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Case Report

Darier's Disease: A Case Report

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Abstract

Darier's disease or keratosis follicularis is an uncommon disorder of keratization which is autosomal dominant and have variable penetrance. It begins in adolescence or early adult life with discrete or confluent, brown or skin colored foul smelling, greasy, crusted, dirty looking, papules or plaques at the seborrhoeic sites, such as scalp, face, nasolabial folds, chest, back, axillae & groins. Palms & soles also may show minute pits & dorsal of hands shows small flat discrete papules. Sometimes cobblestone papules occur on the palate. Nails may have angular nicks. Sometimes Darier's disease is asymptomatic. It may run a chronic course with exacerbation in summer & relative remission in winter. Histopathology shows supra-basal acantholysis with dyskeratosis. Treatment includes avoidance of triggering factors like- sweating, frictions, UVB etc. and topical emollients such as- urea & lactic acid, topical Corticosteroid; topical & systemic retinoid can be given. Here we present a case of Darier's disease with typical skin involvement.

Key words : Darier's disease, Darier- White Disease, keratosis follicularis, Dyskeratosis.

Introduction

Darier's disease (DD), or keratosis follicularis is a rare genodermatosis was described independently by White and Darier in 1889. It is an autosomal dominant disease with variable expressivity. It is more common in female than male and the total ratio is 1 in 50,000-1,00,000 people. Darier's disease is clinically manifested by hyperkeratotic papules primarily affecting head, neck and thorax. Clinical features vary widely, involvement range from typical nail changes only to generalized disease. The disease runs a chronic relapsing course that persists throughout life, although it don't affect general health much. Exacerbations of Darier's disease can occur from exposure to heat, sunlight, trauma, UVB and during menstruation. Histology shows dyskeratosis in spinous layer (corps ronds) and stratum corneum (grains), suprabasal acantholysis and clefts. Basic measures to manage Darier's disease may include using sunscreen, avoiding hot environments. Moisturizers with urea or lactic acid can reduce scaling of the skin. A mild to moderate potency topical steroid and topical antibiotics are often useful for inflammation. Topical medications may include topical retinoids. Oral retinoids is the most effective medical treatment for Darier's disease. Other treatments may include oral antibiotics to clear bacterial infection, oral acyclovir to treat or suppress herpes simplex virus infection; dermabrasion; electrosurgery. We report a 15 years old female having malodourous greyish colored warty plaques in sebaceous distribution areas for three years. There was no mucosal involvement. Skin biopsy revealed abnormal formation of keratin tissue and acantholysis. Topical retinoid adapalene was prescribed along with topical 10% urea cream. Advice given to improve general hygiene, preference of cotton clothes, avoiding heat and sunlight.

Case report

A 15-year-old female presented with eruptions on different parts of the body for the last three years with occasional malodor and bleeding from the same areas. Eruptions were associated with severe pruritus. The rash became itchy and infected during summer. There were no associated systemic complaints. The disease is progressive. Her parents or siblings were not affected by the same disease. The patient presented with greyish colored warty plaques scattered on the

forehead, anterior scalp area [Figure 1], both aspects of forearm [Figure 2], back [Figure 3], abdomen [Figure 4] legs and soles [Figure 5]. The lesions were rough and firm in consistency, mostly non-tender but few on the legs were tender. After removal of the crust, there was eroded surface with bleeding points. Palms and soles showed some pits. Nails were fragile having subungual hyperkeratosis, splintering, triangular nicking and alternate white and red streaks. Oral mucosa appeared normal. No lesions were seen in other parts of the body. Blood counts, sugar, urea, creatinine and electrolytes were all normal.

Skin biopsy from back showed focal suprabasal clefts with acantholytic cells, corps ronds in stratum malpighii, grains in stratum corneum and dermal mild chronic inflammatory cells infiltrate. No granuloma or malignant cells were seen.

As a part of treatment, patient education, counseling was done. She was put on topical 10% urea cream along with advice to improve general hygiene, wearing of cotton clothes, avoidance of heat and sunlight as much as possible. Topical retinoid adapalene was also prescribed. The option of oral retinoids(acitratin or etretinate) was not considered in the treatment plan; because of patient's age at child bearing potential[9]. On follow up visits with monthly intervals the skin condition improved.

