Case for diagnosis *

Caso para diagnóstico

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CASE REPORT

Male patient, 41- years-old, born and living in Belo Horizonte (Minas Gerais), noticed nine months previously asymptomatic skin lesions on his face, trunk and limbs. Schizophrenia was a comorbid condition for which the patient has been treated with haloperidol 10mg/day for 15 years. No similar cases reported in patient´s family.

The dermatological examination revealed extensive and confluent grayish macules with erythematous raised borders affecting the face, neck, trunk and proximal parts of the limbs (Figures 1-3).

Laboratory tests were normal. Histopathologic examination showed irregular atrophy of the epidermis and irregular *foci* of hydropic degeneration of the basal layer, as well as lymphohistiocytic perivascular inflammatory infiltrate and pigment incontinence (Figure 4).

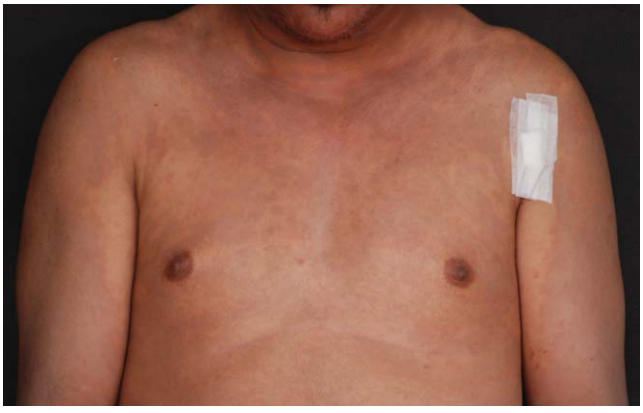

FIGURE 1: Hyperpigmented macules, extensive and confluent, affecting the face, neck, trunk and proximal limb

