Recent Advances in Pathology & Laboratory Medicine
Volume 9, Issue 3&4 - 2023, Pg. No. 11-13
Peer Reviewed & Open Access Journal

Case Study

A Case of Erythema Dyschromicum Perstans

Khushbu Katariya[1], Parul Singhal[2], Lamia[3]

[1]Assistant Professor, [2]Professor, [3]Post Graduate Student, Department of Pathology, Saraswathi Institute of Medical Sciences, Hapur, India.

DOI: https://doi.org/10.24321/2454.8642.202306

I N F O

Corresponding Author:
Parul Singhal, Department of Pathology, Saraswathi Institute of Medical Sciences, Hapur, India.
E-mail Id:
Dr_singhal_parul@yahoo.co.in
Orcid Id:
https://orcid.org/0000-0002-7614-3011

How to cite this article:
Katariya K, Singhal P, Lamia. A Case of Erythema Dyschromicum Perstans. Rec Adv Path Lab Med. 2023;9(3&4): 11-13.

Date of Submission: 2023-10-21
Date of Acceptance: 2023-11-20

A B S T R A C T

Erythema dyschromicum perstans (EDP) is an acquired disease presenting as ashy dermatosis comprising blue–grey pigmented patches over the neck, trunk, and extremities. The pigmentation may become disfiguring when present for a long time. The aetiology of EDP is unclear, and there is no defined treatment or established therapy. EDP has a presentation mimicking lichen planus pigmentosus, idiopathic eruptive macular pigmentation, post-inflammatory hyperpigmentation, fixed pigmented erythema, Addison's disease, and hemochromatosis. All these pigmentation disorders make the diagnosis and treatment of EDP quite challenging. No treatment for EDP has been universal, and combination treatments have worked in different cases to cure this disease.

Keywords: Erythema Dyschromicum Perstans, Ashy Dermatosis

Introduction

Erythema dyschromicum perstans (EDP) is an acquired disease presenting as ashy dermatosis comprising blue–grey pigmented patches over the neck, trunk, and extremities. The pigmentation may become disfiguring when present for a long time. The etiology of EDP is unclear and there is no defined treatment or established therapy. In 1957, Ramirez, for the first time, reported 139 patients in El Salvador having progressive macules with a purplish hue forming patches on the arms, legs, and trunk, labelled as "los cencientos", meaning ashen ones. Herein, we report a case of EDP with extensive lesions, cured by oral steroid therapy.

Case Report

An 11-year-old male patient presented to the Dermatology Outpatient Department of Saraswathi Institute of Medical Sciences with multiple, asymptomatic, violaceous papules present all over the body for 9 months. The lesions first appeared on the trunk and then slowly spread to the whole body, including the back and the extremities. The overall clinical and general body examination was normal. There was no history of previous skin lesions, family history, autoimmune disease, or thyroid involvement.

The dermatological evaluation showed bluish-grey, symmetrical, confluent macules with polycyclic margins at all places. Isolated lesions were seen at some areas which varied in size from 0.5 to 2.0 cm. No erythema was seen at the margins. The palms, soles, nails, and mucous membranes showed no abnormal characteristics. No history of drug intake, itching, fatigue, or systemic symptoms was noted. The laboratory investigations reported normal blood counts, stool examination, and liver function tests.

The differential diagnoses in the clinical context were shortlisted to erythema dyschromicum perstans, lichen planus, idiopathic eruptive macular pigmentation, post-inflammatory hyperpigmentation, fixed pigmented erythema, Addison's disease, and haemochromatosis.

