Case report

ERYTHEMA DYSCHROMICUM PERSTANS

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ABSTRACT

We describe two cases of erythema dyschromicum perstans, one of recent onset and the other present for 6 years, which illustrate the histological features that characterize the early and late stages of this disease. Surface microscopy proved to be a useful in vivo technique for the differential diagnosis of these EDP lesions.

KEY WORDS
erythema dyschromicum perstans, dermatosi cenicienta, skin hyperpigmentation.

INTRODUCTION

Erythema dyschromicum perstans (EDP) was first reported by Ramirez in 1957 who used the term "dermatosi cenicienta" (ashy dermatosis) to describe the characteristic ash-gray lesions he observed in subjects from El Salvador. The disease was later reported in other Latin American countries, as well as in the United States and Europe (2,9). The disease is characterized by the appearance of numerous, blue-gray macules on the trunk, limbs and face, which remain for indefinite periods of time. During the initial inflammatory phase, the borders of the plaques are erythematous.

Case reports

Case 1: A 32-year-old male was referred to our outpatient clinic for numerous, painless gray lesions 3-5 cm in diameter, that were asymmetrically distributed over the entire trunk. The round, flat lesions presented well defined borders and smooth, non-desquamated surfaces (Fig. 1). There were no signs of infiltration. The patient reported that the macules, which were initially erythematous, had appeared suddenly, without apparent cause, three months prior to our examination. After approximately two months they began to assume an ashy gray color. The patient denied any recent infection episodes, allergies, or drug use; he could recall no significant changes in diet or conditions of emotional or physical stress. Previous treatment with topical steroids and hydroquinone-preparations had no effect whatsoever on the lesions.

Blood chemistry studies, routine radiological exams, and sex hormone levels were all within normal limits. Assays to detect organ-specific and non-organ-specific autoantibodies and skin deposits of antibodies were negative. The Wood's

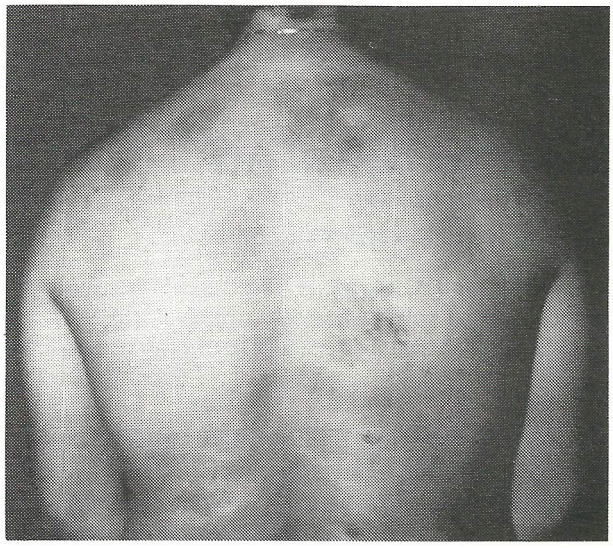

Fig. 1.: Case 1: numerous flat lesions of gray color, 3 - 5 cm in diameter were distributed over the entire trunk.

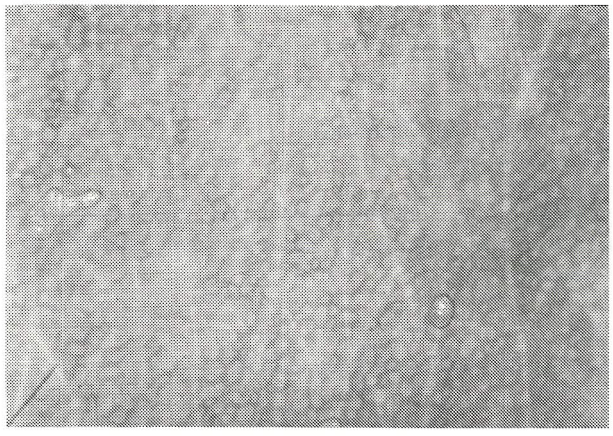

Fig. 2.: Case 1: surface microscopy revealed oval shaped areas of brownish pigmentation forming a network

light examination was normal.

Surface microscopy revealed oval-shaped areas of brownish pigmentation distributed in regular networks (Fig. 2).

Histological examination of one of the pigmented lesions (Fig. 3) showed vacuolization of the cells of the basal layer with perivascular infiltrates (Fig. 4) of lymphocytes and histiocytes; melanophages were also present within the infiltrates.

The lesions have remained stable during the six months we have been following this patient and there have been no changes in surface microscopy findings.