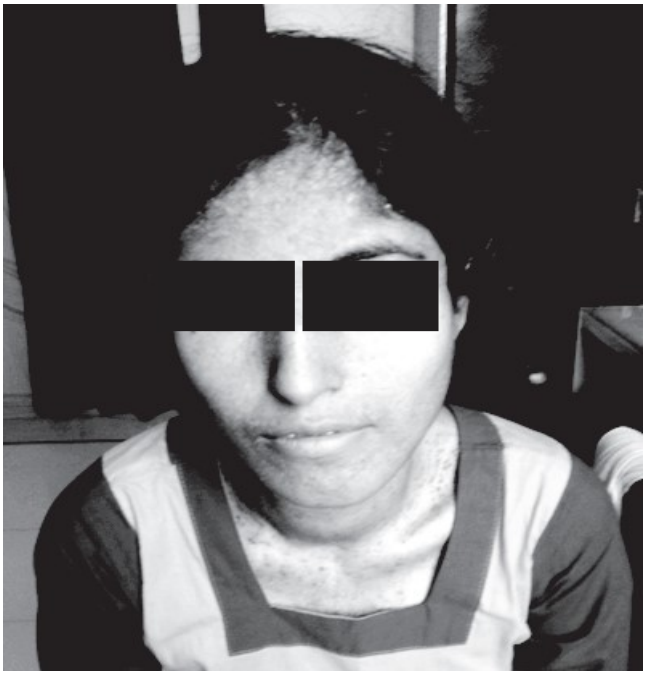
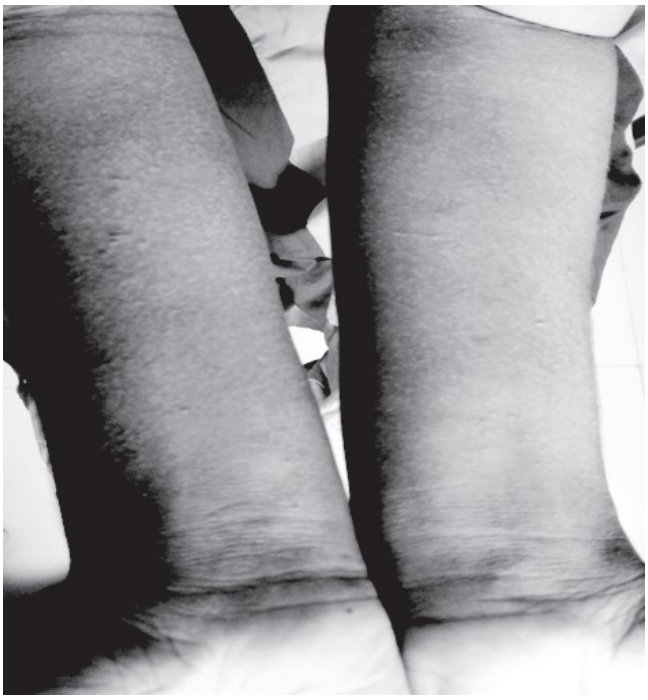


Figure 1



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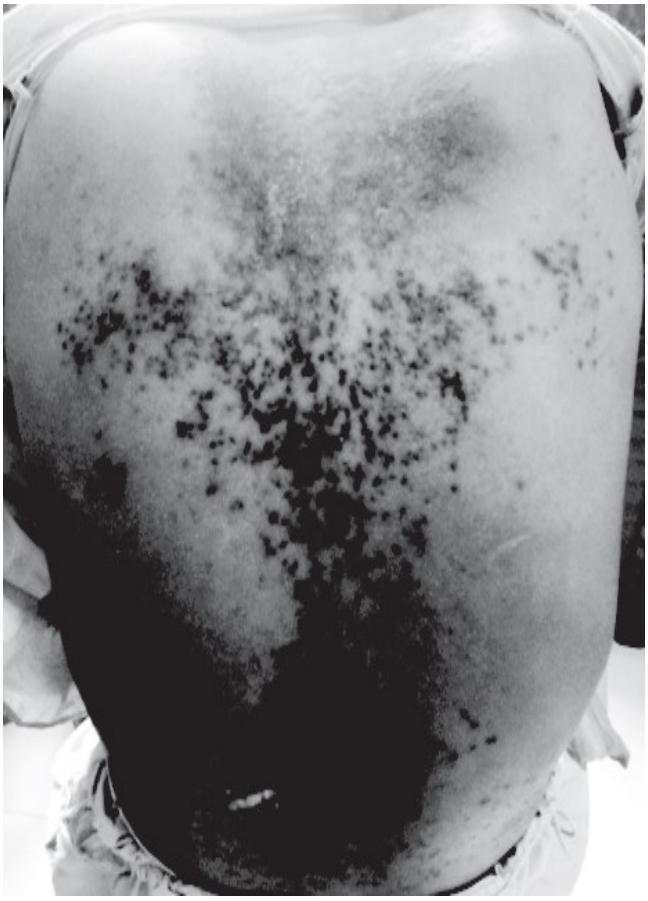

