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CASE STUDY

DARIER'S DISEASE WITH PERIFOLLICULAR HYPOPIGMENTION – A RARE ENTITY

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Key message: We present this case because of the rarity of its occurrence and to the best of our knowledge it's the 15th such case reported ever worldwide.

ABSTRACT

Darier's disease is characterised by skin colored, yellow brown or brown firm rough papules in the seborrhoeic rich areas of scalp, face and trunk. The disorder can also present with a myriad of unusually rare cutaneous lesions like small leukodermic macules. We present such a case of darier's disease with hypopigmented maculopapular lesions because of the rarity of its occurrence.

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INTRODUCTION

Darier-White disease is an autosomal dominant disorder and is characterised by follicular and non-follicular hyperkeratotic papules with waxy scales in a seborrheic distribution. It occurs primarily in late adolescence and early twenties with no sex predilection. The disorder can however also present along with a myriad of unusual and rare cutaneous lesions like linear, unilateral, vesicobullous, cornifying, or solitary hypertrophic lesions. (Griffiths *et al*., 1992) Small leukodermic macules are one such extremely rare manifestation of Darier's disease which was first described by Goddal and Richmond in 1965. (Goddal and Richmond, 1965) We present a case of darier's disease with hypopigmented maculopapular lesions in a young male, because of the rarity of its occurrence and to the best of our knowledge it's the 15th such case reported ever worldwide.

Case Report

A 17 year old male presented with history of itching and appearance of asymptomatic hypopigmented maculopapular lesions over his arms and legs of 3 years duration. Lesions started as asymptomatic perifollicular macules over legs which spread to arms and back over the period. Before presenting to us, he had been treated for follicular vitiligo, seborrheic dermatitis and pityriasis versicolor. In due course, he developed acneiform papules over his chest and back. History of exacerbation of itching and lesions on exposure to sunlight was present. There was no family history of similar lesions. On examination, there were multiple perifollicular hypopigmented maculopapular lesions over the dorsal aspects of both upper and lower extremities, more so distally, along with excoriated acneiform lesions over back, sides of neck and lower abdomen (Fig 1-4). He also had acrokeratosis verruciformis like lesions on dorsum of hands and feet (Fig1). Palmar pits were present (Fig 6). Nails changes were present in the form of longitudinal leuconychia along with notching of distal ends (Fig 7). Oral mucosa and genitalia were unaffected. All routine investigations were within normal limits. Skin scrapings from hypopigmented lesions didn't show any fungal elements thus ruling out the differential diagnosis of pityriasis versicolor. Biopsy was taken from the papular hypopigmented lesion. The histology of the papular lesion revealed the classic features of Darier's disease viz. suprabasal clefts, dyskeratotic "corps ronds" and "grains"; and an acanthotic epidermis topped by a hyperkeratotic, parakeratotic stratum corneum (Fig 5). Patient

