct

Introduction Clinical and histopathological features of acquired spontaneous macular hyperpigmentation appear to overlap in the conditions described as lichen planus pigmentosus, ashy dermatosis, lichen pigmentosus and erythema dyschomicum perstans.

Methods Ten consecutive patients with acquired hyperpigmented macules over 5 mm in size without a history of inflammation were studied. Salient points in the history and examination were recorded and biopsies were assessed in all the patients.

Results Age ranged from 4-60 years. Seven were females. None had a history of lichen planus or erythematous lesions prior to pigmentation. There was no history of contact dermatitis. Face was the commonest site involved but lesions were seen in non-exposed areas as well. Lesions were non itching, ashy gray macules with ill defined margins, sometimes becoming confluent. Size of lesions varied within and between patients. Pigmentary incontinence, epidermal thinning and mild hyperkeratosis were consistent features in all 10 biopsies. Paucity of inflammation was a notable feature in all cases. Focal basal cell degeneration was seen in 3 biopsies.

Conclusion Acquired spontaneous macular hyperpigmentation should not be linked to lichen planus when not associated with same. A consensus opinion on terminology is needed to avoid confusion as different workers seem to label the same condition by different names.

Introduction

Large macules of acquired hyperpigmentation without a history of inflammation or a drug eruption are uncommon. When it occurs it is troublesome, particularly in the dark skinned individuals. Clinical and histopathological features similar to each other have been described in conditions known as lichen planus pigmentosus (LPP), ashy dermatosis (AD), lichen pigmentosus and erythema dyschromicum perstans (EDD). Although some cases of acquired macular hyperpigmentation (AMH) may be associated with lichen planus (LP) it is not the case in most cases.

Aims

To study the clinical and histopathological features of acquired macular hyperpigmentation occuring without a history of inflammation.

Materials and methods

We studied 10 consecutive patients who presented to the dermatology clinics at General Hospital Matara (1993-1995) and General Hospital Kalutara (1996) with acquired hyperpigmented macules larger than 5 mm in size, without a prior history of inflammation. Lesions less than 5 mm were not included as a practical way of excluding small naevi, freckles and typical lesions following lichen planus. Fixed drug eruptions, pigmentation secondary to friction, melasma, freckles and naevi were excluded from the study on clinical examination. Size, site, distribution, margins and degree of pigmentation were the main clinical features recorded.

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A skin biopsy was done from a representative lesion in each of the patients. The specimens were fixed in 10% formol saline and stained with haematoxylin and eosin. The characteristic features of lichen planus were searched for in all biopsies. The other histological features of the epidermis and the dermis were recorded.

Results

The sex distribution was 3 Male: 7 Female. The age of the patients ranged from 4 years to 60 years. Duration of illness ranged from a few months to a few years at presentation. Size of lesions varied from small to large (0.5-10 cm). All the lesions had ill defined margins. Majority of patients had confluent lesions, appearing as large areas of dyspigmentation. The degree of pigmentation varied from patient to patient as well as within a given patient. In most patients the macules were slowly enlarging, and in some, the progression of the disease was very slow after an initial phase of activity. Two patients had slight scaliness but the lesions were not raised above the surface. Figure 1 illustrates a typical case with this condition.

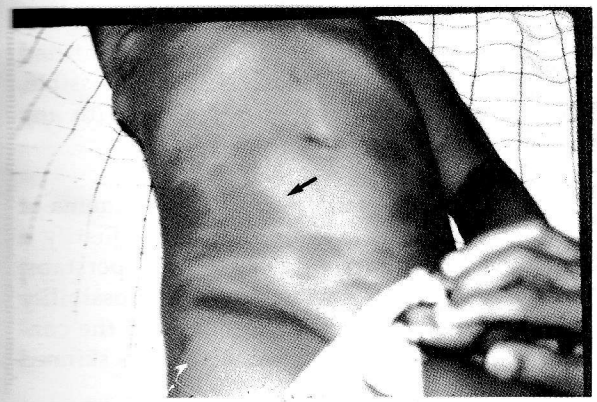

Figure 1. Hyperpigmented ill-defined macules in a four year old child

Face was involved in eight cases but all lesions were not limited to sun exposed areas. The lesions were variably scattered on the face, trunk and the limbs. Mucous membranes were not affected. Finger nails, toe nails, palms and soles were not affected. There was no pruritus of the lesions.

