



DARIER-WHITE DISEASE: A RARE CASE REPORT

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ABSTRACT

Introduction: Darier-White Disease (DWD), also known as follicular keratosis, is a rare autosomal dominant genodermatosis characterized by impaired adhesion between epidermal cells and abnormal keratinization. **Case:** A 15-year-old woman came to the Dermatovenereology and Aesthetics clinic of Dr. Sitanala General Hospital with itching and red papules on the face, chest, and arms that had been present for 2 months before admission. A skin biopsy was performed to confirm the diagnosis. The patient was given tretinoin cream 0.05%, Melanox ES[®], and cetirizine 10 mg. **Discussion:** The diagnosis of DWD was established based on the clinical and histopathological findings. Clinically, multiple erythematous, greasy-appearing miliary papules with ill-defined borders and fine scales were observed in seborrheic areas, including the face, trunk, scalp margins, ears, neck, and upper extremities. Histopathological examination revealed focal

acantholytic changes and dyskeratotic cells (corps ronds cells) within the epidermis, consistent with the diagnosis of DWD.

Conclusion: This case illustrates the importance of proper evaluation, education, and management for clinical improvement and the prevention of complications.

Keywords: Darier-White disease, follicular keratosis

INTRODUCTION

Darier-White Disease (DWD), also known as follicular keratosis, is a rare autosomal dominant genodermatosis characterized by impaired adhesion between epidermal cells and abnormal keratinization ⁽¹⁾. Clinically, this condition manifests as greasy, hyperkeratotic papules and plaques with an unpleasant odour, predominantly affecting seborrheic areas such as the chest, back, and forehead⁽²⁾.

Globally, the prevalence of DWD is estimated to range from 1:30,000 to 1:100,000 people ⁽³⁾. In Indonesia, epidemiological data remain very limited; however, various case reports indicate that this disease is frequently misdiagnosed due to its clinical resemblance to other acantholytic disorders ⁽¹⁾. Disease onset usually begins during puberty, generally between the ages of 6 and 20 years, without significant differences by sex or ethnicity ⁽²⁾.

This disease is caused by mutations in the ATP2A2 gene, which encodes the sarco/endoplasmic reticulum Ca²⁺-ATPase (SERCA2) pump ⁽⁴⁾. Disruption of this pump causes an imbalance in calcium levels within the endoplasmic reticulum, leading to desmosomal protein malformation that triggers acantholysis and cell apoptosis ⁽²⁾. Several external risk factors can worsen clinical manifestations, including ultraviolet (UV) exposure, high heat, humidity, and mechanical friction on the skin ⁽³⁾.

Patients with DWD have a high risk of developing secondary infections, particularly from pyogenic bacteria and viruses such as Herpes simplex, which can progress to Kaposi varicelliform eruption ⁽⁴⁾. In addition to the physical burden, this disease has a substantial psychosocial impact, such as anxiety and depression resulting from visually apparent and malodorous lesions ⁽⁵⁾. Several studies have also shown an association between ATP2A2 mutations and neuropsychiatric disorders, including bipolar disorder and epilepsy ⁽³⁾.

The main management strategy focuses on symptom control and avoidance of triggering factors such as heat and sun exposure ⁽⁵⁾. First-line therapy for mild cases includes topical retinoids and corticosteroids to relieve inflammation and hyperkeratosis ⁽⁴⁾. For severe or extensive cases,

systemic retinoids such as acitretin remain the gold standard for reducing the thickness of keratotic plaques, although they require periodic monitoring for side effects ^(2,5).

This report aims to provide information regarding a case of DWD. It is hoped that this report will provide insight for clinicians in recognizing and managing similar cases so as to prevent complications.

CASE ILLUSTRATION

An 18-year-old unmarried woman presented to the Dermatovenereology and Aesthetics clinic of RSUP Dr. Sitanala with itching and red papules on the face, chest, and arms that had worsened over the 2 months prior to admission. Initial complaints began 6 years earlier, starting with papules on the hands, which then spread to the face and chest. The patient had been taken to a general practitioner's clinic and was given an ointment of unknown name. The patient denied any history of allergy or regular medication use.



Figures 1–7. Erythematous papules on the face, chest, and arms

Dermatological examination revealed multiple miliary-sized erythematous papules with diffuse borders and fine scales on the facial, thoracic, and bilateral brachial regions. A skin biopsy was performed on the right forearm (*antebrachii dextra*), and a sample measuring 0.6 x 0.4 x 0.3 cm was obtained. Histopathological results showed focal acantholytic changes and dyskeratotic cells (*corps ronds* cells) on the surface of the epidermis.

