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Case Report
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Erythema Dyschromicum Perstans in a 10-Year-Old Girl

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Abstract

Observation: Erythema dyschromicum perstans (EDP) is a rare skin disorder characterized by hyperpigmented macules of various size on the trunk, face and extremities. It is an acquired dermatosis that occurs most frequently in Central and South America. Only a few cases have been reported from Turkey. EDP usually appears in adults, but some isolated cases and small series have been reported in prepubertal children. We present a case of EDP in a 10-year-old Turkish girl.

Introduction

Erythema dyschromicum perstans (EDP) or ashy dermatosis is a rare disorder characterized by asymptomatic, slowly progressive, ash-gray macular hyperpigmentation that is most common in Hispanic patients [1]. Only a few cases reported from Turkey [2,3]. The etiology of EDP is still unknown. The therapeutic options are many, but few have been effective. EDP is unlikely to resolve in adults; however, most prepubertal children have a course of spontaneous slow resolution.

Case Reports

A 10-year-old girl presented with a 3-year history of an ashy gray colored macular lesion which first developed on the left side of her neck and thereafter spread to her chest, back and left arm. The parents and her siblings do not have similar skin lesions. There was no history of previous skin eruption or topical and systemic drug consumption. She was otherwise healthy. Dermatological examination revealed numerous blue-grayish macules of varying sizes and shapes, confluent and located in the neck, chest, back, and left arm (**Figure 1**). Mucous membranes, palms, soles and the remaining integument were normal. Histologic examination of a skin biopsy demonstrated epidermis with increased pigmentation at the basal layer, focal degeneration of the basal cells, pigment incontinence, and a perivascular lymphohistiocytic infiltrate in upper dermis (**Figure 2**). A diagnosis of EDP was made on the basis of the clinical and histological findings. The patient was advised to avoid sun exposure, and to use sunscreen.

Discussion

Ramirez first described EDP in 1957. This description of 58 patients included those as young as 7 years of age. They were South Americans, whose families referred to them colloquially as "los cenescientos," or the ashy people, leading to the commonly used name "ashy dermatosis" [1]. The etiology of EDP is unknown. However, a number of etiological factors like ingestion of ammonium nitrite, nematodes infestation, radiographic contrast media, cobalt allergy, and chlorothalonil exposure have been implicated [4]. EDP usually appears in adults,

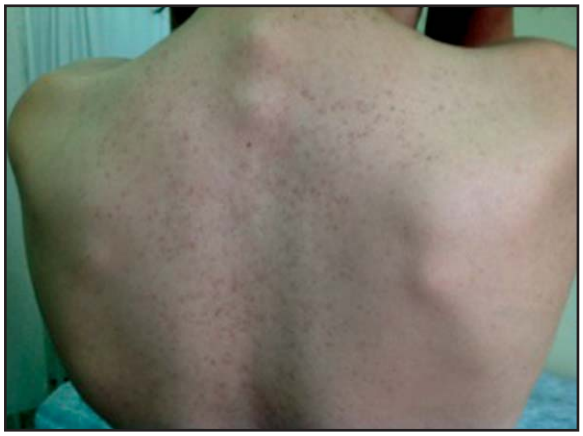

Figure 1. Clinical appearance of the upper back

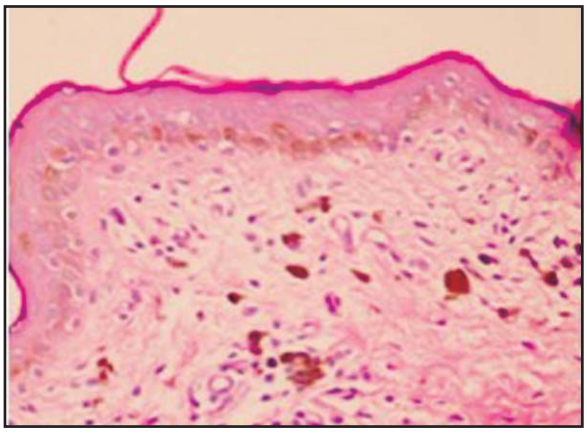

Figure 2. Histologic examination demonstrating focal degeneration of the basal cells, pigment incontinence. HEx400

but some isolated cases and small series have been reported in prepubertal children [**1,5**]. The diagnosis of EDP is based on the appearance of the symmetric, 0.5–2 cm, ashy gray to blue, hyperpigmented oval plaques and patches 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 be evident when the physicians examine the patient. The lesions slowly extend peripherally, become confluent and can affect almost the entire body. The palms, soles, scalp, nails and mucous membranes are usually uninvolved. The lesions are usually asymptomatic [**5**].

The histopathology of EDP is not pathognomonic. The most common histopathological findings are a dermal perivascular lymphocytic infiltrate with many melanophages, vacuolization of the basement membrane zone, necrotic keratinocytes in the basal layer, colloid bodies, exocytosis of lymphocytes, and incontinence of the pigment [**6**].

The differential diagnosis of EDP includes lichen planus pigmentosus, fixed drug eruption, early pinta, argyria, pigmentation from medication eruptions, such as that to carbamazepine, Addison's disease, melasma, macular amyloidosus, confluent and reticulate papillomatosis, and other cutaneous dyschromias [**3, 4**]. There are no effective therapies for EDP.

The administration of topical agents, including steroids and hydroquinones, has been universally unsuccessful. Other therapeutic options for EDP are sulfone medications such as dapsone and clofazimine, may prevent subclinical disease extension in adults. Therapies that have been reported to be of anecdotal utility include oral corticosteroids, antibiotics, ultraviolet light therapy, isoniazid, griseofulvin, and keratolytics [**7**]. We did not recommend any treatment for our patients, because none is consistently effective and none of them is devoid of adverse effects. Also, unlike adults, EDP in children can have an eventual improvement or spontaneous resolution as was seen in a study in prepubertal children [**7**].

We recommend educating the family of pediatric patients about the good prognosis of prepubertal EDP while awaiting spontaneous resolution.

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