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A Sporadic Case of Darier Disease with Acrokeratosis Verruciformis like Lesions: Treated Effectively with Oral and Topical Retinoid

Dr. Rahul Kumar Sharma M.D.*[1], Dr. Smriti Vyas[2]

Department of Dermatology, Venereology and Leprosy, GMCH Udaipur
Department of dermatology, Geetanjali medical college, Udaipur, Hiran magri Extn. Udaipur- 313002

Abstract:

Darier disease (DD) is a type of autosomal dominant genodermatosis with calcium pump defect. We are reporting a case of Darier disease because of its rarity. A 24 year old man presented with multiple pruritic dark warty lesions over the seborrheic areas since 5 years of age. We treated him with retinoids, with which he had considerable improvement. It is necessary to diagnose this condition early as it can adversely affects the quality of life and can be associated with multiple systemic co-morbidities.

Keywords: ***Darier disease, Acrokeratosis verruciformis, Isotretinoin, Keratosis follicularis, Cobblestoning.***

Introduction

Darier disease (DD) is a type of inherited genodermatosis with calcium pump defect.[1] It is also known as Keratosis follicularis.[2] It has an autosomal dominant inheritance.[3] DD is caused by the mutations of ATP2A2, which encodes an endoplasmic reticulum calcium pump, SERCA2.[1,4] The defect in the functioning of calcium pump leads to abnormal keratinization and acantholytic dyskeratosis (loss of adhesion between suprabasal epidermal cells).[1,2,3,4] DD often start in childhood and presents with small dirty warty greasy papules and plaques predominantly in seborrheic areas such as the face, chest and back.[3,4,5] During the evolution of DD, scales and scabs may gradually develop.[5] DD patients may develop multiple other organ diseases(eyes, bone, lungs, and urogenital abnormalities) and neuropsychiatric symptoms.[5,6,7] DD can be associated with internal malignancy.[5,8] We are reporting a case of Darier disease because of its rarity and as this case was having sporadic occurrence which is against of its autosomal dominant inheritance. Moreover he presented with acrokeratosis verruciformis like lesions and other classical findings making it interesting.

Case History

A 24 year old man, farmer by occupation, presented with multiple pruritic dark warty lesions over the seborrheic areas since 5 years of age with slow progression over a period of 10 years. There is a history of progressive darkening of skin lesions and increased pruritus over 15 years. There is no history of similar illness in the family. He was earlier treated and misdiagnosed elsewhere as acne without improvement. The clinical examination revealed multiple greasy dark coloured papules and plaques in seborrheic areas and vegetative plaques in flexures, groin, axillae and perineum. Oral examination revealed dirty white cobblestone like papules on the gingiva and palate. There was mild hyperkeratosis of the palms with small pit like depressions. The finger nails were showing classical longitudinal, red and white streaks and diagnostic V shape nicking supporting the diagnosis of DD. The histopathological examination revealed hyperkeratosis, papillomatosis, acantholytic dyskeratosis, suprabasal cleft, corps ronds and grains suggestive of Darier disease.

We treated this case with oral Isotretinoin (20 mg per day), Adapalene gel 0.1 % and weekly bleach bath, with which he had remarkable improvement in the thickening of skin and resolution of vegetative plaques in the flexures. He was also screened for systemic and neuropsychiatric diseases with relevant investigations, which were normal. He was also advised to have regular dental and eye checkup .The genetic counseling was conducted. He was also educated about the exacerbating factors of this disease and was advised to protect himself from extreme sunlight and hot weather.

Discussion

DD is often confused with conditions such as acne vulgaris, seborrheic dermatitis, psoriasis, familial benign pemphigus, grover disease and atopic dermatitis.[3,5] It is imperative to diagnose this condition early as it can adversely affects the quality of life of patient and can be associated with multiple

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systemic co-morbidities like neuropsychiatric illness and renal disease.[5,6,7,8] So the diagnosis and awareness at an early stage can improve the quality of life of the patients and prevent further complications