A 4 mm punch biopsy of a lesion on the trunk was taken. The skin biopsy revealed focal vacuolar disintegration of the basal epidermal layer. Infiltration of lymphocytes,

histiocytes, and melanophages along with pigment incontinence was seen in the papillary dermis. The clinical and histological findings zeroed down to EDP as the final diagnosis. The patient was prescribed Clofazimine, an antileprosy drug along with corticosteroids. After three months, he demonstrated marked improvement with treatment. Fig 1 and Fig 2

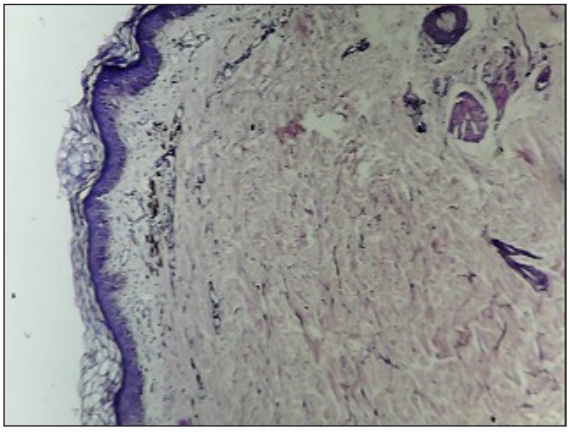

Figure 1.Degeneration of Basal Layer of Epidermis (H&E Stain, 10X)

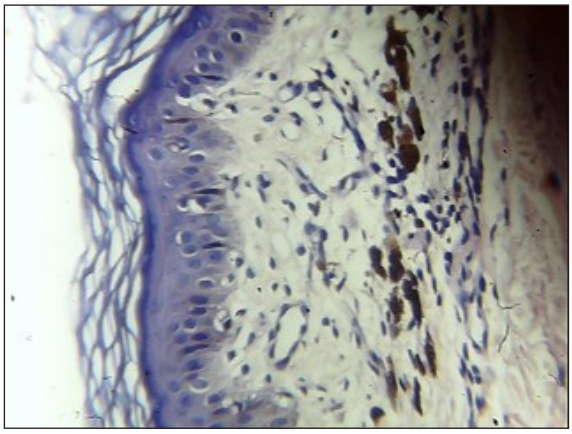

Figure 2.Melanosis in the Upper Dermis and Perivascular Infiltrate at Places (H&E Stain, 40X)

Discussion

Erythema dyschromicum perstans (EDP) is a benign, chronic, progressive skin disorder presenting in both males and females worldwide. Fitzpatrick skin types III-V are more commonly inflicted with EDP.[1] The most common age of presentation is before 40 years, though children and the elderly are not totally spared. EDP is a pigmentation disorder characterised by the formation of greyish-blue (ashy) patches/ macules throughout the body. The lesions have polycyclic margins. The aetiology of EDP is not definite, though genetic susceptibility and allergy to various hair dyes and cosmetics have been proposed. Toxic chemicals like barium sulphate and ammonium nitrate, viral infections, whipworm infestation, and adverse drug reactions are also seen to be causative factors of EDP in a few cases.

Skin lesions clinically present as symmetrical round to oval or irregular-shaped grey or bluish-brown macules and patches distributed all over the body. The lesions are 0.5-3.5 cm in size. Lesions usually start on the trunk and spread to the neck, arms, and occasionally on the face. Early lesions may have a slightly elevated, non-scaly, erythematous border, while older lesions may have an ill-defined border. Mucous membranes are unaffected. Skin biopsy is the only resort to rule out other diseases and confirm EDP.

EDP may persist for years, although spontaneous remission may occur in children. There are a few treatment options for EDP, but none has been shown to be consistently effective. Clofazimine and dapsone have been shown to be effective in some patients. Hydroquinone and topical steroids may yield varying results. Other treatment modalities include ultraviolet light therapy, chemical peels, isoniazid, griseofulvin, and antihistamines.[2]

EDP has a presentation mimicking lichen planus pigmentosus, idiopathic eruptive macular pigmentation, post-inflammatory hyperpigmentation, fixed pigmented erythema, Addison's disease, and hemochromatosis.[3] All these pigmentation disorders make the diagnosis and treatment of EDP quite challenging. Convit et al., in 1961, reported 5 patients with a clinical presentation similar to EDP in Venezuela.[3] However, the lesions were devoid of erythematous margins and self-healed in a few months. The disease was called EDP by these authors owing to the erythematous borders of the active patches. Since then, many dermatologists have tried to determine whether EDP can be placed in a unique category or should be included in a wide dyschromia spectrum, just like LPP.[4,5] Now, it has been concluded that EDP and ashy dermatosis refer to stages of the same disease.[6]

