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Case Report

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Darier disease with seizures- A Coincidental finding or an identified syndrome?

***Oshin Agrawal, **BB Mahajan, ***Saurabh Sharma, ***Sunil Chaudhri, ***Richa Nagpal, ****Deepak Rana, *****N.S. Neki**

*Junior Resident, **Professor, ***Ex Junior Resident,
Dept. of Dermatology, Govt. Medical College, Amritsar
****Junior Resident, Dept. of Radiology, Govt. Medical College, Amritsar
*****Professor & Head, Dept. of Medicine, Govt. Medical College, Amritsar
Corresponding Author: **Dr. Saurabh Sharma,** Ex Junior Resident, Dept. of Dematology,
Govt. Medical College, Amritsar, India, 143001
E-mail: *dr.ssharma1985@gmail.com*

Abstract

Darier's disease is a rare autosomal dominant Genodermatosis characterized by a persistent eruption of hyperkeratotic greasy papules mainly over the seborrheic sites of the body, usually associated with nail abnormalities. The lesions typically appear in the younger age group and are associated with pruritus. We hereby report acase of Darier's disease associated with seizuresin a 30-year-oldfemale.

Keywords: Darier disease, Seizures

Introduction

Darier's disease or Keratosis Follicularis is a rare autosomaldominant disorder, which is characterized by greasy, keratotic, yellowish brown warty papules and plaques, particularly over the seborrheic areas. In about 47% of patients, there is no family history, presumably because of incomplete penetrance. The disease is causedby mutations in the ATP 2A gene, which encodes the sarcoendoplasmicreticulum Ca2+ ATPase.[1] This enzyme transportsCa2+ from the cytosol back to the endoplasmic reticulumlumen thus mediate stability and adhesion of desmosomes. Themutations in this gene affect Ca2+ homeostasis which results in an abnormality in desmosomal stability and adhesion.[2]

Case Report

A Thirty-year-old woman presented to us with the chief complaint of keratotic, greasy papules over the trunk since eight years. It started behind the ears [Figure 1] which gradually progressed to involve the face, trunk, dorsum of hands as well as dorsum of feet. Lesions were associated with moderate itching with no diurnal variation. There was a history of partial remission in winters and aggravation in summers.

Figure 1 : Multiple skin coloured greasy keratotic papules behind the ears

History of seizures was present for which patient was taking medication for six years. Family history was not significant. On examination, erythematous to skin colored keratotic papules were present behind the bilateral ears, face, neck, the central part of the back, inframammary area, buttocks and back of her hands and feet[Figure 2].Moderate greasy scaling was present over the scalp. Palms and soles were thickened.

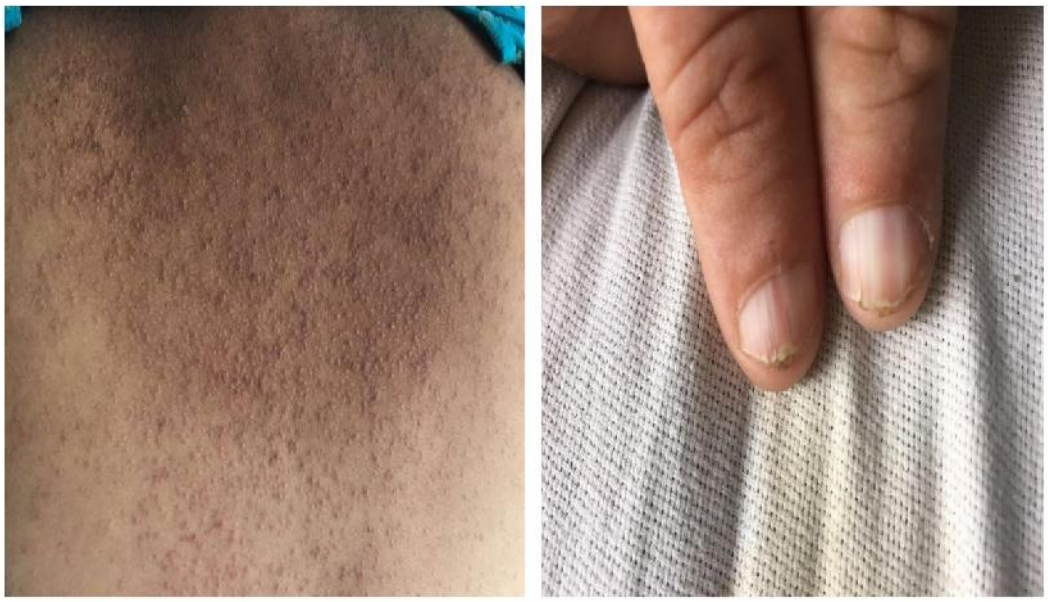

Figure 2: Multiple skin coloured greasy keratotic papules on trunk [Panel a], longitudinal streaks along with V shaped nicks

