Erythema Dyschromicum Perstans: Report of Five Cases and Review of the Literature

ERITEMA DISKROMIKUM PERSTANS: 5 OLGU SUNUMU VE LİTERATÜRÜN GÖZDEN GEÇİRİLMESİ

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Abstract

Erythema dyschromicum perstans (EDP) is a rare disorder characterized by asymptomatic, slowly progressive, ashy- gray macular hyperpigmentation of the skin. Most cases reported to date are of Latin American and Indian patients. Rare cases have been reported from Turkey. We present five cases that were diagnosed with EDP clinically and histopathologically.

Key Words: Pigmentation disorders, hyperpigmentation

Özet

Eritema diskromikum perstans (EDP) deride asemptomatik, yavaş ilerleyen gri-kahverengi renkte makuler hiperpigmentasyon ile karakterize nadir bir dermatozdur. Bugüne kadar sunulan olguların çoğu Latin Amerikan ve Hintli hastalar olup Türkiye'den az sayıda olgu bildirilmiştir. Bu makalede, klinik ve histopatolojik olarak EDP tanısı alan beş olgu sunulmaktadır.

Anahtar Kelimeler: Hiperpigmentasyon, pigmentasyon bozuklukları

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Ashy dermatosis or erythema dyschromicum perstans (EDP) is an unusual disorder of skin pigmentation that primarily occurs in young adults. The disorder is characterized by slate-gray oval macules and patches with erythematous borders that range in diameter from 0.5 to 3 cm.[1] The lesions appear in 2 stages: An initial inflammatory stage and a stage of pigmentary changes.[2,3] Lesions initially appear on the trunk and spread centrifugally to the extremities.[1] Both sexes are equally affected, and no genetic predisposition has been found.[4] This condition occurs anywhere from 5 years of age through adult life.[5] No treatment of choice is presently available. We report 5 patients with EDP and review the previously reported articles.

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Case Reports

Case 1

A 16-year-old girl presented with 2-month-long history of progressive cutaneous pigmentary changes. These began as truncal, asymptomatic blue-gray macules and patches, which slowly spread to her extremities and neck. Past medical history was not contributory; there was no history of drug intake. There was no family history of heritable skin diseases. Physical examination revealed diffuse, nonmucosal patches of brownish-gray macules located on her neck, trunk, abdomen and extremities (Figure 1). Routine laboratory findings were within normal limits.

Case 2

A 17-year-old girl presented with 6-month-long history of hyperpigmented macules on the trunk, abdomen and extremities. First, she noticed several gray patches on her abdomen. During 6 months, the patches gradually increased in number to involve most of the trunk and extremities. The

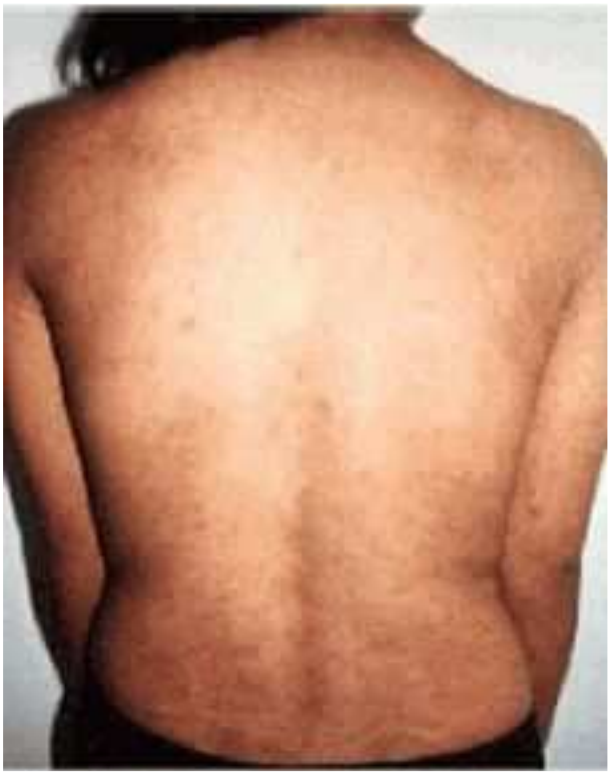

Figure 1. Clinical presentation of case 1 with blue- gray macules and patches.

patient's medical history and review of the systems was typical, and her family history was noncontributory. On physical examination, the patient had brownish-gray macules on the trunk, abdomen and extremities. Findings of routine laboratory investigations were within normal limits.

