

Erythema Dyschromicum Perstans: A Case Report

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Abstract

Erythema dyschromicum perstans (EDP) is an acquired dermatosis characterized by ash gray (ashy dermatosis) or blue macules. It can appear at any age but is more common in young adults. Lesions may occur on the neck, chest arms and face, but most frequently on the back. It does not generally lead to subjective complaints, but may rarely cause itching. Diagnosis is made with clinical findings and can be corroborated by histopathological examination. This report describes the case of a 15-year-old female patient diagnosed with EDP and is intended to emphasize the need to include the condition at differential diagnosis.

Keywords: Ashy dermatosis, erythema dyschromicum perstans, hyperpigmentation

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Introduction

Erythema dyschromicum perstans (EDP) is a rare chronic and idiopathic pigmentation disease characterized by ash gray macules, also known as ashy dermatosis. It was first described by Ramirez in 1957 [1,2]. The etiology is uncertain [2,3]. Lesions can be seen in the form of indurated, erythematous gray-blue, oval or polycyclic macules. They are more common on the trunk, axillar region and upper extremities and symmetrical or asymmetrical [4]. The condition can appear at any age, but is more common in young adults. It does not generally lead to subjective complaints, but may rarely cause itching. Diagnosis is made with clinical findings and exclusion of diseases at differential diagnosis. Histopathology is not specific, but can support diagnosis [1].

We describe a rare case of EDP diagnosed in a 15-year-old female patient together with a review of the literature in order to emphasize the need to consider the condition among similar dermatoses at differential diagnosis.

Case Report

A 15-year-old female patient presented to our clinic with eruptions in the axillar region. She stated that these had appeared 1 year previously that she had been examined several times before and had been prescribed creams with various different diagnoses, although the condition had failed to resolve. There was no subjective complaint such as itching apart from the eruptions. There was no characteristic in her own or the family history, and systemic examination was normal. Dermatological examination revealed diffuse ash gray colored pigmented macular lesions of varying diameters in the right axillar region (Figure 1). Laboratory tests including complete blood count, erythrocyte sedimentation rate, liver and kidney function tests, blood electrolytes, thyroid function tests and complete urine tests were within normal limits. Anti-HBs, anti-HCV, anti-HIV, VDRL and ANA were all reported negative. A preliminary diagnosis of EDP was made on the basis of the clinical findings and an incisional biopsy was taken. Histopathological examination revealed mild vacuolar degeneration in the basal layer, a few colloid bodies and lymphocyte and melanophage infiltration in the papillary dermis (Figure 2). EDP was diagnosed with exclusion of other diseases at differential diagnosis. The patient was started on 100 mg/day dapsone therapy, and the condition was brought under control.



Figure 1. Diffuse ash gray colored pigmented macular lesions of varying diameters in the right axillar region

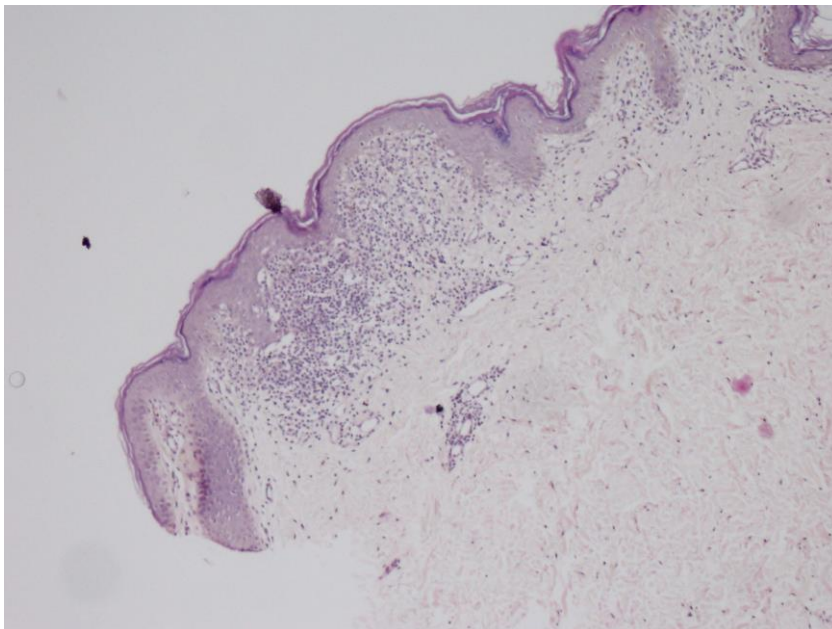


Figure 2. Histopathological examination revealed mild vacuolar degeneration in the basal layer, a few colloid bodies and lymphocyte and melanophage infiltration in the papillary dermis

