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CASE REPORT

DARIER'S DISEASE MIMICKING CONDYLOMA ACCUMINATA, UNUSUAL PRESENTATION: A CASE REPORT

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ABSTRACT: Darier white disease is determined by autosomal dominant gene or it is determined by mutation of gene. The follicular and non-follicular hyperkeratotic papules with waxy scales are constant features. However the disorder rarely presents along with a myriad of unusual cutaneous lesions. Linear, unilateral, bullous cornifying or solitary hypertrophic lesions are some such varieties. We are reporting a case of Darier's disease presented with condyloma accuminata like growth present over external genitalia for its rarity.

KEYWORDS: Darier's disease, warty dyskeratoma, condyloma accuminata.

INTRODUCTION: Darier white disease is determined by autosomal gene or it is determined by mutation of gene.[1] The follicular and non-follicular hyperkeratotic papules with waxy scales are constant features. However the disorder rarely presents along with a myriad of unusual cutaneous lesions. Linear, unilateral, bullous cornifying or solitary hypertrophic lesions are some such varieties.[2] We are reporting a case of Darier's disease presented with condylomata accuminata like growth over external genitalia for its rarity.

CASE REPORT: A 35 year old female born to non-consanguineous marriage, presented with skin lesions of 15 years duration, consisting of multiple brown black, warty, greasy, follicular and non-follicular papules, some of which also showed keratin plugs, mainly over the seborrhoeic areas, few papules were also present over the dorsa of fingers, hands, feet and both shins. Red and white bands and V shaped nicking at the free margins were noted on the finger nails.

Hyperkeratotic verrucous (Warty) growth present over the external genitalia and preianal area, few areas showing erosions. [Fig. 1 & Fig. 2]

Routine investigations were within normal limits. VDRL, HIV1 & 2 and HbsAg were negative. Punch biopsy from the lower back and excision biopsy from external genitalia were showed hyperkeratosis, suprabasal lacunae, corps-ronds and grains and hyperkeratotic invagination filled with keratinous material and acantholytic cells suggestive of Darier's disease. [Fig. 3]

DISCUSSION: The disease was first reported independently by Darier and White in 1889. Darier's disease (DD) is a rare congenital acantholytic disorder transmitted as an autosomal dominant trait. It may also occur as mutation. Both sexes are affected with equal frequency. It usually begins in childhood but can manifest at any time.[3]

Mutation in the gene ATP2A2 cause Darier's disease. ATP2A2 located on 12q23-24, encodes the endoplasmic reticulum Ca 2+ ATP, which is a calcium pump. This pump maintains low cytoplasmic Ca2+ve level by actively transporting Ca ions from the cytosol in to the lumen of the endoplasmic reticulum.[4]

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Electron microscopy reveals loss of desmosomes, breakdown of desmosome-keratin intermediate filament attachment, and perinuclear aggregates of keratin intermediate filaments.[5]

The lesion may first appear as skin colored or yellow brown papules with a greasy, warty texture. The lesion are especially common in seborrhoeic areas such as the forehead, scalp, margin of the scalp, nasolabial folds, ears, chest and back. In less than 10% of patients, flexural disease predominates, with large, warty, vegetative papules in the axillae, groins or perineum. Frequently associated with pruritus. Heat, sweat, humidity, sunlight, UVB exposure, lithium, oral corticosteroids and mechanical trauma have been reported to exacerbate this condition.[6]

Involvement of hands include punctate keratosis, palmar pits and haemorrhagic macules, acrokeratosis verruciformis like lesions are present in half the patients. Nail changes like white and red longitudinal bands, longitudinal ridges, V shaped nick and subungual hyperkeratosis are frequently found.[7] Mucosal lesions appear as white papules with a central depression (cobblestone) lesions commonly seen in the mouth, but can also occurs in genital mucosa.[8]

The histopathological finding, irrespective of clinical presentation, characterized by focal changes of acantholysis and presence of dyskeratotic cells resembling Corps ronds and grains associated with suprabasal clefts around the preserved villi, hyperkeratosis and parakeratosis. These findings are classical of Darier's disease, but are also found in Hailey Hailey disease, Grover's disease and warty dyskeratoma.[9]

In 1984, Chorzelski et al reported a 23-year old female with multiple asymptomatic dome-shaped papules on the labia majora. Clinically, this was different from Hailey-Hailey disease in that papules were persistent for more than six years and there were no vesicles, fissuring and erosions. Histology showed hyperkeratosis, parakeratosis with some areas showing intraepidermal vesiculation and acantholysis similar to Hailey-Hailey disease, while other areas were more like Darier's disease showing dyskeratotic cells throughout the epidermis, including Corps ronds and grains. Skin and serum immunofluorescence were negative. The author regarded this as a new entity and named the disease as Papular acantholytic dyskeratosis (PAD).

