

ERYTHEMA DYSCROMICUM PERSTANS

A Report of Two Cases in Fair-skinned Patients

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Abstract. Erythema dyschromicum perstans is documented in two fair blond Caucasian females of polyglot northern European ancestry. Its emergence in these patients, along with three other cases in the literature, affirms that this diagnosis should not be limited to darkly pigmented people of Spanish or Indian descent.

Erythema dyschromicum perstans (EDP) is a disfiguring cutaneous disorder often termed ashy dermatosis. With the exception of three patients, this disease has been described entirely in darkly pigmented individuals (1, 2, 3). We are reporting two additional light skinned, blond individuals of northern European descent with EDP, suggesting that this unusual condition may be more widespread than is presently recognized. In fair-complexioned people, deliberate tanning may lessen the disfigurement.

CASE REPORTS

Case 1

A 12-year-old post-pubescent blond Caucasian female complained of grey spots over her abdomen, back, shoulders, and anterior thighs (Fig. 1). She described an episode of hives during the preceding month, but not in the same areas. She had been treated for tonsillitis with ampicillin on three occasions in the preceding year, but had not taken antimalarials, heavy metals, or other medications. Although the cutaneous lesions have not changed since the first episode, she has noticed that the grey areas are less apparent under a summer tan. Her parentage included a mother of Dutch, Swedish, Irish, and Welsh extract and a father from Irish and French stock.

Physical Examination: Multiple grey macules (Fig. 1) without erythema or scale were evident on the abdomen, low back, and anterior thighs. A nevus unius lateris extended from the left breast to the left forearm. Histologic examination of a typical macule showed findings consistent with EDP (Fig. 2). Laboratory tests including complete

blood count, erythrocyte sedimentation rate, urinalysis, glucose, calcium, phosphorus, blood urea nitrogen, uric acid, total protein, albumin, cholesterol, total bilirubin, alkaline phosphatase, lactic dehydrogenase, and serum glutamic oxalacetic transaminase were within normal limits. No additional lesions or change in the original macules have occurred in 2 years.

Case 2

This 12-year-old post-pubescent blond female was concerned about grey-brown macules which had developed on her trunk and were spreading to her arms, neck and thighs. She was in good health, but she had been receiving small doses of phenobarbital because of a epileptic seizure at age 4. Recent electroencephalograms have been normal. There was no history of antimalarial, heavy metals, or other drug ingestion. Her lineage consisted of a father who was Irish and Dutch and a mother whose parentage included French, Irish, German and a great-grandmother who was said to be an American Indian.

Numerous grey macules without erythema, induration or border elevation were patently obvious on physical examination. Histologic study of a typical macule showed vacuolization of the stratum malpighian, basal layer degeneration, and a perivascular dermal infiltrate of lymphocytes and pigment-laden macrophages. Laboratory examination included normal blood count, urinalysis, glucose, phosphorus, calcium, blood urea nitrogen, uric acid, cholesterol, total protein, albumin, total bilirubin, alkaline phosphatase, lactic dehydrogenase, and serum glutamic oxalacetic transaminase. No ova or parasites were found at stool examination. No clinical change has been observed in 18 months, although the grey areas are less apparent under a summer tan.

DISCUSSION

Erythema dyschromicum perstans reportedly has affected approximately one hundred and fifty people, both males and females, and in almost every decade of life. Nearly all have been dark-complexioned individuals of Spanish or Indian

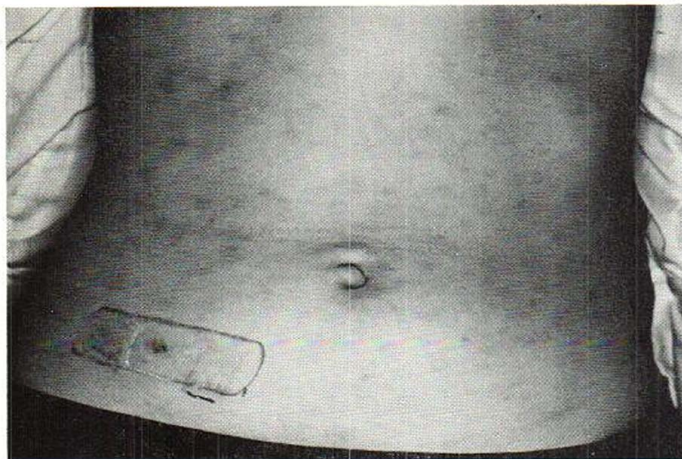


Fig. 1. Case One: The grey macules of EDP stand out in striking contrast with the lightly pigmented skin of the abdomen. Biopsy site on right lower quadrant.

descent from Central American countries (2, 3, 5). In some patients, elevated advancing erythematous borders precede the pigmented areas. The unsightly grey or grey-tan macules begin on the

trunk and spread to the exposed areas of the body, bringing the patient to the physician. Only three lightly pigmented females ages 5, 6 and 58 years have been reported in the literature (1, 6). This report of two additional fair blond girls emphasizes that the disorder occurs in light- as well as dark-skinned persons.

Microscopic findings in this disorder are only diagnostic when coupled with the clinical picture. Vacuolization of malpighian cells leading to spongiosis and occasional microvesicular formation, liquefaction degeneration of the basal layer cells, edema of the papillary dermis, incontinence of melanin pigment, and a mild lymphocytic perivascular infiltrate in the upper one-third of the dermis are the usual findings (Fig. 2). Several other disorders, including lupus erythematosus, Riehl's melanosis, dermatomyositis, drug eruptions and old lesions of lichen planus may resemble these histologic findings. However, of these diseases, only fixed drug eruptions might be confused clinically with EDP.

Ultrastructural studies have added little to our understanding of the pathogenesis of this disorder. The basal and lower malpighian cells show perinuclear translucent areas devoid of cytoplasmic organelles. In the lower epidermis, intracellular spaces are widened and there is desmosomal retraction. The papillary dermis contains melanin granules within mononuclear cells (4).

Treatment has been ineffective although tanning appears to lessen the cosmetic stigma. Personal communications with other dermatologists has led us to the opinion that this disorder may



Fig. 2. Histopathology of EDP includes vacuolization of malpighian cells, degeneration of basal layer cells, papillary dermis edema, and incontinence of melanin pigment (arrow).

be more common than is evident in the literature. However, it is apparently rare in fair-skinned persons. To our knowledge, this disorder has not been reported in lightly pigmented males which may be due to the diminished cosmetic concern in men.

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