Discussion

Ashy dermatosis or EDP is a comparatively rare skin disease from the idiopathic acquired hypermelanosis group [5]. The etiopathology is uncertain, although several factors have been

implicated. These include infections (parasites and HIV), toxins (ammonium nitrate, cobalt, radiocontrast materials, pesticides and fungicides), drugs (ethambutol, penicillins and benzodiazepines), endocrinopathies (thyroid diseases and diabetes mellitus), atopy and dyslipidemia. Immunopathological investigation of active lesions has shown that immune-mediated mechanisms are probably involved in the pathogenesis of EDP [2,5].

The disease is most common in the 1st-3rd decades, but can be seen at any age, albeit rarely. It is also more common in women and subjects with dark skin. Lesions may be of sudden onset or progress gradually in the form of ashy gray-blue asymptomatic macular lesions 0.5-2 cm in size, oval, polycyclic or irregular in shape. Early lesions may take the form of protruding erythematous papules with margin activation. They generally appear on the trunk, the proximal aspects of the extremities, the neck and face. The disease is usually asymptomatic, although mild itching may sometimes be seen [2,4]. Gray-colored asymptomatic macules 0.5-1 cm in size was also observed in the axillar region in our case. As a 15-year-old female, our case was compatible with the literature.

Diagnosis is made with clinical findings and histopathological tests. Histopathology is important in order to rule out other diseases at differential diagnosis, but is not pathognomonic. Histopathological examination of biopsy from active lesions reveals a picture compatible with lichenoid dermatitis, such as vacuolar degeneration in the basal layer, colloid bodies and increased epidermal melanin. Edema in the papillary dermis, mild or moderate lymphocytic infiltration and dermal melanophages can also be seen. Loss of vacuolization in the basal layer and decreased dermal infiltrate is determined in the inactive phase [1,2,5]. Presence of fibrinogen and IgM and C4 colloid deposition may be observed at direct immunofluorescent microscopy [3]. Lichen planus pigmentosus, post-inflammatory pigmentation, figurate erythema, pityriasis rosea, multiple fix drug eruption, hemochromatosis, Addison disease, melasma, leprosy, idiopathic eruptive macular pigmentation, macular amyloidosis, confluent and reticular papillomatosis and secondary syphilis may be considered at differential diagnosis [2,5]. At histopathological examination in our case we observed mild vacuolar degeneration in the basal layer, colloid bodies and lymphocyte and melanophage infiltration in the papillary dermis. EDP was diagnosed with histopathological exclusion of other diagnoses and the existing clinical findings.

There is still no effective therapeutic option, although various treatments have been tried. Solar protectors, chemical peeling, antibiotics, local and systemic corticosteroids and vitamins have been reported to be ineffective, while partial effects have been observed with isoniazid, griseofulvin, clofazimine and dapsone [6]. Bahadır et al. reported that lesions on the arms, face and neck of a 19-year-old male patient resolved completely in 3 months with dapsone therapy and that no new lesions occurred [7]. Akarsu et al., however, administered dapsone for 4 months to a 36-year-old patient diagnosed with EDP and reported no improvement in lesions [8]. We started our patient diagnosed with EDP on dapsone therapy at 100 mg/day and brought the lesions under control. However, the patient failed to attend for subsequent check-up. We were therefore unable to assess the outcome of the treatment. Further clinical studies with sufficient case numbers are needed to determine the effectiveness of these treatments.

In conclusion, EDP is a rare dermatosis. Therefore, although clinical studies with sufficient patient numbers are problematic, case reports need to be issued at the very least. This will increase our knowledge of EDP and enable it to be considered among other diseases at differential diagnosis.

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