PAD is an uncommon disorder of the genitalia, usually asymptomatic but sometimes may be pruritic. It is more commonly reported in females, with lesions occurring on the vulva and perianal area, but there has also been a report of a male patient with lesions over the penile shaft.[10] Clinically, PAD must be differentiated from genital papules of infectious origin, Hailey-Hailey disease, localized Darier's disease of genital area and various pre-malignant and malignant conditions.[9]

In our case female patient presented with condyloma accuminata like growth over external genitalia and perianal area, which is rare clinical morphology of Darier's disease and confirmed by histological findings. It is clinically differentiated by PAD and histologically by warty dyskeratoma. We are reporting this case for its morphological rarity.

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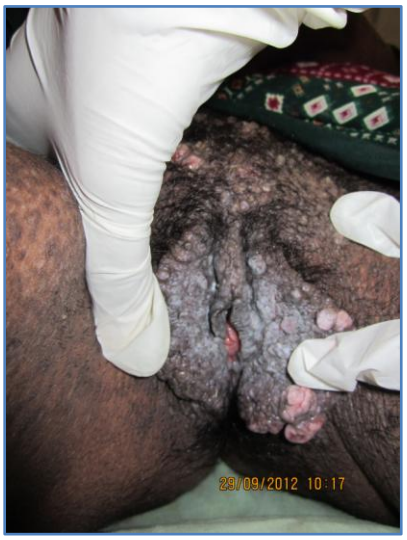

Fig. 1: Hyperkeratotic verrucous growth present over external genitalia

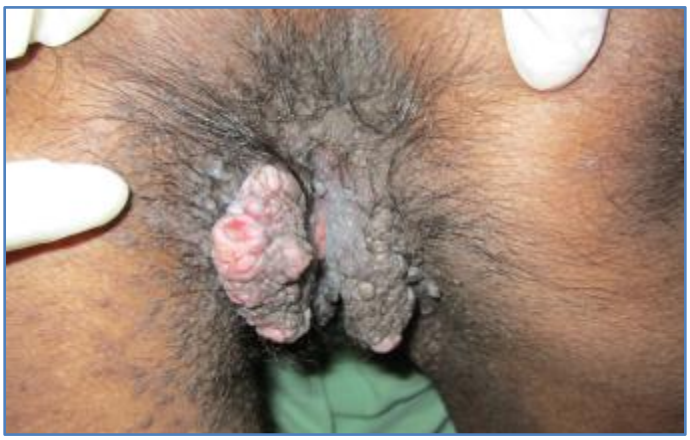

Fig. 2: Hyperkeratotic verrucous growth present over perianal area

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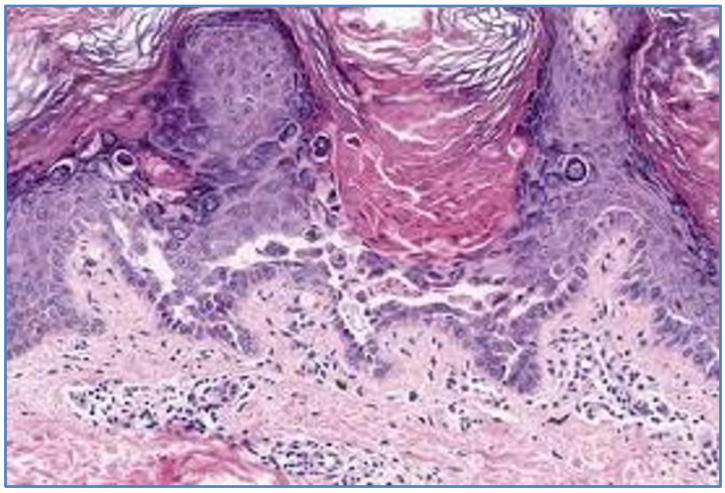

Fig. 3: Showing hyperkeratosis,suprabasal lacunae,corps ronds & acantholytic calls

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