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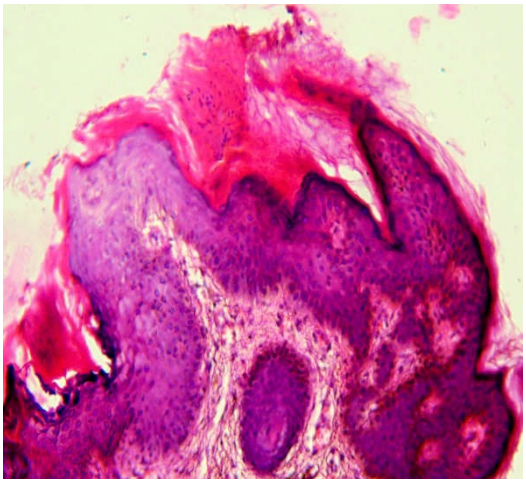
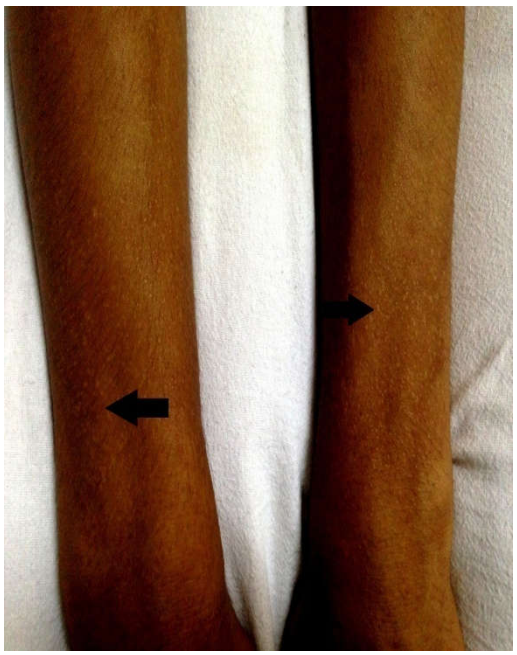
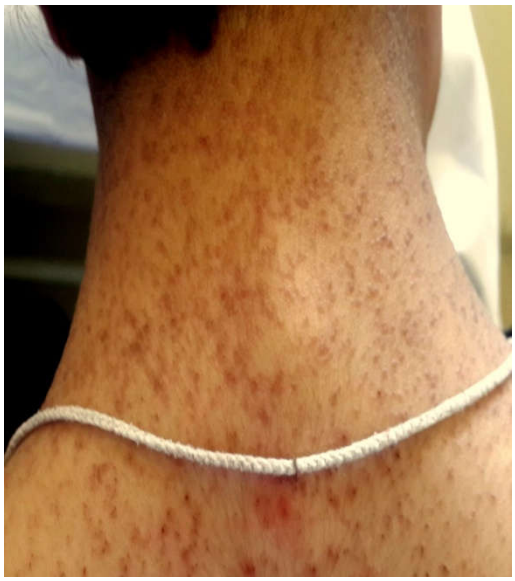
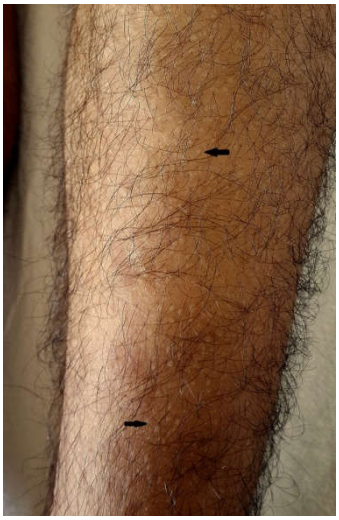
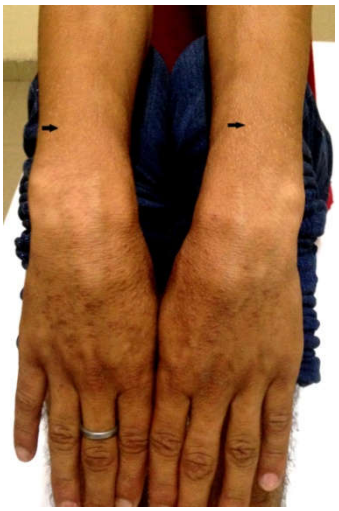
responded well to oral isotretinoin in the dose of 0.5mg/kg/day along with topical sunscreens and emollients with decrease in itching and lesions becoming less prominent.

