

Case report

Idiopathic eruptive macular pigmentation

Viviane Marins de Arruda Câmara, MD, Omar Lupi, MD, PhD, and Juan Piñeiro-Maceira, MD, PhD

From the Departments of Dermatology and Pathology, Federal University of Rio de Janeiro, and Instituto de Dermatologia Prof. Rubem Azulay, Santa Casa do Rio de Janeiro, Brazil

Correspondence

Omar Lupi, MD, PhD
Rua Frei Leandro, 16 apt 501
Rio de Janeiro/RJ 22.470-210
Brazil
E-mail: omarlupi@globo.com

A 23-year-old, dark-skinned man presented at the dermatology department with pigmented macules over the trunk and proximal thighs of 2 months' duration. He reported that the number of lesions had increased progressively for a few weeks and then remained stable. Skin examination showed oval brown macules and patches, 5–40 mm in diameter, involving mainly the anterior trunk and proximal thigh, but also the neck and dorsum (Fig. 1). The lesions were asymptomatic. There was no previous history of an inflammatory process, erythema, scaling, or drug intake. The mucous membranes, palms, and soles were clear. The pigmentation was not influenced by sunlight. Darier's sign (urticaria or erythema around the macules after scratching or rubbing of the lesions) was absent. The previous use of emollients, keratolytics, and topical antifungal agents did not alter the aspect of the lesions. The results of physical examination and routine laboratory tests (blood cell count, blood chemistry) were normal. Venereal Disease Research Laboratory (VDRL) test and direct skin examination and culture for fungus were negative. A biopsy specimen was obtained from a pigmented macule. The histologic study showed hyperkeratosis and acanthosis, epidermal basal layer pigmentation with an irregular distribution, focal mild lymphohistiocytic infiltrate in the papillary dermis and dermal–epidermal junction, and discrete pigmentary incontinence (Figs 2–4). The mast cell population was normal. A further biopsy 1 year later showed similar findings.

After 3 years of follow-up, mild spontaneous fading occurred in some of the trunk lesions, but most remained unchanged.

Discussion

The term “idiopathic eruptive macular pigmentation” (IEMP) was first proposed by Degos *et al.*¹ in 1978 to designate cases of a pigmented dermatosis observed by themselves and other authors, previously published under various terms.

The disorder is characterized by asymptomatic pigmented macules, involving the neck, trunk, and proximal limbs. Most cases have been published in the French literature^{1–4} and involve children and adolescents. The first reference in the English literature was in 1996, when de Galdeano *et al.*⁵ published five cases of IEMP and established diagnostic criteria. Recently, additional cases have been reported.^{6–9} We report a case of IEMP in a young male adult.

The age of onset of the disease ranges from 1 to 31 years. Males and females are affected equally (Table 1). The eruption is composed of oval, circumscribed, homogeneous pigmented macules that appear without previous erythematous, papular, or hypochromic lesions, mostly on the trunk, neck, and proximal limbs (Table 1). Pruritus is absent. No specific treatment has been proposed. The lesions tend to resolve spontaneously, although one case only resolved after 11 years' duration,¹ and one patient showed two periods of remission but still has lesions after 21 years of follow-up.⁷ The most common histo-

logic features are epidermal thickening with basal cell pigmentation with an irregular distribution, discrete pigmentary incontinence, and mild inflammation with focal lichenoid changes. The histology is similar at different times in the evolution of the disease.

Twenty-five years after the first report, the pathogenesis of the disease remains unclear. Sunlight is not important, as most lesions occur in photoprotected areas. Hormonal factors may be involved in increased pigment production, as most patients are children or young adults.⁶ Clinically, inflammation is not prominent, but a subclinical focal interface inflammatory process is demonstrated in the histopathologic analysis; a subclinical interface inflammatory process cannot be discounted.⁵

According to de Galdeano *et al.*,⁵ the following criteria should be fulfilled for the diagnosis of IEMP.

- 1 Eruption of brownish, nonconfluent, asymptomatic macules involving the trunk, neck, and proximal extremities in children and adolescents.
- 2 Absence of preceding inflammatory lesions.
- 3 No previous drug exposure.
- 4 Basal cell layer hyperpigmentation of the epidermis and prominent dermal melanophages without visible basal layer damage or lichenoid inflammatory infiltrate.
- 5 Normal mast cell count.