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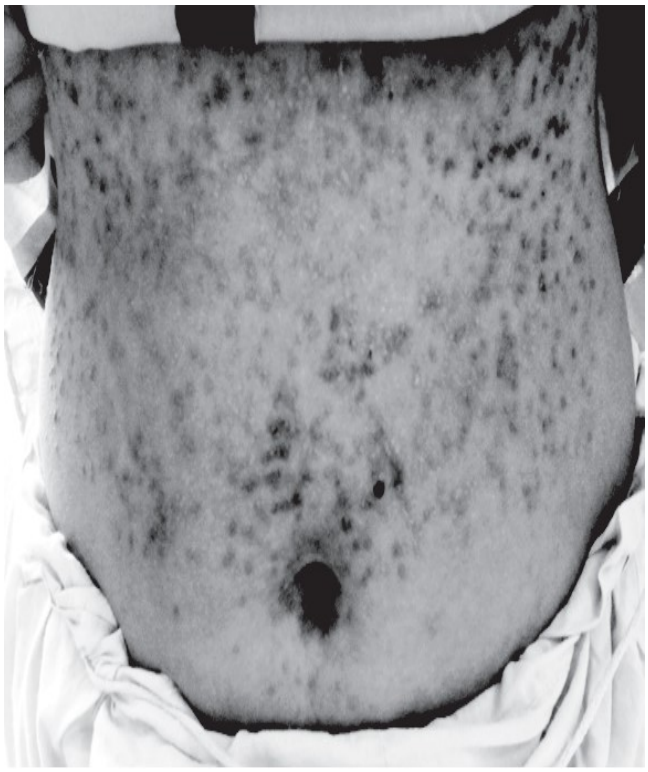

Figure 4

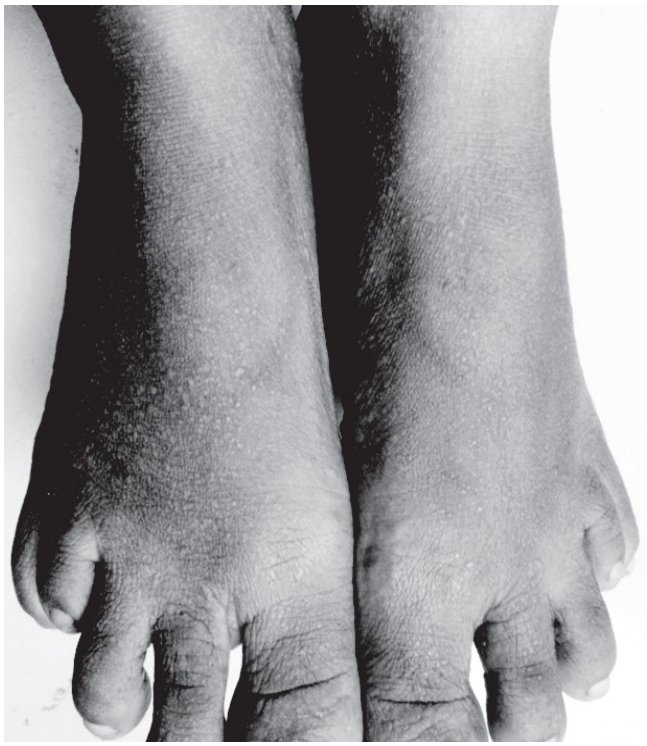

Figure 5

Discussion

Darier's disease, also known as-Darier–Whitedisease, keratosis follicularis, is an autosomal dominant disorder discovered by French dermatologist Ferdinand-Jean Darier. Darier's disease is a raregeneticdisorder manifested by scaly and/or crusted papules. Darier's disease is classified as a hereditary acantholytic dermatosis[1]. The abnormal gene in Darier's disease is located on chromosome 12q23-24.1 and is identified as ATP2A2. This gene codes for the SERCA enzyme or pump (SarcoEndoplasmic Reticulum Calcium-ATPase)[1,2]. This enzyme is required to transport calcium within the cell. The exact mechanism by which this abnormal gene works is not clear but it appears that it works by causing disruption of attachment of cells of skin[1]. The skin cells or keratinocytes are stick together by desmosomes and the gene causes failure of proper assembling of desmosomes[1,2]. Failure of assembling may be due to insufficient calcium. The symptoms and signs of Darier's disease vary between individuals. In an affected person the severity of the disease can fluctuate over time[3].