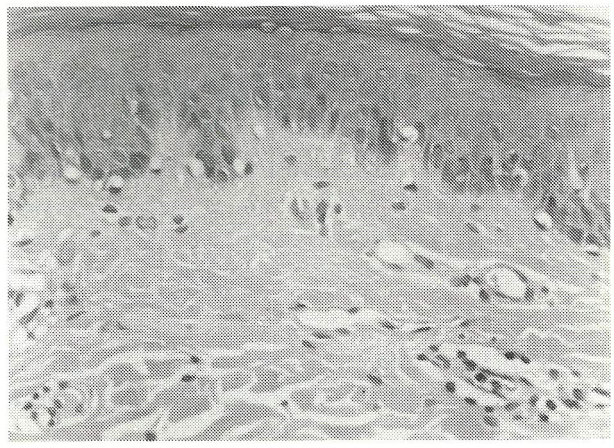

Fig. 3.: Case 1: histopathology: vacuolization of cells in the basal layer, an increased number of melanocytes.

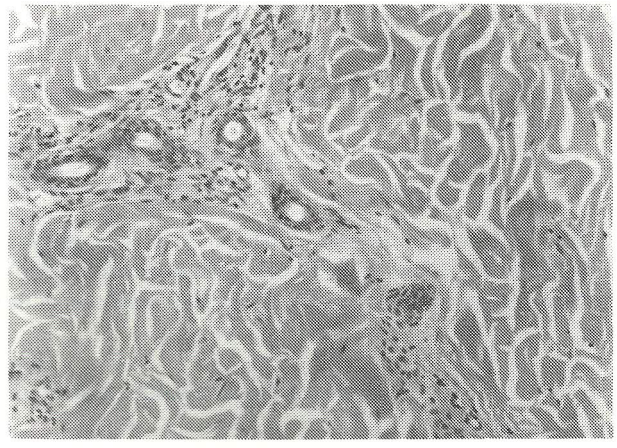

Fig. 4.: Case 1: histopathology: perivascular infiltrate composed of lymphocytes, histiocytes and a few melanophages.

Case 2: The patient was a 23 year-old female. Her parents were non-consanguineous and she had been born at term. At the age of 18, she had noted brownish plaques on her back following sun exposure. The lesions were not associated with any subjective symptoms and resolved spontaneously within few months after their appearance. However, new lesions had subsequently appeared on the neck, trunk, abdomen and arms (Fig. 5). Confluence of the lesions produced a reticular pattern. The patient's sister reportedly had the same type of lesions, but she unfortunately refused to be examined by our staff.

The clinical features presented by this patient were suggestive of a number of cutaneous pigmentary disorders. Studies were begun to rule out the presence of autoimmune

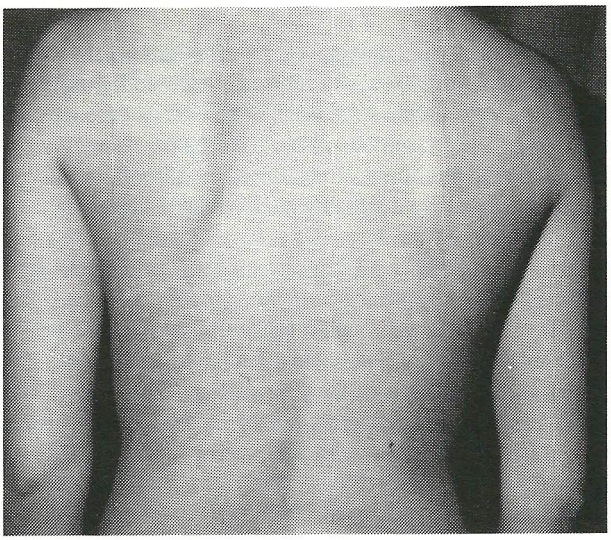

Fig. 5.: Case 2: brownish pigmentation forming a reticular pattern

disorders, neoplastic processes and systemic disease. Routine blood chemistry results, serum complement fractions, serum ACTH levels and thyroid function were all within normal limits; assays for LE cells, autoantibodies (ANA, ASMA, anti-DNA), S100 protein and direct immunofluorescence on lesional skin for IgG, IgA, IgM, C3 and fibrinogen all yielded negative results. Numerous radiological studies, ultrasound tomography of the thyroid, kidneys and adrenals, venous doppler studies, esophagogastroduodenoscopy, esophageal manometry, esophageal clearance and gastric fat absorption were all normal as well. Serum cortisol levels were decreased (20 ng/ml - normal values: 50-200 ng/ml) and assays for APCA autoantibodies were positive (+++). Ultrasound tomography of the pelvis revealed a micropolycystic right/left ovary. Surface microscopy showed slightly enhanced skin transparency.