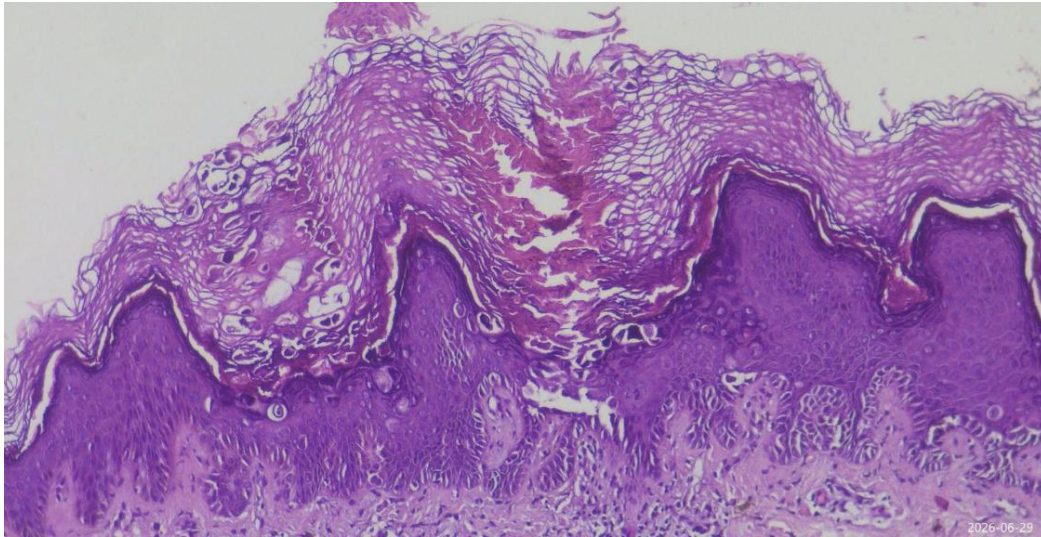


Figure 8. Histopathological appearance of the biopsy specimen

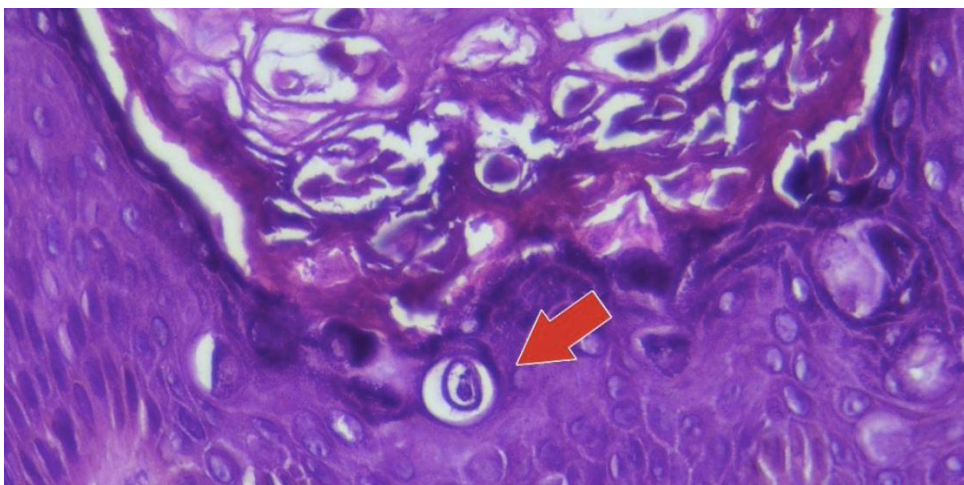


Figure 9. Corps ronds cell

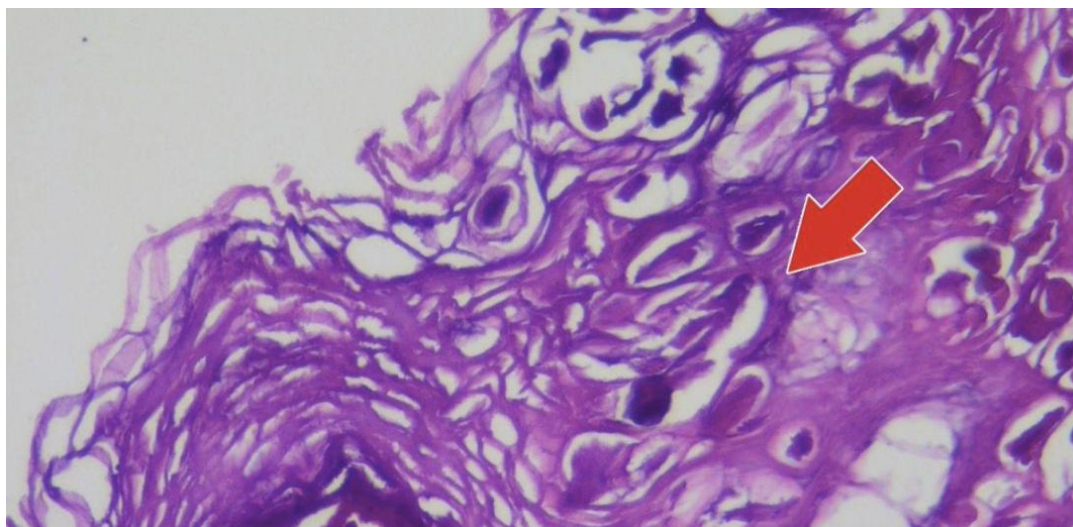


Figure 10. Grain cell

Based on the history taking, physical examination, and supporting investigations, the patient was diagnosed with Darier-White Disease. The patient was given a compounded cream of tretinoin 0.05% and Melanox ES[®], along with cetirizine 10 mg for the itching complaint.

