A case of zosteriform Darier's disease with seasonal recurrence

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ABSTRACT

Darier's disease is an uncommon genodermatosis characterized by keratotic papules in seborrheic distribution. The disease can rarely present in unilateral zosteriform pattern, as a mosaic form following the Blaschko's line. We present a 35-year-old woman with zosteriform pattern of Darier's disease over right infra mammary region. The lesions occurred strictly during summers. Histologically, suprabasal acantholysis with abundant dyskeratotic cells were seen.

INTRODUCTION

Darier's disease (DD), described independently by Darier and White in 1889, is an autosomal-dominant disease characterized by persistent eruption of hyperkeratotic papules, histological examination of which shows suprabasal acantholysis with distinct overlying dyskeratosis.[1] Expressivity is variable but the penetrance is complete in adults.[2] DD is caused by mutation in the ATP2A2 gene at chromosome 12q24.1, which encodes the sarco endoplasmic reticulum calcium pumping ATPase type 2 (SERCA2).[3] Clinically, the disease is characterized by keratotic papules or plaques in the seborrheic areas with occasional nail or mucosal involvement. The disease shows aggravation during summers and sun-exposure; heat and humidity may exacerbate the condition.[4] Localized pattern of DD was first reported in 1906.[5] Since then, many localized variants like localized, unilateral, linear,segmental or zosteriform DD[4-9] have been described. We present a case of zosteriform DD confined to the right infra-mammary region in an adult female. Interestingly the lesions recurred every summer and then regressed spontaneously.

CASE REPORT

A 35-year-old woman presented with mildly pruritic, unilateral, hyperpigmented, keratotic papules localized to the right infra mammary region [Figure 1]. For the past 3 years, these lesions occurred strictly in summers, from March to June, and regressed spontaneously afterwards. There was no history of similar skin lesions in any of the family members. No oral, nail or hair abnormalities were detected. General physical examination was normal. Routine hematological and biochemical investigations were normal. Histopathological examination of the skin biopsy showed hyperkeratosis, elongation of rete ridges, suprabasal acantholysis and marked dyskeratosis in the form of corps ronds and grains [Figures 2 and 3] along with a mild lymphohistiocytic inflammatory infiltrate in the dermis consistent with DD.

DISCUSSION

DD is an inherited autosomal-dominant condition with prevalence of around 1/50,000–100,000.[1] Around 10% of the cases of DD present in a localized pattern.[4] Localized pattern

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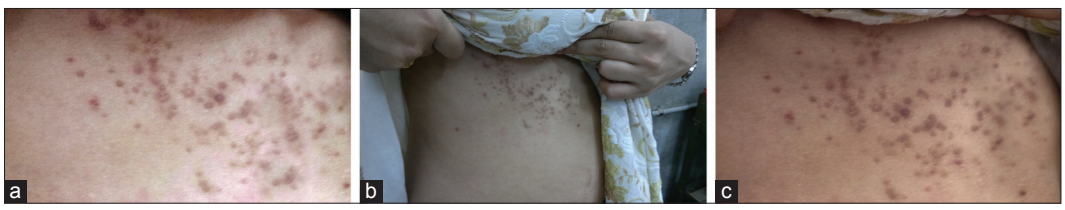

Figure 1: (a-c) Hyperpigmented, keratotic papules localized to the right infra-mammary region in a zosteriform distribution