Histological examination of a lesional biopsy revealed papillomatosis with hyperpigmentation and an increased number of basal melanocytes. There was focal edema of the basal keratinocytes with orthokeratotic hyperkeratosis. Numerous melanophages were seen within the papillary dermis and around the vessels.

COMMENT

Erythema dyschromicum perstans is a rare disease that can be classified among the cutaneous hypermelanoses (Tab. 1). Diagnosis is often difficult, as in Case 2, because the pattern of hyperpigmentation is similar to those produced by a variety of other diseases, both cutaneous and systemic.

EDP must be differentiated from acromelanosis and Dohi's reticular acropigmentation, both of which produce lesions on the digits with possible involvement of the limbs as well.

Table 1. Differential diagnosis concerning erythema dyschromicum perstans

Acromelanosis
Congenital dyskeratosis
Heterochromia extremitatum
Dohi's reticular acropigmentation
Anomalous flexural reticular pigmentation (Dowling-Degos disease)
Kitamura's reticular pigmented acrodermatitis
Naegeli-Franceschetti-Jadasshon syndrome
Idiopathic eruptive macular hyperpigmentation
Incontinentia pigmenti
Reticular pigmented dermatopathy
Diffuse congenital mottling of the skin
Progressive familial hyperpigmentation
Prurigo pigmentosa
Ziprowski-Margolis syndrome
Scleroderma

Kitamura's reticular acropigmentation (also reffered to as Dowling-Degos disease) (4, 5, 6) is characterized by hyperpigmented epheloid (freckle-like) lesions arranged in network-like patterns and by an increased number of melanocytes.

Prurigo pigmentosa, which initially presents with intensely pruritic vesicular papules on the trunk, also produces a brown-colored network of pigmentation in its advanced stages.

Idiopathic eruptive macular hyperpigmentation is characterized by asymptomatic, blue-gray macules on the trunk, neck and proximal portions of the limbs, but these lesions, unlike those of EDP, regress spontaneously; vacuolar degeneration of the basal layer is also absent in this idiopathic form.

Incontinentia pigmenti is almost exclusively seen in females. The hyperpigmentation is distributed along Blaschko's line in a spray-like pattern. This disease is usually noted at birth or within the second month of life and is frequently associated with abnormalities of the skeletal, ocular and nervous systems. In the hyperpigmentary phase, the disease is characterized by pigment incontinence within the dermis and little or no pigment within the basal layer.

In localized forms of scleroderma, histology reveals a thickened dermis containing bands of homogeneous, compact collagen and few elastic fibers. The epidermis is flat and atrophic with regression of interpapillary crests.

Ziprkowski Margolis syndrome is characterized by the presence of deaf-mutism and heterochromia of the iris.

Lichen planus shares a number of histological features with EDP, and numerous cases have been reported in which lichen is either associated with or precedes EDP (1, 8, 10).

For these reasons, many authors use the term "lichen planus pigmentosus" for EDP. Electron microscopy has, however, demonstrated differences in these two lesions. In EDP, the desmosomes and hemidesmosomes are normal or only slightly retracted, while in lichen planus, these structures are broken or missing. The perinuclear vacuoles seen in EDP are absent in lichen planus, which is associated, instead, with the presence of intranuclear bodies.

EDP is a dermatosis characterized by grayish plaques caused by an accumulation of melanin within the dermis; its etiology is currently unknown. Many authors believe that the disease is caused by long-term, accidental consumption of chemical substances present in the environment. Given the sporadic nature of the disease, others have hypothesized the role of an idiosyncratic predisposing factor (3, 7).

The histological findings change as the disease progresses (1). In the initial phase (illustrated by case 1), there is vacuolization of the basal keratinocytes, and dermal infiltrates of lymphocytes and histiocytes appear around the small vessels. Colloid bodies and mononucleate cell exocytosis can be seen within the epidermis. When the disease reaches an advanced stage, as it had in Case 2, clumps of melanophages and increased numbers of melanocytes appear within the dermis and are responsible for the increased pigmentation.

We felt that the cases described here deserved to be reported, not only because of the apparent rarity of this disease (which is, in our opinion, related more to the difficulties involved in its diagnosis), but also because they illustrate the histological evolution of the lesions over time. We would also like to emphasize the diagnostic importance of surface microscopy in patients with lesions of this type. This rapid, nontraumatic exam revealed oval-shaped, regular networks of brownish pigmentation which helped us to distinguish our patients lesions from those of fixed drug eruption, mycotic infections or lichen.

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