Case 3

An 11-year-old girl presented with 2-year-long history of progressive cutaneous pigmentary changes.These began as truncal, asymptomatic reddish macules. The macules changed to a brownish color, and the lesions spread to other parts of the trunk and extremities during the last year. Past medical history was not contributory, and there was no history of drug intake or other systemic illness. There was no family history of skin diseases. Physical examination revealed diffuse brownish macules on the neck, trunk, abdomen and extremities (Figure 2). Results of laboratory studies did not show any abnormalities.

Case 4

A 54-year-old woman presented with 2-month-long history of asymptomatic, brownish-gray macules which first started on her breasts, and spread to the trunk. Past medical history and family history was normal. On physical examination, the patient had brownish-gray macules located on these locations. Results of the Routine laboratory investigations were within normal limits.

Case 5

A 55-year-old man presented with 5-month-long history of cutaneous pigmentary changes. These began as asymptomatic gray macules on his upper extremities. The patient's medical history and review of systems was typical, and his family history was noncontributory. Physical examination revealed gray macules on his arms. Results of laboratory studies were within normal limits.

Punch biopsy of a macular lesion was performed from all of the patients. Microscopical examination revealed similar findings in all of the cases. Examination of each biopsy specimen demonstrated an atrophic, flattened epidermis with orthokeratosis, mild vacuolar alteration, occasional colloid bodies and focal exocytosis. An infiltrate of scattered lymphocytes and numerous melanophages in the papillary dermis were seen (Figure 3). These findings were consistent with EDP.

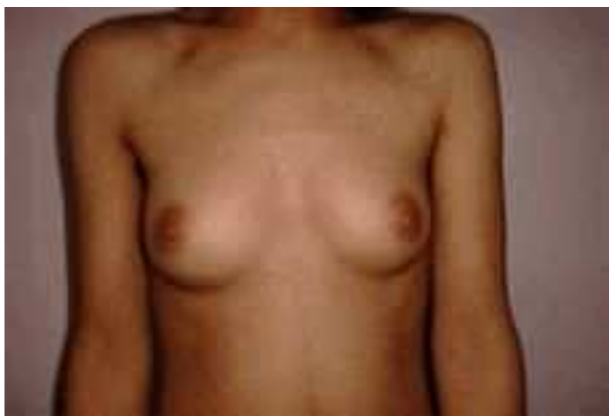

Figure 2. Diffuse brownish macules on the neck and trunk of case 3.

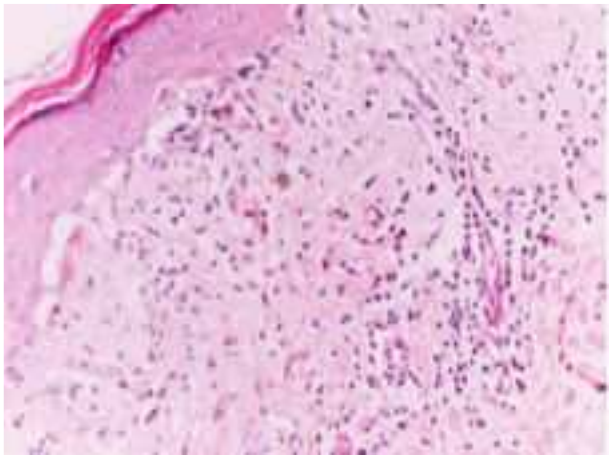

Figure 3. Atrophic, flattened epidermis with orthokeratosis, mild vacuolar alteration, occasional colloid bodies and focal exocytosis with an infiltrate of scattered lymphocytes and numerous melanophages in the papillary dermis (HE, X100).