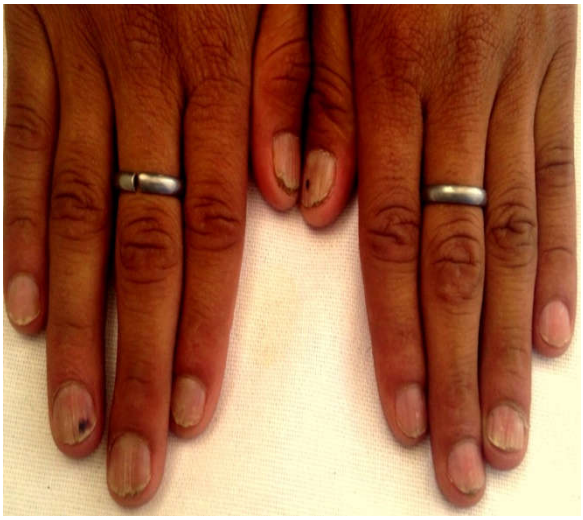


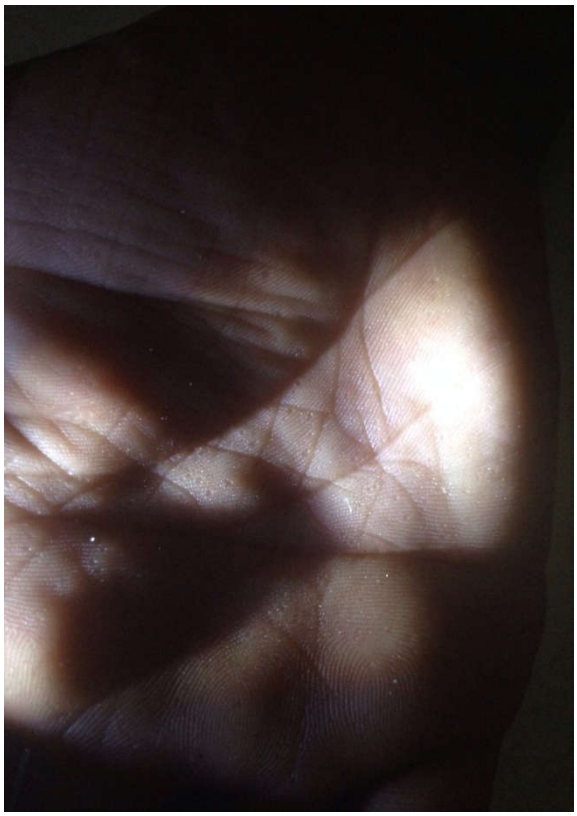












DISCUSSION

Darier's disease is an autosomal dominant condition characterized by a persistent eruption of hyperkeratotic papules in a seborrheic distribution. It occurs due to mutations in the *ATP2A2* gene which is responsible for maintaining high calcium concentration in the endoplasmic reticulum. The distinctive lesion of Darier's disease is a firm, rough papule, which is skin coloured, yellow–brown or brown. Seborrhoeic areas of the trunk and face, particularly the scalp margins, temples, ears and scalp, are most often involved. Rarely flat and freckle-like pale macular lesions (guttate leukoderma) in pigmented skin might be present. There have been early reports of such lesions since 1965. (Goddal and Richmond, 1965; Cattano, 1968; Cornelison *et al*., 1970; Bedi and Garg, 1978; Garg and Sait, 1985; Berth-Jones and Hutchinson, 1989) Udagani *et al* described such a case of 35 year old male with follicular, non-follicular, hypopigmented hyperkeratotic papules present over the scalp, face, shoulders and neck although plams, soles and nails were not involved. (Udagani *et al*., 1991) In our case though age of presentation was earlier (17 years) and palms and nails were involved. A similar case of leukodermic macules in Keratosis follicularis was reported by Jacyk and Visser (1992). Another case of Darier's disease with depigmented macules was described by Tolat SN in which a 42 year old male presented with numerous, discrete, depigmented macules scattered over his chest, abdomen, back, buttocks and thighs. A family history revealed that several members from 3 generations (vertical transmission), were similarly affected. (Tolat *et al*., 1994) In our case, no family history was present and hypopigmented maculopaular lesions were present only on distal aspects of legs and forearms only. Another similar case of Darier's disease with hypopigmented macules was reported by Bleiker and Burns (1998). Peterson *et al.* described a case of 45-year-old black woman presenting increasing number of "light spots" on her face, upper trunk, and legs along with pruritic eruption on the dorsum of her hands along with nail changes. Her sister and daughters also had similar complaints. (Peterson *et al*., 2001) In our case face was not involved, age of presentation was earlier and no family history. Gupta S and Shaw JC also reported a case of unilateral Darier's disease with unilateral guttate leukoderma. (Gupta and Shaw, 2003) In our case however, bilateral involvement was present. Another case of a 38 year old female was reported with hyperpigmented keratotic papules distributed over seborrheic areas and unusually hyperkeratotic papules and plaques over the extremities, especially over the shins along with hypopigmented macules over the extremities. Dorsa of the hands showed typical dome shaped brown papules, with a few of the finger nails showing distal notching and red longitudinal streaks. Palmar pits and typical oral mucosal lesions were also noted. (Pise *et al*., 2006) In our case, mucosal involvement was not present and unusually hyperkeratotic lesions were absent. Most recently a case of 22 year old male was reported by Sornakumar *et al.* in which patient presented with multiple perifollicular 3-4 mm hypopigmented lesions over his face, trunk, back and over the extremities. Genital and oral mucosae were not involved. Hair, nails, palms and soles were not involved. Biopsy confirmed the diagnosis of darier's disease. His father had similar hypopigmented lesions since child hood. (Sornakumar and Srinivas, 2010) In or case again, there was no family history and palms and nails were involved. Most patients with mild disease require no treatment other than emollients, simple hygiene and sun protection. Topical tretinoin and isotretinoin, adapalene and tazarotene are effective. Antiseptics are helpful in infected conditions. In severe disease, oral retinoids are usually effective; bothacitretin and isotretinoin are used. Dermabrasion, photodynamic therapy or laser treatments have also been proved useful although in limited areas. In our case, patient responded well with oral isotretinoin in the dose of 0.5mg/kg/day along with topical sunscreens, emollients and symptomatic treatment.

What's known: Dariers disease generally presents as skin colored, yellow brown or brown firm rough papules.

What's new: Flat lesions may also appear as pale macules (guttate leukoderma) in pigmented skin and are thought to be rare.

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