

Case study

ERYTHEMA DYSCHROMICUM PERSTANS, the spotty possibility – A rare case report

ABSTRACT:

Erythema dyschromicum perstans / ashy dermatosis is a less common dermatological disorder with an unknown etiology. It is characterized by asymptomatic, grey, symmetric and confluent macules all over the body. Herein, we report a case of a 50 years old man who presented clinically, with diffuse, patchy pigmentation having ill-defined, erythematous borders involving bilateral arms and forearms. Histopathological examination of the lesion biopsy demonstrates vacuolar degeneration of the basal cell layer, dermal perivascular mononuclear cell infiltrate and increased epidermal melanin pigment and presence of dermal melanophages. For the present case, treatment with clofazimine has proven to be effective.

Keywords:

Erythema dyschromicum perstans, ashy dermatosis, dermal melanophages, clofazimine

INTRODUCTION:

Erythema dyschromicum perstans (EDP) is a pigmentary disorder on the spectrum of acquired macular pigmentation. It is also known as ashy dermatosis due to its clinical presentation. Predominantly, it is found in patients with skin phototypes III-IV.⁽¹⁾ The most commonly affected sites include trunk, neck, face and upper limbs but any region of the body can be affected. Onset can occur at any age but the mean age of occurrence is usually in the second and third decade of life. Demographically, this dermatological disease occurs quiet commonly in Asian and central/south American continents. It can affect both the sexes but slightly more common in women. The exact etiopathogenesis involving ashy dermatosis is still unknown but its association with contrast agents, infections and parasitic infestations has been widely reported.

CASE HISTORY

A 50 years old male patient with type III complexion, approached the dermatology department, in our college with a history of recent dye application over scalp hair and subsequently presenting with numerous, asymptomatic, brown-grey macules with mild erythematous borders, **(Figure 1)** involving the face initially and later progressing to bilateral arms and lateral aspects of chest. He had no known family history of skin or autoimmune diseases. A four millimeter skin punch biopsy was taken and sent to the pathology department for histopathological examination.

Histopathology examination revealed mild hyperkeratotic and atrophic epidermis with basal cell vacuolar degeneration and pigmented incontinence overlying the fibro-collagenous dermis. **(Figure 2)** Also, the superficial dermis showed moderated lymphocytic infiltration admixed with pigment laden macrophages surrounding capillaries. **(Figure 3)**

A diagnosis of Erythema dischromicum perstans (EDP) was made.

DISCUSSION

Pathologically, EDP may involve immune mediation. It has also been postulated that direct damage to melanocytes and basal layer keratinocytes results from abnormal immune response of the body to particular foreign antigens.

The hair dye applied by our patient could have been responsible for such postulated immune reaction. Typically, the active lesions in this condition show basal cell vacuolar degeneration with pigmentary incontinence by presence of many melanophages in the upper dermis. There is also lymphohistiocytic infiltration of dermal capillaries documented in many such cases.

In case of residual macules, there is predomination of the pigment incontinence, while the cellular infiltration and basal cell vacuolar degeneration may range anywhere from minimal to intense. Thus, the patient was diagnosed with EDP from both clinical and histopathological findings.

Differential diagnosis of this condition includes Addison's disease, pigmented lichen planus, idiopathic eruptive macular pigmentation and post-inflammatory hyperpigmentation. Pigmented lichen planus is the most commonly considered differential diagnosis of EDP. In pigmented lichen planus, the lesions are usually characterized by bright violaceous/purple, flat and solid papules that are often crossed by whitish lines (Wickham Striae). Lichen planus commonly involves mucous membrane and is associated with mild pruritis,⁽²⁾ neither of which were present in our patient. We excluded features like Ultra-violet (UV) light induced aggravation of lesions⁽³⁾ and systemic autoimmune/ infectious pathogenesis⁽⁴⁾ of post-inflammatory hyperpigmentation

and Addison disease respectively, as a result of thorough history elucidation and clinical examination.

Use of various therapies including topical and systemic corticosteroids, UV light, antimalarial agent and antibiotics has been reported without clear evidence of benefit.^(5,6)

Some limited data from small studies suggest significant benefit from treatment with clofazimine.^(7,8) Thus, it was tried and utilized in our current scenario.

Conclusion:

In our case, clofazimine along with topical mometasone has been used with some success, apparently because of their anti-inflammatory and immune-modulating effects. Patient was treated with both for three months. After the treatment period, a betterment of the erythematous component of the pigmented lesions was noticed. But for many patients however, longstanding EDP can be chronically disfiguring and a disconcerting problem that is resistant to treatment modalities.⁽⁶⁾ Clinicians should suspect this entity whenever a large pigmented process is presented so that therapy can be initiated much earlier and lessen the residual impact of the lesions.

