Erythema dyschromicum perstans showing resolution in an adult

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INTRODUCTION

Erythema dyschromicum perstans (EDP) is a rare cutaneous eruption of unknown etiology that often resolves in prepubertal children but is generally thought to persist in adults. We present a case of EDP in an adult that cleared significantly after 6 years.

CASE REPORT

A 25-year-old Hispanic woman with type IV phototype complexion presented in December 2006 with a slightly pruritic eruption of 1 month's duration consistent with pityriasis rosea (PR). It began with a typical herald patch on the left flank followed by scaly pink papules on the left forearm spreading to the trunk and proximal upper extremities.

The patient's medical history was significant for alopecia areata at age 20, which had been treated successfully with intralesional triamcinolone. She had no known family history of skin or autoimmune disease. Medications included an oral contraceptive pill, which she had been using for several years. Initial workup included a rapid plasma reagin, the results of which were negative. She was treated for presumptive PR with twice-weekly narrowband ultraviolet B therapy for 8 weeks (maximum dose, 1680 mJ/cm^2), along with daily topical mometasone.

One month after completing this regimen, the eruption continued to evolve, although she had not taken any new medications (prescribed or over the counter) nor had she changed her lifestyle. Skin examination found multiple 0.3- to 1.5-cm slate-gray to brown macules and patches, many with a pink rim, distributed over the trunk, buttocks, proximal extremities, and neck. The lesions on the back and Abbreviations used:

- EDP: erythema dyschromicum perstans PR:
- pityriasis rosea

abdomen were along lines of cleavage (Fig 1, A). Lesions were not present on the palms, soles, or oral mucosa.

Biopsy of several lesions on the abdomen and arm found a mild interface dermatitis with pigment incontinence (Fig 2, A and B) supporting a diagnosis of EDP. Absence of spongiosis ruled out a diagnosis of PR. A more extensive workup was done: a complete blood count and comprehensive metabolic panel were within normal limits. Autoimmune studies included a normal thyroid stimulating hormone and negative rheumatoid arthritis screen, antinuclear antibody, SS-A, SS-B, and gliadin IgG/IgA antibody. The patient's total 25-OH vitamin D level was found to be low at 12 ng/mL (reference range, 20-100). Over a 2-year period, additional treatment with topical clobetasol, pimecrolimus, and tacrolimus (each for over 3 months) and oral dapsone for more than 2 months (25 mg/d initially, then 50 mg/d) were not helpful in preventing progression of her pigmented eruption onto her distal extremities, and she was lost to follow-up.

Five years later, the patient returned because of a new 2-cm patch of alopecia on her scalp. This finding prompted a full examination, which found significant clinical resolution of her pigmented lesions (Fig 1, B). The patient felt that exposure to summer sunlight over the preceding 2 to 3 years had helped.

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Fig 1. EDP over time. **A**, Patient at 1-year follow-up, with symmetric widespread coalescing slate-gray to brown macules and patches on the abdomen, back, and proximal extremities. Lesions on the abdomen and back are distributed along lines of cleavage. **B**, Patient at 6-year follow-up, with significant clearing of lesions. Note: Identifying tattoo on patient's right upper back has been obscured.

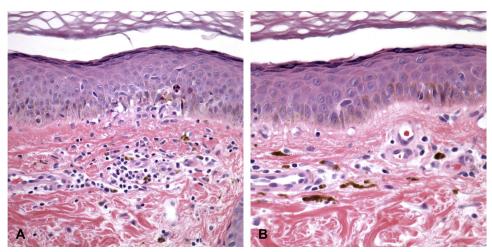


Fig 2. Representative histopathology of EDP. **A**, Interface dermatitis with basal vacuolar change, a few apoptotic keratinocytes, and mild superficial inflammatory infiltrate composed of small lymphocytes. The stratum corneum shows a preserved basket-woven pattern. **B**, Variable basilar pigmentation, pigment incontinence, mild vascular ectasia, and slight flattening of the rete pattern. Spongiotic changes were not identified. (**A** and **B**, Hematoxylineosin stain; original magnification: ×400.)

DISCUSSION

EDP, or ashy dermatosis, is a rare asymptomatic eruption of unknown etiology. It was first described in El Salvador by Ramirez in 1957 who termed affected patients *los cenicientos* because of the characteristic ashy color of the skin lesions.¹ Clinically, it is characterized by slowly progressive gray-brown to blue-gray macules and patches symmetrically distributed over the neck, trunk, and proximal arms. A thin erythematous peripheral border may be present in acute lesions (as in our case). It occurs more commonly in persons of Latin American descent with skin types III and IV.²

Our patient presented with an eruption suggestive of PR, including a herald patch on the right flank and a distribution of lesions along cleavage lines, before developing the slate-gray to brown macules of EDP. However, spongiotic changes were not identified in several biopsies to support PR. It is possible that PR triggered the onset of EDP; however, an association between these conditions has never been reported, to our knowledge. Interestingly, the lesions of EDP may follow skin cleavage lines, with a pattern that resembles PR^{2,3}; however, the postinflammatory hyperpigmentation of PR is transient and fades in a matter of weeks compared with the chronic nature of EDP.³ The distribution of lesions along lines of cleavage exemplified in this case shows that EDP may present similarly to PR and may result in early misdiagnosis of this disease entity.

Another unusual aspect of our case is near complete resolution of lesions with long-term follow-up. Although the rash of EDP tends to resolve in 2 to 3 years in prepubertal children,^{4,5} it is generally believed that lesions persist in adults.

Our case, showing striking resolution 6 years after disease onset, shows that lesions of EDP may clear in adults as well.

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