None of our patients had lichen planus at presentation nor developed same during the follow-up period which varied from 2 months to 3 years. They also did not show erythema at the edges of the lesions. Only two patients said that they used perfumes, but the areas of hyperpigmentation did not correspond to the areas where perfume had been used. There was no history of using oils on the body other than on the scalp. There was no uniform pattern of using a particular type of soap or an optical whitener. There was no history of exposure to any noxious chemicals. There was no significant drug history. There were no major coexisting systemic diseases in the patients studied.

There was no irregular acanthosis, hypergranulosis, diffuse basal cell degeneration or band like mononuclear cell infiltration of the dermis characteristic of LP. The main histopathological features of the biopsies are given in Table 1. Pigmentary incontinence, epidermal thinning and mild hyperkeratosis were the most consistent findings. There was no acanthosis, saw toothing, hypergranulosis, spongiosis, dermal oedema, vascular dilatation or eosinophilic infiltration in any biopsy. Figure 2 shows the histopathology of a representative case.

Table 1. Histopathologic features

Microscopic features	*Number*
Pigmentary incontinenance	10 (100%)
Epidermal thinning	10 (100%)
Minimal or no inflammation	10 (100%)
Hyperkeratosis	10 (100%)
Focal basal cell degeneration	3 (30%)

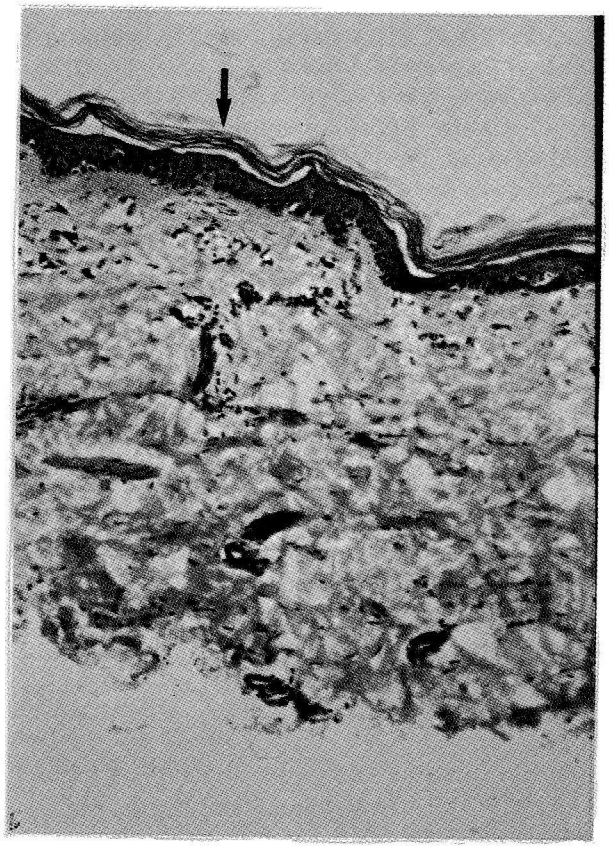

Figure 2. Photomicrograph of a skin biopsy stained with haematoxylin and eosin (x100) showing epidermal thinning, minimal inflammation, hyperkeratosis and pigmentary incontinence

Discussion

Clinically, the acquired spontaneous macular hyperpigmentation (ASMH) described in our patients was characterized by widespread bluish gray ('ashy') or dark, small and large macules occuring variably on the face, trunk and limbs. Some authors consider this to be a variant of erythema dyschromicum perstans[1]. Some others consider this to be a variant of lichen planus and thus call it lichen planus pigmentosus[2,3]. In Japanese medical publications a similar condition has been described as lichen pigmentosus[4].

Most reports of this clinical entity have been from India, South America, and the Middle East[1-9]. The lesions on the face also have some similarities with Riehl's Melanosis[9], although lesions were not limited to the face in our patients. Even some major dematopathology books use the terms EDD, AD and LPP interchangeably[10-12].

Histological differential diagnosis in our patients would include "burnt out" or atrophic lichen planus, post inflammatory pigmentation and the conditions described as ED, AD, and LPP. Pigmentary incontinence and minimal inflammation were conspicuous in all our biopsies. Features characteristic of LP, such as diffuse basal cell degeneration, hypergranulosis and irregular acanthosis were not seen. Thus there was not sufficient evidence to link the condition to lichen planus either clinically or histopathologically in our series.