Degenerative changes in the basal keratinocytes and melanocytes occur because of hypersensitive immune reactions to unknown antigens. CD8+ T lymphocyte predominance in the dermis and epidermal accumulation of HLA-DR+ and ICAM 1+ keratinocytes are noted.[7,8] A genetic susceptibility related to MHC (mostly HLA-DR4) is also commented upon.[9] Patch testing to locate trigger antigens causing EDP, LPP, or contact dermatitis with pigmentation has also been documented, which showed that 40% of patients having EDP and 36% of patients suffering from LPP reported positive patch testing.[10,11] No treatment for EDP has been universal, and combination treatments have worked in different cases to cure this disease.

Conclusion - No treatment for EDP has been universal, and combination treatments have worked in different cases to cure this disease.

ISSN: 2454-8642
DOI: https://doi.org/10.24321/2454.8642.202306

Conflict of Interest: None

References

1. Chang SE, Kim HW, Sin JM, Lee JH, Na JI, Roh MR. Clinical and histological aspect of erythema dyschromicum perstans in Korea: a review of 68 cases. J Dermatol. 2015 Nov;42(11):1053-7. [PubMed] [Google Scholar]
2. Ramirez CO. Los cenicientos: problema clinico. Proceedings of the First Central American Congress of Dermatology; 1957 Dec 5–8. San Salvador; 1957. [Google Scholar]
3. Convit J, Kerdel-Vegas F, Rodriguez G. Erythema dyschromicum perstans: a hitherto undescribed skin disease. J Invest Dermatol. 1961;36:457-62. [Google Scholar]
4. Bhutani LK. Ashy dermatosis or lichen planus pigmentosus: what is in a name? Arch Dermatol. 1986 Feb;122:133. [PubMed] [Google Scholar]
5. Ghosh A, Coondoo A. Lichen planus pigmentosus: the controversial consensus. Indian J Dermatol. 2016;62(5):482-6. [PubMed] [Google Scholar]
6. Zaynoun S, Rubeiz N, Kibbi AG. Ashy dermatosis–a critical review of literature and a proposed simplified clinical classification. Int J Dermatol. 2008 Jun;47(6):542-4. [PubMed] [Google Scholar]
7. Baranda L, Torres-Alvarez B, Cortes-Franco R, Moncada B, Portales-Perez DP, Gonzalez-Amaro R. Involvement of cell adhesion and activation molecules in the pathogenesis of erythema dyschromicum perstans (ashy dermatitis). The effect of clofazimine therapy. Arch Dermatol. 1997 Mar;133:325-9. [PubMed] [Google Scholar]
8. Pinkus H. Lichenoid tissue reactions. A speculative review of the clinical spectrum of epidermal basal cell damage with special reference to erythema dyschromicum perstans. Arch Dermatol. 1973 Jun;107:840-6. [PubMed] [Google Scholar]
9. Correa MC, Memije EV, Vargas-Alarcon G, Guzman RA, Rosetti F, Acuna-Alonzo V. HLA-DR associated with the genetic susceptibility to develop ashy dermatosis in Mexican Mestizo patients. J Am Acad Dermatol. 2007 Apr;56(4):617-20. [PubMed] [Google Scholar]
10. Tienthavorn T, Tresukosol P, Sudtikoonaseth P. Patch testing and histopathology in Thai patients with hyperpigmentation due to erythema dyschromicum perstans, lichen planus pigmentosus, and pigmented contact dermatitis. Asian Pac J Allergy Immunol. 2014 Jun;32(2):185-92. [PubMed] [Google Scholar]
11. Combemale P, Faisant M, Guennoc B, Dupin M, Heyraud JD. Erythema dyschromicum perstans: report of a new case and critical review of the literature. J Dermatol. 1998 Nov;25(11):747-53. [PubMed] [Google Scholar]

ISSN: 2454-8642
DOI: https://doi.org/10.24321/2454.8642.202306