Figure 1 Multiple brown macules involving the trunk

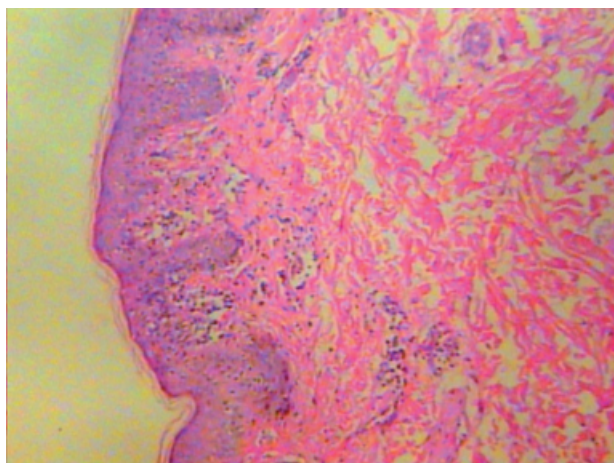


Figure 2 Mild inflammation in the papillary dermis and dermal-epidermal junction with focal lichenoid changes (hematoxylin and eosin, $\times 40$)

IEMP must be differentiated from drug eruption, postinflammatory pigmentation, mastocytosis, lichen planus pigmentosus, and erythema dyschromicum perstans (ashy dermatosis).

Drug eruption and postinflammatory pigmentation are excluded by a lack of history of medication use or of clinical

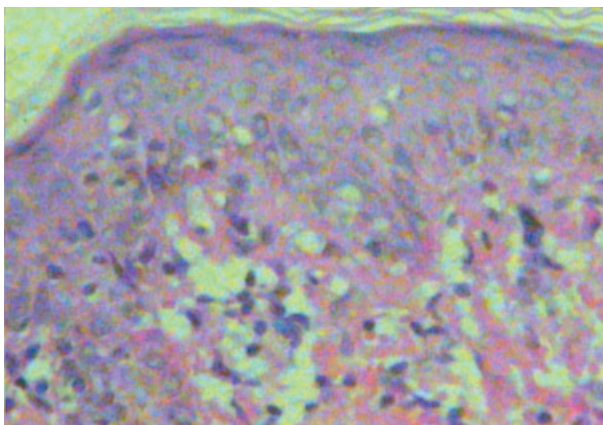


Figure 3 Mild inflammation in the papillary dermis. Melanin pigmentation with an irregular distribution in the epidermal basal layer (hematoxylin and eosin, $\times 250$)

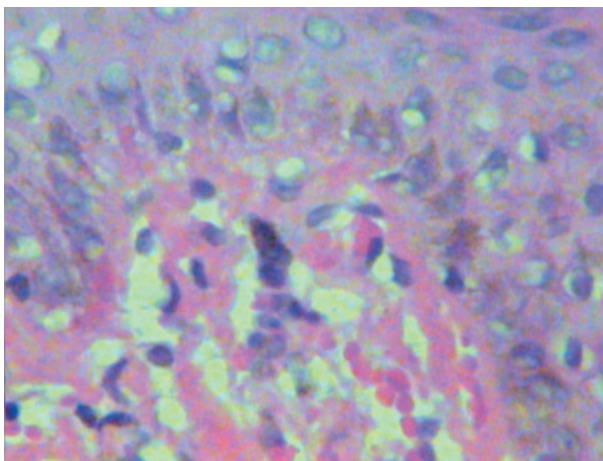


Figure 4 Detail of the epidermal basal layer showing irregular melanin pigmentation; there are a few extravasated red cells and a melanophage in the papillary dermis (hematoxylin and eosin, $\times 400$)

manifestations of a previous dermatosis that commonly affects the dermal-epidermal interface, such as lichen planus, benign lichenoid keratosis, and erythema multiforme.⁶

The normal number of mast cells and absence of Darier's sign exclude a diagnosis of mastocytosis, an important differential diagnosis in the pediatric population.