Although some histologic features resemble features of "burnt out" lichen planus they need not necessarily mean that they are due to LP or a variant. The authors are in agreement with the concept that 'a lichen planus like or lichenoid picture is actually a cutaneous reaction rather than a single disease entity' as stated by Pinkus[4]. In most of our cases the size of lesions were much larger than the areas involved in plaques of lichen planus. It is well known, that following lichen planus darkly pigmented macules may persist in the affected areas. However none of our cases had a history of LP. Furthermore, lichen planus is universal, but the condition described here is most commonly reported in dark skinned races.

Some authors suspect that this dermatosis may be a response to some unidentified enviromental toxin[1]. Although this is possible, we could not find any relationship with any such factor in our patients.

None of the patients had any erythema at the edge or centre of the lesions to justify a diagnosis of erythema dyschromicum perstans. However one can not exclude the possibility that a slight erythema at the onset of the condition may have been missed by the dark skinned patients.

Pigmentation secondary to use of perfumes etc. is highly unlikely as most of our patients

did not use any. Even in the two who had used perfumes, the pigmentation did not correspond to those areas.

The time of presentation of patients varied from months to years after the onset of symptoms. Therefore in this study we cannot address the issue, whether the biopsy findings would have been different at the onset of the disease.

Lack of adequate response to any form of treatment is a constant disappointment to patients with AMH of this type. There has been a single report of use of high doses of vitamin A therapy in this condition[8]. However this form of therapy was not attempted in our patients. Bleaching agents such as hydroquinone 2% were disappointing. Topical steroids and hydroquinone reduced the intensity of hyperpigmentation in some patients.

There is no accepted classification for large macules of acquired spontaneous hyperpigmentation at the moment. In the quest for identifying the aetiologic factors of this type of hyperpigmentation it may be important to subdivide the pigmented patches according to the size. Small patches of hyperpigmentation may occur secondary to conditions such as pityriasis rosea, which may not be detected by the patient. Therefore some cases with clinico-histopathologic appearences described in this paper may be secondary to skin conditions which are not noted by patients while others may be due to a still unknown specific aetiology.

We propose that acquired spontaneous macular hyperpigmentation without coexisting or preceding inflammation should be separated from lichen planus and its variants. More research should be done on the aetiopathology of ASMH. It is opportune to have a consensus of opinion by a team of experts on the terminology of this type of hyperpigmentation to avoid confusion, as workers in different regions appear to name the same clinical entity by different names.

References

1. Naidorf KF, Cohen SR. Erythema dyschromicum perstans and lichen planus. *Archives of Dermatology* 1982; **118**: 683-685.

2. Bhutani LK, Bedi TR, Pandhi RK, Nayak NC. Lichen planus pigmentosus. *Dermatologica* 1974; **149**: 43-50.

3. Kark EC, Litt JZ. Ashy dermatosis -a variant of lichen planus? *Cutis* 1980; **25**: 631-633.

4. Pinkus H. Lichenoid tissue reactions, *Archives of Dermatology* 1973; **10**: 840-847.

5. Knox JM, Dodge BG, Freeman RG. Erythema dyschromicum perstans. *Archives of Dermatology* 1968; **97**: 262-272.

6. Sanchez NP, Pathak MA, Sato SS, Sanchez JL, Mihm MC, Fitzpatrick TB. Circumscribed dermal melaninoses: classification, light, histochemical and electron microscopic studies on three patients with erythema dyschromicum perstans type. *Inernational Journal of Dermatology* 1982; **21**: 25-31.

7. Verhagen ARHB, Koten JW. Lichenoid melanodermatitis. *British Journal of Dermatology* 1979; **101**: 651-658.

8. Bhutani LK, George M, Bhate SM. Vitamin A in the treatment of lichen planus pigmentosus. *British Journal of Dermatology* 1979; **100**: 473-474.

9. Bleehan SS, Ebling FJG, Champion RH. Disorders of skin colour In: Champion RH, Burton JL, Ebling FJG eds. Text book of Dermatology. Fifth ed. London: Blackwell Scientific Publications 1992; 1595-1597.

10. Weeden D, Systemic Pathology/The skin, Third edition, Churchil Livingstone, Edinbourough: 1992; 35-37.

11. Ackerman AB. Histologic Diagnosis of Inflammatory Skin Diseases, Second edition, Baltimore: Williams and Wilkins Company, 1997; 502-507.

12. Lever WF. Schaumburg-Lever G. Histopathology of the Skin. 7th ed. Philadelphia: JB Lippincott; 1990; 154-171.