Erythema Dyschromium Perstans – A Rare Case Report of Dermal Hyperpigmented Skin Lesion

KEYWORDS

Dr. Priyadarshini D
Assistant professor, Department of Pathology, Shridevi Institute of Medical Sciences and Research Hospital
Near Sira Gate, Tumkur -572106, India

Dr. Satish Arakeri
Assistant professor, Department of Pathology, DMWIMS, Naseera nagar, Meppadi (PO), Kalpetta, Kerala-673577, India

ABSTRACT

A 20-year-old healthy man with skin complained of a 6 months history of progressive hyperpigmentation on the trunk and abdomen. There was no preceding dermatitis. The lesions were asymptomatic. He did not use any prescription or over-the-counter drugs or any herbal supplements. A full review of systems was negative. Physical examination revealed diffuse symmetric grayish brown patches on the trunk and abdomen (Figure 1). The lesions were nonpalpable and without erythema. Two adjacent 4-mm punch biopsies were performed on the skin over abdomen, one consisted of unaffected skin and one of hyperpigmented skin. (Figure 2) shows affected skin with an interface dermatitis and significant pigment incontinence. There was no evidence of depositional process of substances such as heavy metals, drugs, or tattoo. Differential diagnosis included erythema dyschromicum perstans, fixed-drug reaction, or interface drug reaction. (Figure 3) Fontana Masson stain revealed prominent dermal melanin deposition.

Materials.
1. Punch biopsy or elliptical biopsy needle
2. 10% Formalin
3. Automated tissue processor
4. Rotary Microtome
5. Clean non grease glass slides
6. Haematoxylin & Eosin stain
7. Fontana Masson special stain
8. Microscope

Method
The site of the biopsy was selected, Elliptical skin biopsy is taken. As fixative 10% formalin can be used in almost all instances, The entire skin biopsy was submitted for routine processing and embedded in paraffin wax. 3-5mm thick paraffin sections of the skin biopsy were stained with haematoxylin and eosin. Fontana Masson special stain is used to confirm the epidermal and dermal hyperpigmented skin lesions.

INTRODUCTION

Erythema dyschromium perstans was first described in El Salvador by Ramirez in 1957 and was termed at that time dermatitis cenicienta or ashy dermatosis. The name erythema dyschromicum perstans did not emerge until four years later, in 1961, when Dr. Marion Sulzberger coined the term after describing a patient in Venezuela.

The etiology of EDP is unknown but several theories abound. Some authors consider it to be a variation of lichen planus actinicus. Other authors have considered the geographic distribution of EDP and suggested environmental factors as the etiology. However, all attempts to identify pollutants within the geographic distribution have been unsuccessful.

CASE REPORT

A male patient aged about 20 years presented with multiple brownish grey macules over trunk and abdomen since 6 months. There was no preceding dermatitis. The lesions were asymptomatic. He did not use any prescription or over-the-counter drugs or any herbal supplements. A full review of systems was negative. Physical examination revealed diffuse symmetric grayish brown patches on the trunk and abdomen

A preliminary haematoxylin and eosin staining followed by Fontana Masson staining for melanin is done in our study to differentiate epidermal and dermal hyperpigmentation.

Histopathology revealed vacuolar degeneration of basal layer, mild perivascular lymphocytic infiltrate and dermal pigment incontinence and Fontana Masson stain showed prominent dermal pigment incontinence.

FIGURE 1: Multiple brownish grey macules over trunk and abdomen with diffuse hyperpigmentation

FIGURE 2: Erythema dyschromium perstans H&E stain 40X showing basal cell degeneration and perivascular lymphocytic infiltrate and dermal pigment incontinence
DISCUSSION
Erythema dyschromium perstans also known as Ashy dermatosis is a slowly progressive, diffuse darkening of the face, arms, neck and trunk. It is a hypermelanotic disorder of the idiopathic variety characterized by bluish grey hyperpigmented macules of variable shapes and sizes. They may be symmetrical in distribution or unilateral. Early lesions may be reddish in colour, often with a more pronounced border, and they may be somewhat elevated. However, this phase is not always observed. The patient is otherwise well with no associated disease or blood test abnormality. Erythema dyschromicum perstans most often affects darker skinned patients, most frequently Latin Americans and Indians. However it has also been reported in people of lighter skin colour and various ethnicities. Erythema dyschromicum perstans may occur at any age but it appears to be more frequent in young adults. Women are affected more often than men.

The lesions have an active red border and lichen planus pigmentosus, occupational dermatosis with hyperpigmentation; drug related dermatosis, universal acquired melanosis, and familial progressive hyperpigmentation are the common entities confounding the diagnosis.

Disorders of pigmentation can result from migration abnormalities of melanocytes from neural crest to the skin during embryogenesis, impairment of melanosome transfer to keratinocytes and alteration in melanin synthesis. Since treatment strategy differs for epidermal and dermal hyperpigmented lesions, detailed histopathologic examination focused on existence of melanin deposits or melanocytes is important. Epidermal pigmentation disorders respond better than dermal pigmentation disorders which are difficult to treat. Triretinoin and hydroquinone are histologically effective for treating epidermal melanin deposits but not dermal melanosis or dermal melanocytes.

Tlougan B et al in 2010 studied a case of Erythema dyschromium perstans in a male patient aged 39 years presented with brown-grey discrete and coalescing patches over Upper extremities, trunk, neck and face. Sparse, superficial, perivascular lymphocytic infiltrate with focal vacuolar changes in basal layer and dermal melanophages.

Chakrabarti N et al in 2012 reported a case of erythema dyschromium perstans in a male patient aged 20 years. Bluish grey macules over face arms, neck and trunk with nonspecific histological features.

Differential diagnosis for erythema dyschromium perstans include:
- Lichen planus pigmentosus
- Multiple lesions of fixed drug eruption
- Postinflammatory hyperpigmentation
- Urticaria pigmentosa
- Incontinentia pigmenti
- Pinta

Treatment options for erythema dyschromium perstans include:
- Topical steroids
- Exposure to ultraviolet radiation
- Pigment lasers and Chemical peels

The most successful systemic treatment has been clofazimine. Dapsone, griseofulvin, hydroxychloroquine, isoniazid and corticosteroids have been used successfully in a few cases.