Lichen planus pigmentosus is an uncommon variant of lichen planus characterized by hyperpigmented dark-brown macules in sun-exposed areas.¹⁰ Erythema dyschromicum perstans is an idiopathic hypermelanosis with an ashy color that may coalesce and present an erythematous raised border. Some authors believe that these entities are clinical variants of

Table 1 Gender, age, and clinical characteristics of cases of idiopathic eruptive macular pigmentation

Reference	Sex/age (years)	Lesions	Location	Duration
Degos <i>et al.</i> ¹	M/12	Brown macules	Trunk, neck	11 years
Degos <i>et al.</i> ¹	F/12	Gray and brown macules	Trunk, neck, arm	–
Degos <i>et al.</i> ¹	M/15	Gray and brown macules, 10–20 mm	Trunk, neck, arms, dorsum	7 years
Degos <i>et al.</i> ¹	F/19	Brown erythematous macules	Trunk, arm, legs	> 7 months
Degos <i>et al.</i> ¹	M/9	Brown macules, 15–20 mm	Trunk, neck, arm, legs	2 years
Degos <i>et al.</i> ¹	M/18	Gray macules	Trunk	> 1 year
Degos <i>et al.</i> ¹	F/17	Blue–gray macules	Trunk, neck, arm, legs	1.5 years
Dupre <i>et al.</i> ³	F/24	Gray macules, 10–20 mm	Trunk, arms, legs	2.5 years
Dupre <i>et al.</i> ³	M/9	Brown macules, 10–20 mm	Trunk	2 years
Plantin <i>et al.</i> ⁴	M/4	Blue–gray macules, 20–30 mm	Trunk, neck, legs	> 6 months
de Galdeano <i>et al.</i> ⁵	F/16	Brown macules, 8–10 mm	Trunk, neck, legs	2 years
de Galdeano <i>et al.</i> ⁵	M/6	Macules, 5–25 mm	Trunk, neck, arm, legs	1 year
de Galdeano <i>et al.</i> ⁵	F/2	Light-brown macules, 3–4 mm	Trunk, neck, arm, legs	> 4 years
de Galdeano <i>et al.</i> ⁵	F/11	Brown macules, 5–10 mm	Trunk	2 years
de Galdeano <i>et al.</i> ⁵	M/5	Brown macules, 5–20 mm	Trunk	1 year
Jang <i>et al.</i> ⁶	F/4	Brown macules, 3–5 mm	Trunk, face, arm, legs	1 year
Jang <i>et al.</i> ⁶	M/17	Brown macules, 8–13 mm	Trunk, legs	3 years
Jang <i>et al.</i> ⁶	M/8	Brown macules, 3–5 mm	Trunk, arm, legs	2 months
Jang <i>et al.</i> ⁶	M/10	Gray macules, 5–10 mm	Trunk	3 months
Jang <i>et al.</i> ⁶	F/10	Brown patches, 10–20 mm	Trunk, neck, arm, legs	6 years
Jang <i>et al.</i> ⁶	M/12	Brown macules, 3–10 mm	Trunk	5 years
Jang <i>et al.</i> ⁶	M/16	Brown macules, 5–20 mm	Trunk, neck, arm, legs	1 year
Jang <i>et al.</i> ⁶	F/20	Brown macules, 3–7 mm	Trunk, neck, arm, legs	4 years
Jang <i>et al.</i> ⁶	M/7	Dark-brown patches, 10–30 mm	Trunk, face, arm, legs	> 6 years
Jang <i>et al.</i> ⁶	F/1	Dark-brown patches, 5–20 mm	Trunk, neck, arm, legs, face	2 years
Mehta <i>et al.</i> ⁷	F/3	Brown macules, 5–20 mm	Trunk, neck, arm, legs, face	> 21 years
Milobratovic <i>et al.</i> ⁸	F/31	Brown macules, 20–70 mm	Trunk, arm, legs	> 2 years
Trcko <i>et al.</i> ⁹	F/10	Brown macules	Trunk, arm, legs	2 years
Present case	M/23	Brown macules, 5–40 mm	Trunk, neck, legs, dorsum	> 3 years

the same disease,¹¹ as the histologic findings are similar.^{12,13} Ultrastructural investigation in IEMP shows numerous mature melanosomes in basal and suprabasal keratinocytes and macrophages containing clustered melanosomes, but no vacuolar pattern of the basal cell layer, discontinuity of the basal lamina, colloid bodies, or a lichenoid infiltrate that might indicate a diagnosis of lichen planus pigmentosus or erythema dyschromicum perstans.^{5,13}

It is important to consider IEMP in the differential diagnosis of pigmented disorders of the skin. It affects children and adolescents most frequently, but can be found in young adults. The rarity of reported cases may be related to the medical unfamiliarity with this entity. Different clinical and histopathologic patterns support IEMP as a separate entity, distinct from other pigmentary disorders.

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