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UNDER PEER REVIEW

Figure 1: Clinical picture of the grey colored macules which show mild hyperemia at the edges



Figure 2: Section showing thinned out epidermis with basal cell vacuolar degeneration overlying a fibrocollagenous dermis. Lymphocytes and pigment laden macrophages noted in the dermoepidermal junction

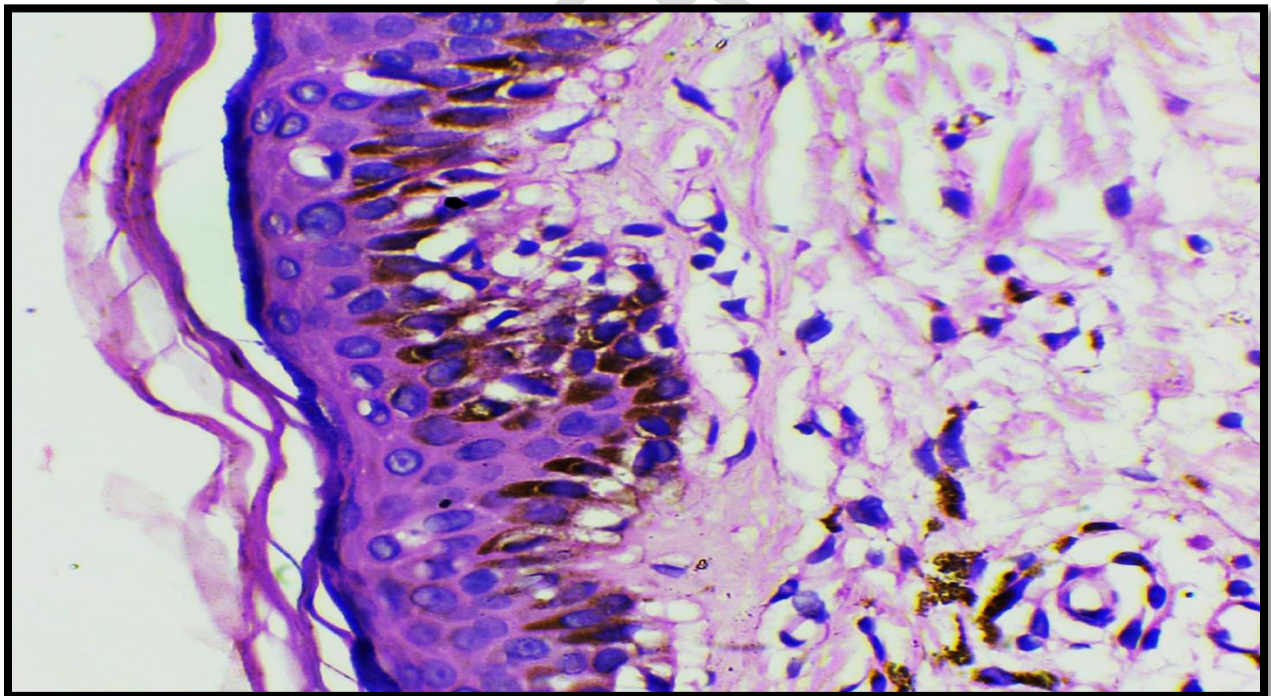
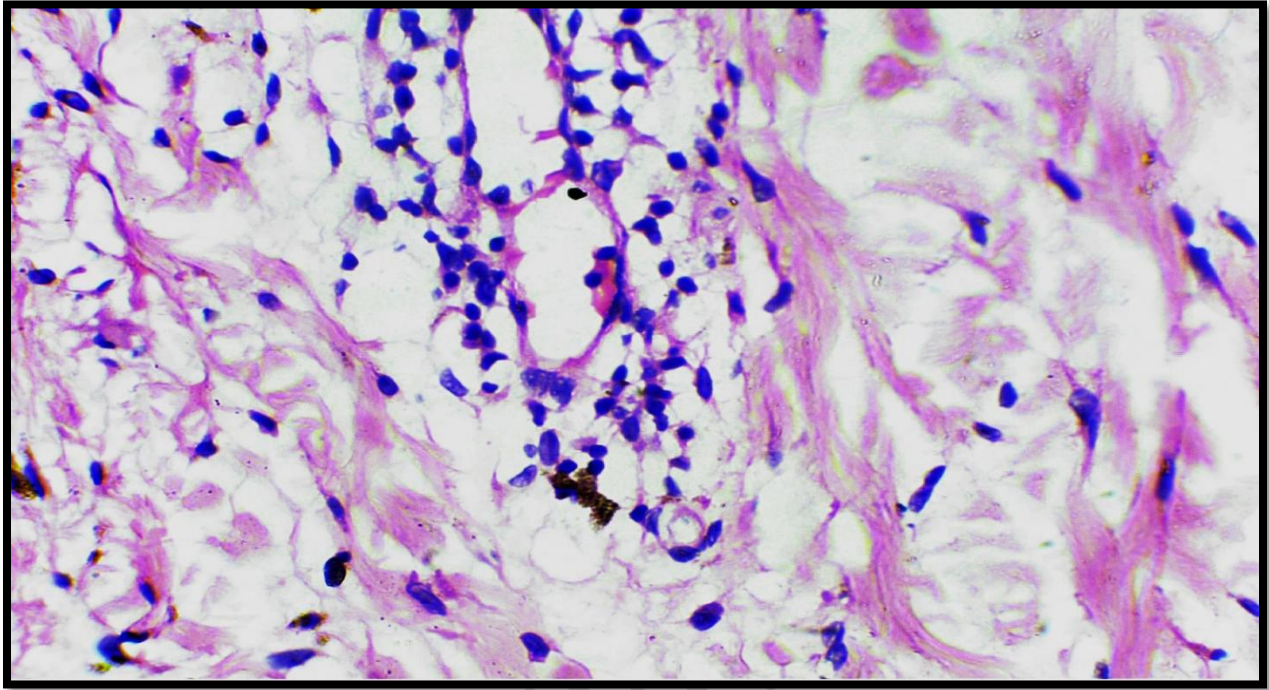


Figure 3: Capillaries surrounded by lymphocytes



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