Defining erythema dyschromicum perstans, ashy dermatosis, lichen planus pigmentosus and idiopathic eruptive macular pigmentation: A global consensus needed

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The conditions erythema dyschromicum perstans (EDP), ashy dermatosis, lichen planus pigmentosis (LPP) and idiopathic eruptive macular pigmentation (IEMP) are commonly confused and poorly defined. It is debated whether they are indeed discrete entities, or variations in presentation and description of the same disease process[1]. Part of the confusion stems from the poor understanding of the pathogenesis of these conditions.

Ashy Dermatosis was first described as 'Dermatosis Cenicienta'[2] by Ramirez, who reported 139 patients with macular grey hyperpigmentation, sometimes with an erythematous border. Histopathologically, basal vacuolar change was seen, in addition to dermal melanosis and a perivascular inflammatory infiltrate. The name erythema dyschromicum perstans was used by Convit, Kerdel-Vegas and Rodriguez-Garcilazo in 1961 to describe what many consider to be the same condition[3,4]. The subtle erythema described in typical cases of EDP is not usually seen in darker skinned patients. Most authors agree that erythema dyschromicum perstans and ashy dermatosis are the same condition[9,10,11,12], however this is not universally accepted[13,15].

IEMP was originally described by Degos[5]. The main differences between EDP/ashy dermatosis and IEMP were proposed to be an absence of preceding erythema and an absence of vacuolar interface change[5,6]. It has also been argued that IEMP can be differentiated from EDP due to a brownish, rather than grey pigmentation[14]. Histopathologically IEMP is described as showing only basal keratinocytic hyperpigmentation, in contrast to the vacuolar change and pigmentary incontinence seen in EDP/ashy dermatosis. Some authors however disagree with the distinction of these conditions based on clinical colour or histopathology[7], suggesting that the presence of basal vacuolar change depends on the time of biopsy in relation to the activity of the condition. The diagnosis of IEMP has been further complicated by the description IEMP with papillomatosis, where some lesions show velvety thickening[17]. In a strict sense this description is a contradiction to the 'macular' description of IEMP.

Based on the presence of interface change on histopathology, and a similar clinical appearance to 'burnt out' lichen planus, it has further been suggested that EDP/ashy dermatosis may be variants of lichen planus and be the same condition as lichen planus pigmentosus[3,8], though some argue against this suggested grouping[10,16]. Most reported cases of EDP/ashy dermatosis have no other evidence of typical lichen planus. EDP and AD most commonly present in children, in whom lichen planus is uncommon. EDP does not show Max-Joseph spaces on histopathology[8], though this could be explained by the temporal variation hypothesis.

The disagreement between authors in regards to the ways in which to differentiate or unify EDP, ashy dermatosis, lichen planus pigmentosus and IEMP suggest that the clinical and histopathological presentations of these conditions are somewhat blurred[16]. In addition, there is a poor understanding of the pathogenesis of this group of conditions and treatment for all is generally ineffective[8].

We suggest that until further evidence is accrued, the terms EDP and ashy dermatosis be considered to describe a morphological spectrum of acquired macular pigmentation associated with evidence of a current or resolved vacuolar interface dermatitis with post-inflammatory pigment alteration. Patients with clinical or histopathological features of lichen planus with post inflammatory pigment alteration can be described as having lichen planus pigmentosus. Frequent resolution and the finding of basal hyperpigmentation without significant pigmentary incontinence suggest that IEMP may have a different aetiology. From a pathological standpoint, familiarity with racial and individual variations in pigmentation and close correlation with the clinical findings and biopsy site are critical to microscopic assessment of these findings. It is accepted that these terms may describe slight clinical and histopathological variations in the same disease process, or several disease processes which present in a similar manner (acquired macular pigmentation).

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More critical from a clinical standpoint is that these idiopathic and treatment resistant conditions be separated from other causes of hyperpigmentation with identifiable specific and potentially treatable causes and those with important systemic implications. To this end, the important clinical and histological differential diagnoses are outlined in Tables 1 and 2 respectively. Figure 1 shows a proposed simplified clinical diagnostic flow chart of the approach to these conditions based on published data and authors' synthesis. This proposed algorithm helps to categorize acquired macular hyperpigmentation due to unknown causes.