The skin lesions are characterized by greasy, scaly papules. Affected sites include seborrhoeic areas of the face, scalp and neck, central chest and back, skin folds, such as armpits, groin, under the breasts and between the buttocks [4]. Several of the small papules may grow together and form larger warty lesions. Crusted rash similar to seborrhoeic dermatitis may be found [5].

Darier's disease is usually diagnosed by its appearance. Family history is also important. Confirm diagnosis may require a skin biopsy[4,5]. The histologic feature are characteristic, focal acantholytic dyskeratosis[1,5,6].

Flare-ups may be caused by exposure to sunlight or heat, bacterial infection, herpes simplex infection. In most patients, general health remains good [6]. Darier's disease has occasionally been reported in association with neuropsychiatric disease. The lifetime prevalence of major depression (30%), suicide attempts (l5%), and suicidal thoughts (31%) appears higher than in the general population, highlighting the need for careful assessment.[1] Our patient refuses to go to school for her dipressive condition.

The implications of Darier's disease are cosmetic and aesthetic[5]. Milder forms of the disease respond to general measures which include improvement of hygiene, avoidance of heat and sunlight, wearing cotton clothes[6,7]. Using sunscreen is also proved beneficial. Moisturizers having urea, topical retinoids like adapalene, 0.1% tretinoin, tazarotene gel are proven effective in decreasing hyperkeratosis[6,7]. Oral retinoids also decrease hyperkeratosis. Oral antibiotics and acyclovir are often used to suppress secondary bacterial and viral infections. Severe inflammatory exacerbations may respond to ciclosporin[8]. Topical 5-Fluoro urocil, Laser surgery, Electrosurgery, Dermabration, Photodynamic therapy are other treatment options with unproven efficacy.[8] As a part of treatment, patient should be informed about the complications, emotional status should be evaluated and the patient should receive genetic counseling[5,6,8].

References

1. Lowell A.G, Kortz S.I, Bavbana A.G. Fitzpatricks Dermatology in General medicine,eighth edition, Vol-1,2012, p.550-556
2. Judge MR, McLean WHI, Munro CS. Disorders of keratinization. In : Burns T, Breathnach S, Cox N, Griffiths, eds. Rook's Textbook of Dermatology, 8th edition, Vol. 1. Massachusetts, Oxford, Victoria: Blackwell Science; 2010. p.19.81–6.
3. Puri N. Original Article: A clinical and histopathological study of Darier's disease. J Pak Assoc Dermatologists. 2011;21:230–4.
4. Burge SM. Darier's disease, keratins and proteases: A review. J R Soc Med. 1989;82:673–6. [PMCID: PMC1292374] [PubMed: 2480450]
5. Burge SM, Wilkinson JD. Darier-White disease: A review of the clinical features in 163 patients. J Am Acad Dermatol. 1992;27:40–50.
6. Abe M, Inoue C, Yokoyama Y, Ishikawa O. Successful treatment of Darier's disease with adapalene gel. *Pediatr Dermatol*. 2011 Mar-Apr. 28(2):197-8.
7. Dicken CH, Bauer EA, Hazen PG, Krueger GG, Marks JG Jr, McGuire JS, et al. Isotretinoin treatment of Darier's disease. *J Am Acad Dermatol*. 1982 Apr. 6(4 Pt 2 Suppl):721-6.
8. Katz TM, Firoz BF, Goldberg LH, Friedman PM. Treatment of Darier's disease using a 1,550-nm erbium-doped fiber laser. *Dermatol Surg*. 2010. 36(1):142-6.
9. Lowell A.G, Kortz S.I, Bavbana A.G. Fitzpatricks Dermatology in General medicine,eighth edition, Vol-2,2012, p.2767.