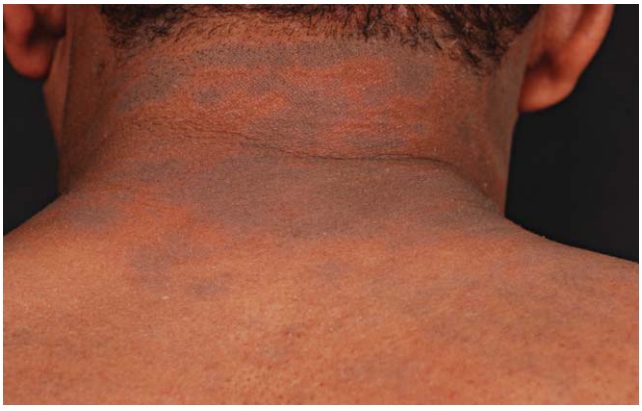

FIGURE 3: Gray macules on the posterior neck area

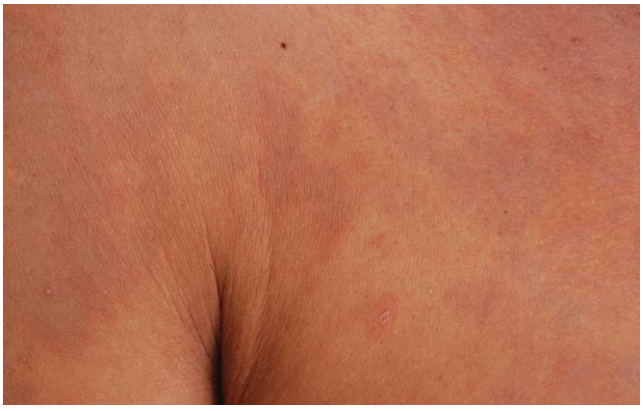

FIGURE 2: Close-up of perilesional erythematous halo

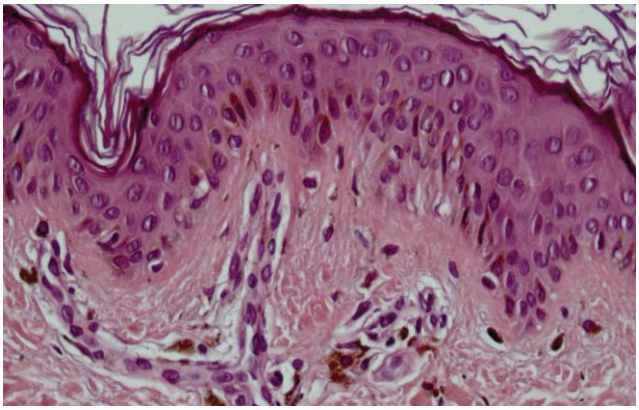

FIGURE 4: Atrophic epidermis, foci of degeneration of the basal layer and pigmentary incontinence (HE, 100x)

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DISCUSSION

Erythema perstans dyschromicum or ashy dermatosis is a rare chronic, benign disorder belonging to the group of acquired idiopathic hypermelanoses.[1-3] Initially described by Ramirez in 1957 (in El Salvador), the condition tends to be more prevalent in Central and South America,[1,2,4] primarily affecting young adults (under 20), with a slight preference for females and darker-skinned individuals. [1-3]

The etiology remains elusive, but associations with endocrinopathies, nematode infestations, pesticide exposure, HIV infection, allergy to cobalt and radiological contrast material have been reported. [2,5] The presence of HLA-DR4 *0407 allelic subtype is a risk factor in Mexican mestizos. [3] It is believed that the immune system is involved due to the presence of interleukins and inflammatory mediators in the lesions. [6]

The clinical picture is characterized by hyperchromic macules, blue-grayish, oval or polycyclic, with erythematous elevated borders with a diameter measuring from 3 mm to several centimeters. The lesions are asymptomatic, with centrifugal growth. The disorder tends to affect mainly the trunk and upper extremities, sparing the mucous membranes, palms, soles and scalp. [2]

Histopathologic findings are nonspecific and include vacuolization of the basal layer, necrosis of basal keratinocytes, colloid bodies, exocytosis of lymphocytes, pigmentary incontinence and perivascular lymphocytic inflammatory infiltrate. [6]

The differential diagnosis includes lichen planus pigmentosus, post-inflammatory hyperpigmentation, figurate erythemas, fixed drug eruption, Addison's disease and hemochromatosis. Among these, lichen planus pigmentosus is the most clinically and histologically similar to ashy dermatosis, which has resulted in it being thought to be a variant of lichen planus. At present, however, they are considered separate entities by most authors. [7]

Evolution of the disorder is chronic and benign and spontaneous remission is rare in adults. In children clinical improvement can tend to occur within two to three years. [8,9] Many treatments have been proposed but there is no standard therapy and the results are in any case inconsistent. Reports exist of improvement with dapsone and clofazimine, as well as with other regimens such as systemic and topical corticosteroids, antibiotics, griseofulvin, isoniazid, antimalarials, keratolytic agents, phototherapy and psychotherapy. [1,8,10] ❑

Abstract: Dyschromicum erythema perstans, or ashy dermatosis, is a rare chronic acquired skin disease characterized by gray hyperpigmented patches with erythematous borders. Its etiology is unknown and there is no specific treatment for the condition. We report a case of ashy dermatosis in a 41-year-old patient with extensive lesions on the trunk and limbs.
Keywords: Drug eruptions; Hyperpigmentation; Lichenoid eruptions; Melanins

Resumo: *Erythema dyschromicum perstans* ou dermatose cinzenta é enfermidade cutânea adquirida, rara, de evolução crônica, caracterizada por máculas acinzentadas, com bordas eritematosas. A etiologia ainda é desconhecida, não havendo tratamento definido para a afecção. Apresenta-se um caso desta dermatose em paciente de 41 anos, com lesões disseminadas no tronco e membros.
Palavras-chave: Erupção por droga; Erupções liquenóides; Hiperpigmentação; Melaninas

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