Discussion

Erythema dyschromicum perstans (EDP) is an acquired pigmentary anomaly of unknown etiology first described by Ramirez in 1957.[6] Although EDP appears to be more prevalent in Latin Americans, it also may appear in whites and other races.[7-11] Rare cases have been reported from Turkey. Bahadır et al. described a 16-year-old Turkish boy whose eruption was notable both for its occurrance in a Turk and for its successful response to dapsone.[12]

The etiology of EDP remains unknown, although not conclusive associations with endocrinopathies,[13] ingestion of ammonium nitrate.[14] treatment with dithiazide iodide for whipworm infestation,[15] an X-ray contrast study,[10] exposure to the fungicide chlorothalonil,[16] vitiligo,[17] human immunodeficiency virus infection[18,19] and chronic hepatitis C[20] have been reported. EDP may be viewed as originating at an early stage in an inflammatory reaction to a chemical, an ingested agent, or contactant, followed by a persistant slate-colored hyperpigmentation. An abnormality in cell-mediated immunity might play a role, possibly as a result of the involvement of cell adhesion and activation molecules in its pathogenesis.[21]

There is no clear sexual predilection, but some authors report that EDP may be more common in females.[22] Four of our cases were female whereas only one case was male. EDP usually appears in adults, but some isolated cases and a small series of 8 cases[1] and a report of 14 cases[23] have been reported in prepubertal children. Our one case was prepubertal and four cases were adult.

The disease is characterized by the rapid onset of ash-colored macules that sometimes have erythematous and slightly elevated margins of about 1-2 mm in width. The border eventually disappears within several months, so it may be no longer being evident when the physicians examine the patient.[10] During our examination, in no place could an inflammatory border be appreciated in our cases.

The lesions are asymptomatic and occasionally pruritic.[23] Our all cases were asymptomatic. The shapes of the lesions can vary from oval or round to polycyclic, which is the more frequent, the size may vary from 3 mm to large, confluent patches, meaning several centimetres in diameter and the color from slate gray to lead-colored. The trunk and proximal extremities are more commonly involved following by the neck and the face. The lesions slowly extend peripherally, become confluent and can affect almost the entire body, without preference for exposed or unexposed areas. The palms, soles, scalp, nails and mucous membranes are usually spared.[22,23] Although the lesions were widespread on the trunk and abdomen in our four cases, the lesions were located only on the arms in one patient.

Histologic findings of EDP are not pathognomonic, but it is characteristic; active borders in early lesions show vacuolar degeneration of the basal cells, and pigmentary incontinence is observed with many melanophages in the upper dermis. Dermal blood vessels are sleeved with an infiltrate of lymphocytes and histiocytes.[2,13]

Distinguishing ashy dermatosis from lichen planus pigmentosus is not always easy.[24] EDP may at times resemble a fixed drug eruption, early pinta, and argyria, pigmentation from medication eruptions, such as that to carbamazepine, Addison's disease, melasma, macular amyloidosis, confluent and reticulate papillomatosis, and other cutaneous dyschromias.[25]

There are no effective therapies for EDP. The administration of topical agents, including steroids and hydroquinons, has been universally unsuccessful.[1] Recent reports, however, have suggested sulfone medications such as dapsone[3,12] and clofazimine[21,26] may prevent subclinical disease extension in adults. Therapies that have been reported to be of anectodal utility include oral corticosteroids, antibiotics, ultraviolet light therapy, isoniazid, griseofulvin and keratolytics.[1] We started clofazimine to case 2 and case 4. After 3-month period, the lesions fade in both cases. In case 1, we started dapsone two months ago, there has not been any change in the lesions yet. Case 3 and case 5 did not demand treatment.

As EDP infrequently appears in whites and other races, and rare cases have been reported from Turkey, we found it worthwhile to report 5 cases of EDP. It is notable that 5 patients from Ankara were diagnosed as EDP in such a short time, one-year period. It may suggest that environmental factors may play a role in EDP.

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