DISCUSSION

Darier-White Disease is a rare, autosomal dominantly inherited genetic skin condition characterized by reddish to brownish papules and greasy hyperkeratotic plaques, predominantly located in seborrheic areas, usually beginning after puberty ⁽⁶⁾. Commonly affected areas include the seborrheic regions of the trunk, face, scalp margins, temples, ears, scalp, neck, and intertriginous areas such as the axillary and inguinal folds, and in women, the area beneath the breasts ⁽⁶⁾. Histopathological wise, DWD is characterized by suprabasal clefting within the epithelium, accompanied by acantholysis and the presence of dyskeratotic cells known as 'corps ronds' and 'corps grains' ⁽⁶⁾.

DWD can be diagnosed clinically, but confirmation with histopathological examination is required. Clinically, this patient presented with multiple erythematous, greasy-appearing miliary

papules with fine scales in seborrheic areas, namely the face, trunk, scalp margins, ears, neck, and arms. This finding is consistent with the clinical presentation of DWD. Histopathological examination of the patient also showed focal acantholytic changes and dyskeratotic cells (corps ronds cells) on the surface of the epidermis, consistent with the histopathological theory of DWD.

Several diseases show clinical and locational similarities to DWD, including: seborrheic dermatitis, which has the same predilection site as DWD, namely seborrheic areas; the distinguishing feature is that, clinically, it presents with an erythematous base covered by greasy, yellowish-white scales, in contrast to DWD, which presents as reddish-brown, greasy papular lesions. Acanthosis nigricans, which has lesions in flexural areas similar to DWD, but the lesions tend to be dark-coloured and hyperkeratotic with papillary eruptions, and most commonly occurs in obese patients with endocrine disorders. Additionally, there is Grover's disease, which is clinically similar to DWD, presenting as papulovesicular lesions on the trunk accompanied by itching, with a similar histopathological appearance to DWD. However, the distinguishing feature is that Grover's disease typically affects older individuals, whereas DWD more commonly affects younger individuals after puberty. In addition, Grover's disease is usually triggered by cold temperatures ⁽⁷⁾.

The goal of treatment for DWD is to improve the appearance of the skin, relieve symptoms, and prevent or treat infectious complications ⁽⁸⁾. Important management measures include maintaining hygiene to prevent secondary infection, and avoiding excessive heat, humidity, friction, and sun exposure ⁽⁶⁾. For mild cases, topical therapy such as emollients containing urea or glycerine can be given to reduce hyperkeratosis. Topical antiseptics such as chlorhexidine or octenidine, as well as topical antibiotics and antifungals, can be used to prevent or treat secondary infection. Topical corticosteroids can be given during acute inflammatory flares. For the management of severe or refractory chronic disease, oral retinoids such as acitretin and isotretinoin are the first-line treatment. Acitretin at a dose of 0.25–0.5 mg/kg/day or isotretinoin at a dose of 0.5 mg/kg/day can effectively reduce hyperkeratosis and malodour. For women of reproductive age, oral alitretinoin can be used ⁽⁶⁾. Oral retinoids have an efficacy of nearly 90% in patients with worsening DWD ⁽⁷⁾.

This patient was also given Melanox ES[®], a topical preparation containing lactic acid (3%), kojic acid, and Rumex occidentalis extract, as well as vitamin E ⁽¹⁾. According to literature, topical

alpha-hydroxy acids (AHAs) such as lactic acid act as potent keratolytic agents ⁽³⁾. This ingredient works by loosening the bonds between corneocytes to stimulate desquamation, making it effective in thinning thick hyperkeratotic plaques and smoothing the patient's skin texture ^(3,4). On the other hand, kojic acid and Rumex occidentalis extract act as tyrosinase enzyme inhibitors, working to suppress excess melanin production, thereby fading post-inflammatory hyperpigmentation (PIH) patches resulting from chronic inflammation and secondary bacterial infection that may occur in the lesion area ^(5,6).

CONCLUSION

Management of DWD is aimed at reducing the hyperkeratotic condition and minimizing the psychosocial impact on the patient. The use of sunscreen and avoidance of direct sun exposure and heat can alleviate the patient's clinical manifestations and prevent recurrence. Because DWD can cause social disability, it is important for clinicians to carry out proper evaluation, education, and management.

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