Figure 1 Multiple greasy dark coloured papules and plaques on seborrheic areas

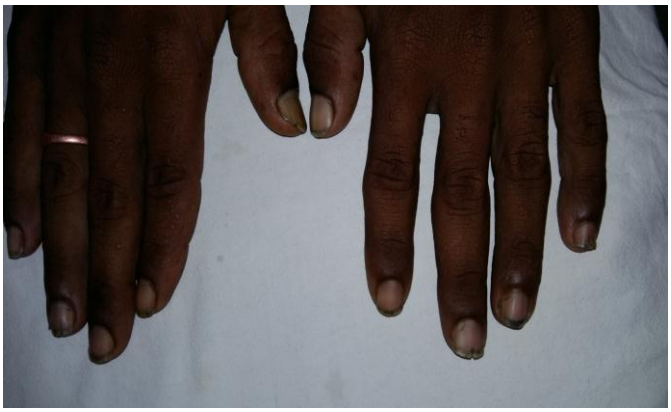

Figure 2 Acrokeratosis verruciformis like lesions over dorsal aspect and nails showing red streaks and diagnostic V shape nicking

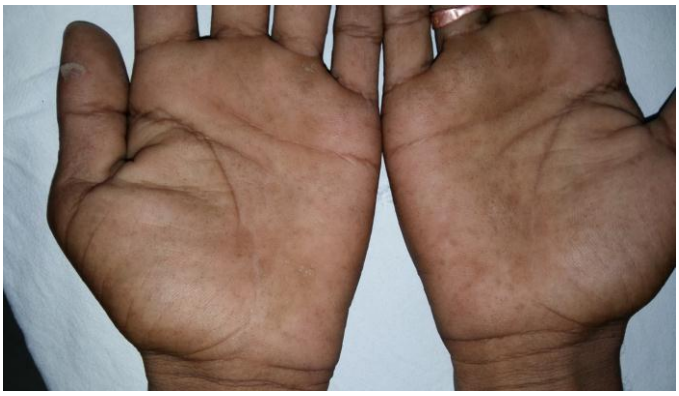

Figure 3 Mild hyperkeratosis and palmar pits

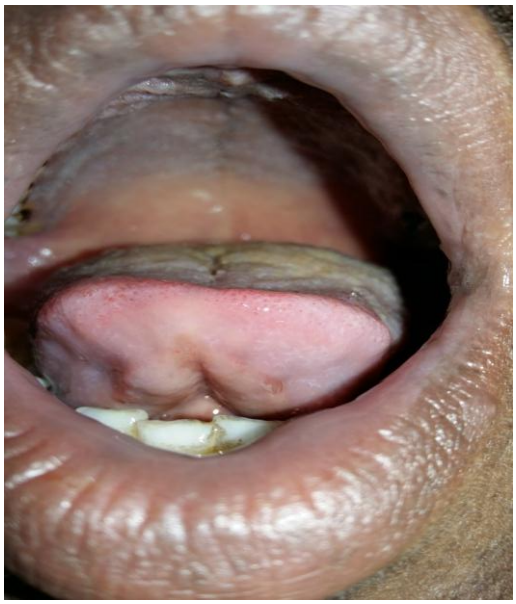

Figure 4 Dirty white cobblestone like papules on the palate

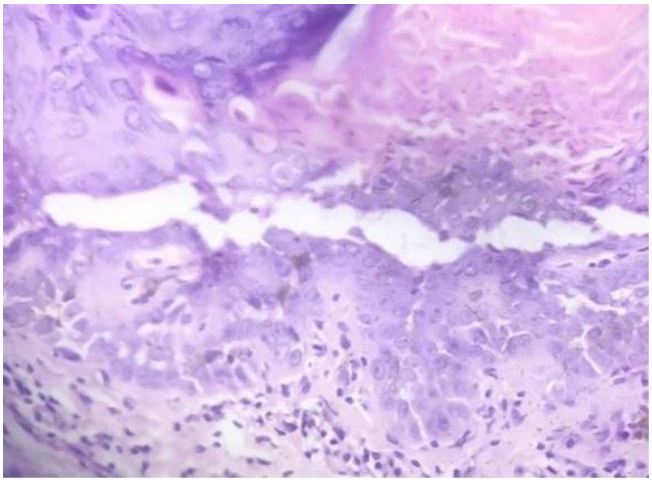

Figure 5 Histopathological examination revealed hyperkeratosis, papillomatosis, suprabasal cleft, corps ronds and grains suggestive of Darier disease

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