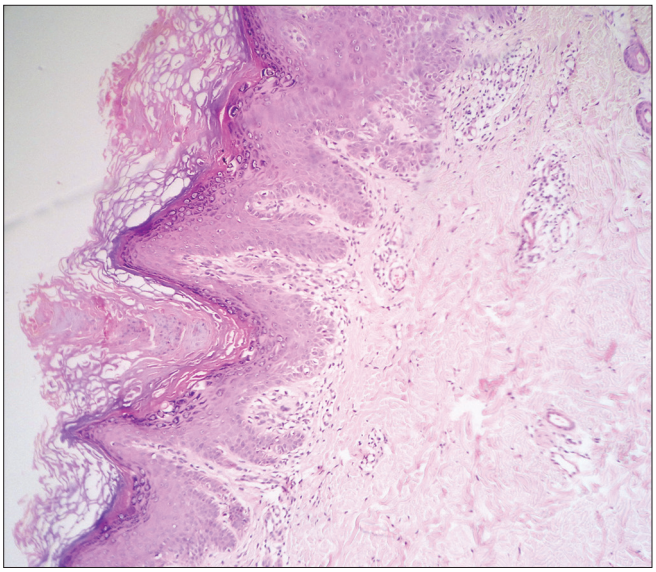

Figure 2: Photomicrograph shows hyperkeratosis, elongation of rete ridges, suprabasal acantholysis and dyskeratosis [H & E, 40x]

of DD was first reported in 1906.[5] Since then, various localized variants like unilateral, linear, segmental or zosteriform have been described.[4-9] The skin lesions in the localized variants are usually confined to a limited area and other features that are associated with classical DD are usually absent. The lack of family history, absence of other signs of DD and the limited distribution of the lesions favor a nevoid origin.

Two types of segmental variants of DD have been observed. The more common type I has a unilateral distribution along the Blaschko's line.[10] The severity and the histological findings within the streaks do not differ from those of generalized DD. This distribution is the result of genetic mosaicism due to post zygotic somatic mutation early in embryogenesis. If there is associated gonadal mosacism, a patient with segmental manifestation may have an offspring with generalized DD.[9]

In the type II segmental variant, patients with generalized DD have linear streaks with increased severity.[6] Type II mosacism occurs in patients with heterozygous germline mutation who also have a somatic loss of heterozygosity of wild type allele in a segmental area leading to homo- or heterozygosity and therefore increased severity in a linear array.

Segmental (localized, linear or zosteriform) forms of DD may be clinically and histologically indistinguishable from epidermal nevi with acantholytic dyskeratosis (ADEN).[11] It is believed that ADEN possibly represents the localized, mosaic form of DD, and it has been suggested to replace the term "ADEN" by "segmental DD," induced by post-zygotic mutation.[10] A case of ADEN on the sole of a 1-year old infant that showed histological features of DD has recently been reported,[12] further supporting this view. The diagnosis of DD can definitely be confirmed if SERCA mutations are found.[11]

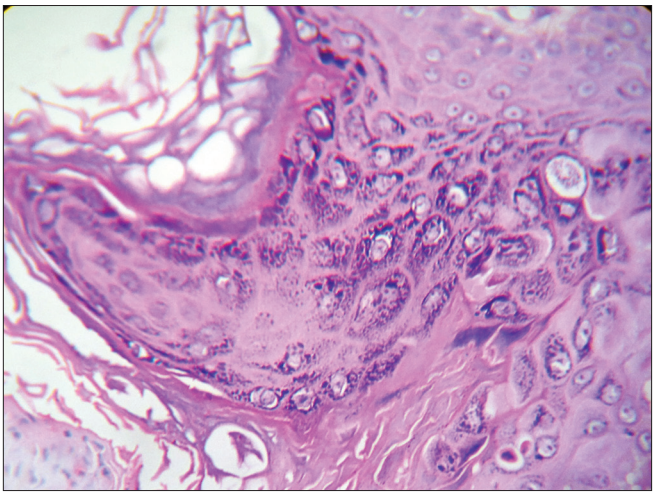

Figure 3: Photomicrograph shows characteristic corps ronds and grains [H & E, 100x]

However, cases of segmental DD not associated with SERCA mutations, mutations of other probably as yet unknown genes and controlling the differentiation of epidermal keratinocytes has been hypothesized.

Our patient seems to exemplify type I segmental DD. Another interesting observation noted in our patient was history of lesions occurring strictly during summers and their complete spontaneous disappearance during the other seasons. A similar observation has also been reported by Plantin[7] *et al*. and Lin[4] *et al*.

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