
Erythema Dyschromicum Perstans: A New Manifestation Of Sjogren's Syndrome

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Citation

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Abstract

Erythema dyschromicum perstans (EDP), otherwise known as ashy dermatosis, is a benign and chronic skin disorder characterized by hyperpigmented lesions of various sizes on the trunk, face and extremities. Its etiology is obscure and current treatment modalities remain ineffective. We report a patient with this unique rash that subsequently developed primary Sjogren's syndrome. Various dermatological manifestations of Sjogren's syndrome that are well known include xerosis, pruritus, angular cheilitis, eyelid dermatitis, annular erythema, and vasculitis. EDP however, is not typical of any of the cutaneous features associated with Sjogren's Syndrome nor has it been reported in such patients.

CASE REPORT

A 38-year-old woman first presented with a sudden eruption of scattered non-pruritic skin lesions on the face and arms in 2008. The lesions continued to evolve into extensive patchy hyperpigmented areas on the face, neck and chest and showed no signs of resolution. There is no history of photosensitivity or alopecia. She was seen by multiple dermatologists and tried multiple ointments and creams with no improvement. Her past medical history includes migraines and anemia and is on iron supplementation. She subsequently developed keratoconjunctivitis sicca and was referred for a rheumatology evaluation.

On examination, she had extensive hyperpigmented lesions with a bluish discoloration on her face and forehead without papules, vesicles, or discoid lesions. General examination was unremarkable. Laboratory tests including complete blood count, erythrocyte sedimentation rate, comprehensive metabolic panel, ferritin, iron, thyroid stimulating hormone, serum protein electrophoresis and complement levels were normal. Further investigations revealed a positive SS-A, however the ANA, SS-B and lupus anti-coagulant were all negative.

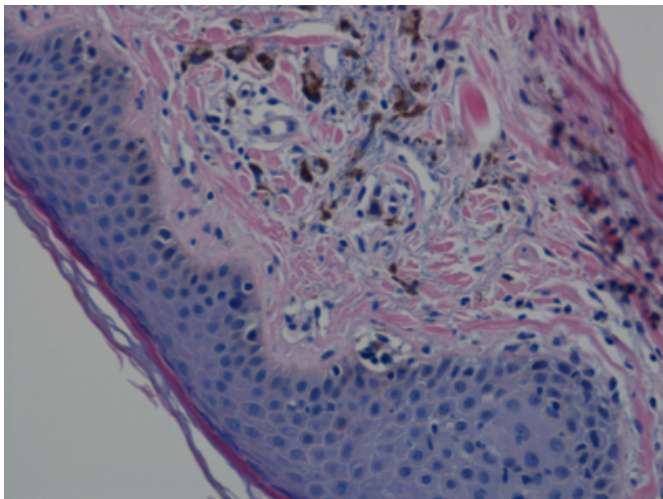
She underwent a shave biopsy of the left forehead to determine the etiology of the vague rash. It revealed sections of skin showing a lichenoid infiltrate of lymphocytes with vacuolar changes and dyskeratotic keratinocytes with many scattered melanophages in the papillary and reticular dermis consistent with erythema dyschromicum perstans. The

changes noted are compatible with a cutaneous manifestation of collagen vascular disease. She was subsequently diagnosed with primary Sjogren's syndrome (pSS) based on her symptoms and serology.

Figure 1



Figure 2



DISCUSSION

It is reported that the skin is affected in nearly half of Sjogren's syndrome patients. Cutaneous features include xerosis, pruritus, angular cheilitis, eyelid dermatitis, annular erythema, and vasculitis mainly presenting as palpable purpura. Most of them are nonspecific and less severe than the oral, ocular or musculoskeletal symptoms. Among the most frequent skin involvement found were xerosis and angular cheilitis in a review of 93 SS patients by Bernacchi et al (1). The peculiar cutaneous finding in this patient has not been reported in pSS. Erythema Dyschromicum Perstans is a relatively rare disease first described in Latin America in 1957 (3). It is a benign but chronic condition. The etiology is unknown although an immunologic basis has been suggested (2). A genetic susceptibility with HLA-DR association was studied in Mexican Mestizo patients by Correa et al and showed the most frequent allele was HLA-DR4 (65%) compared to 23% in ethnically matched controls (2). EDP is clinically manifested as slowly spreading well-defined blue-grey macules that may affect extensive areas but usually the palms soles, scalp, nails and mucous membranes are spared (4). No systemic symptoms or associations exist to date. Therapy remains ineffective in most patients because of persistent residual pigmentation, however success has been reported in some cases with the use of clofazimine (5), and fortunately with our patient as well.

References

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