There were longitudinal white and red streaks along with V-shaped nicks at the free edges of both the thumb nails. Rest of nails were normal[Figure 2]. All mucosae were normal.

Histopathologically, biopsy revealed acanthosis, premature keratinization, suprabasal acantholysis and acantholytic cells in lacunar space, corps ronds and grains are seen. In the papillarydermis, mild perivascular lymphohistiocyticinfiltrate also noticed [Figure 3]. All routine investigations werewithin normallimit. On the basis of these clinical and histopathological findings, a diagnosis of Darier's disease was made.

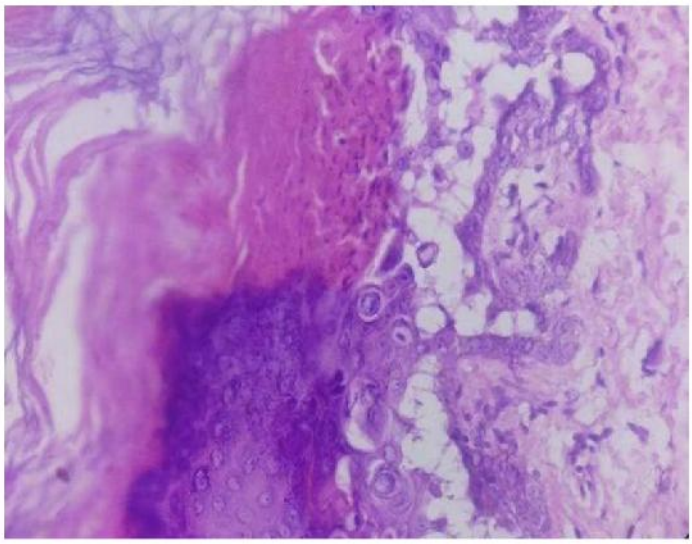

Figure 3: Suprabasal acantholysis, acantholytic cells in lacunar space and corps ronds with grains [H and E stain; X10]

Discussion

This disease was first described by Prince Marrow in 1886 and simultaneously by Darier andWhite in 1889, independently. In 1917, the first case with oral manifestation was reported by Reenstierna[3]. The prevalence of this disorder in population is1: 100,000. The sex incidence is equal, although the males appear to be more severely affected than females. The oral mucosa is affected in 50% of the cases[4-5]. Disease gets precipitated by heat and humidity, mechanical trauma like friction, sunlight, and secondary bacterial infections[6].

Histologically, suprabasal clefts are present in the epithelium, with acantholysis and dyskeratotic cells present as "corps ronds" which are usually presented in the granular layer and consist of irregular eccentric and sometimes pyknotic nuclei. The grains are seen in horny layer[7].

Associated anomalies have been described which include seizures, mental retardation, andpsychosis[8]. Our patient also presented with associated h/o seizures. It may be just coincidental finding or associated with the disease as part of some unidentified syndrome.

Although *Seborrheic* dermatitis is also known to occur with antiepileptic medications so the differential diagnosis of *Seborrheic* dermatitis was also kept in this case. Other clinical differentials include Acrokeratosis verruciformis of Hopf and Plane warts, Hailey-Hailey disease.

In treatment options, we can use topical agents like retinoids, salicylic acid, and lactic acid[9].Systemicretinoidsand lasers which include erbium: YAG lasers are also helpful in chronic recalcitrant cases[10].

Darier's usually begins in childhood but can manifest at any age and in our case, it manifested in the third decade. Our case is associated with seizures (rare association with Darier's disease) for which she is taking regular medicine from a physician. After thorough investigations and proper counseling, the patient was put on isotretinoin after taking opinion from physician and patient responded well to the treatment.

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