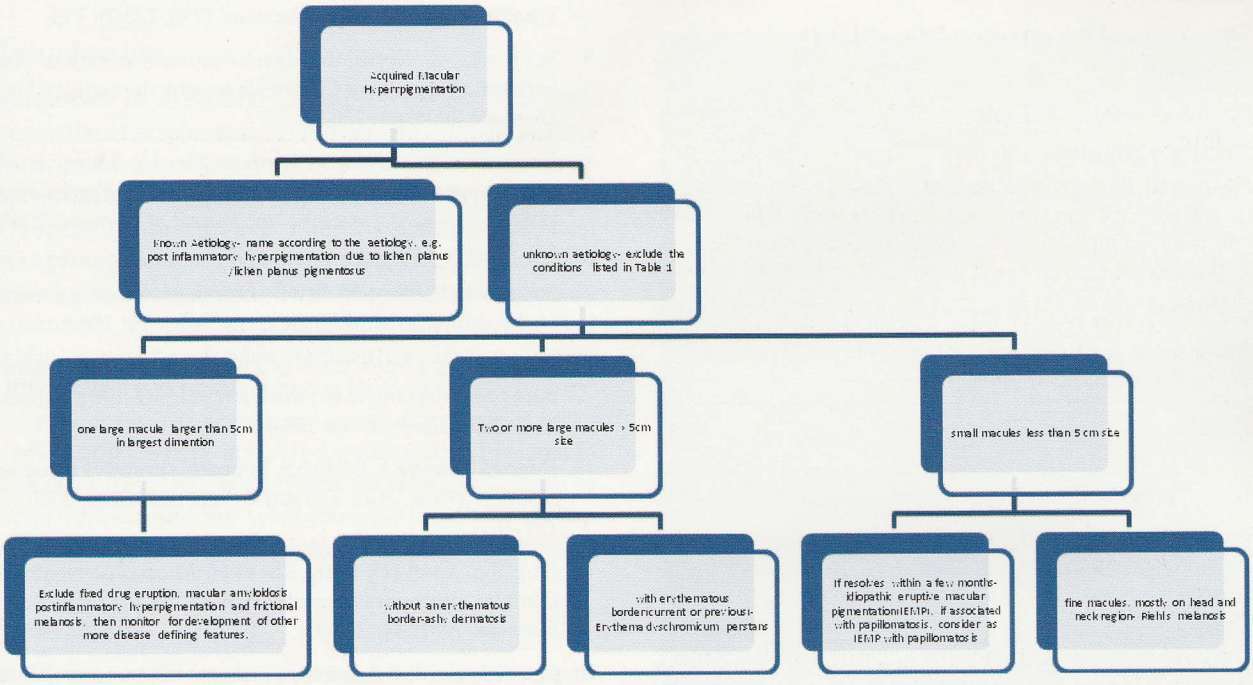

Figure 1. *Proposed algorithm for acquired macular hyperpigmentation based on current published data and authors' synthesis.*

Table 1. Clinical differential diagnoses of EDP, AD, LPP, IEMP

Post inflammatory
Fixed drug eruption
Pigmented contact dermatitis/ Riehl's melanosis
'Burnt out' graft versus host disease
Post inflammatory hyperpigmentation, e.g. viral exanthem, pityriasis rosea

Systemic disorders
Addison's disease
Mastocytosis
Pigmentation secondary to dermatomyositis

Non-melanin pigmentation
Argyria
Drug induced pigmentation (e.g. amiodarone, minocycline)
Ochronosis

Others (miscellaneous)
Mycosis fungoides
Melasma
Hori's nevus
Macular amyloidosis
Confluent and reticulate papillomatosis
Melanoderma secondary to advanced melanoma
Aberrant persistent Mongolian blue spots
Phacomatosis pigmentovascularis
Pigmented seborrhoeic keratosis
Pityriasis versicolor
Ephelides
Solar lentigo
Post-radiotherapy hyperpigmmentation
Erythema ab igne
Phytophotodermatitis
Dowling-Degos disease

Table 2. Histopathological mimics of EDP/AD/LPP

Resolved benign lichenoid keratosis
Fixed drug eruption
Riehl's melanosis/ pigmented contact dermatitis
Resolved drug eruption (post inflammatory hyperpigmentation)
Drug induced hyperpigmentation (slowly progressive/ ongoing)
Resoved viral exanthem
Dermatomyositis
"Burnt out" graft versus host disease
Ephelides
Lentigens
'Normal skin' with occasional dermal melanophages (in dark skinned races)

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