IJCRT.ORG ISSN : 2320-2882

INTERNATIONAL JOURNAL OF CREATIVE RESEARCH THOUGHTS (IJCRT)

An International Open Access, Peer-reviewed, Refereed Journal

Histopathological Diagnosis of Linear Darier's Disease in a Verrucous Epidermal Nevus–Like Lesion: A Rare Case Report.

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ABSTRACT:

A rare localized variation of Darier's disease, linear Darier's disease is characterized by aberrant keratinization and epidermal acantholysis. Clinically, it might resemble other linear dermatoses like verrucous epidermal nevus and appear with hyperkeratotic papules grouped along the lines of Blaschko, making diagnosis challenging. We describe a young woman who had several verrucous, hyperkeratotic plaques arranged in a straight line. Initially, verrucous epidermal nevus was suspected based on the clinical presentation. However, the diagnosis of linear Darier's illness was confirmed by histological analysis of the skin biopsy, which showed supranasal acantholysis and dyskeratosis with distinctive corps ronds and grains. In order to distinguish between clinically similar dermatological disorders and make an appropriate diagnosis, this case emphasizes the significance of histological investigation and clinicopathological correlation.

KEYWORDS:

Linear Darier's Disease, Verrucous Epidermal Nevus, Histopathology, Skin biopsy, Hyperkeratotic plaques, Blaschko's lines

INTRODUCTION:

A mutation in the ATP2A2 gene, which is found on chromosome 12q23-23, causes Darier's disease, a rare genodermatosis. Cutaneous mosaicism is the result of this somatic mutation, which seldom happens during the postzygotic stage of embryogenesis. The mosaic form of Darier's disease, also known as linear Darier's disease, manifests clinically as localized patterns on Blaschko's lines or linear, zosteriform.(1)

Clinical signs of Darier's disease include keratotic, crusted red-brownish papules distributed over seborrheic areas, including the face, lateral parts of the face, scalp edges, and trunk. Large verrucous plaques are typically formed when the papules unite. On the dorsal parts of their hands, about half of the patients have flat, glossy, warty papules. 15% of individuals may have painless, white papules in their mouths.(2)

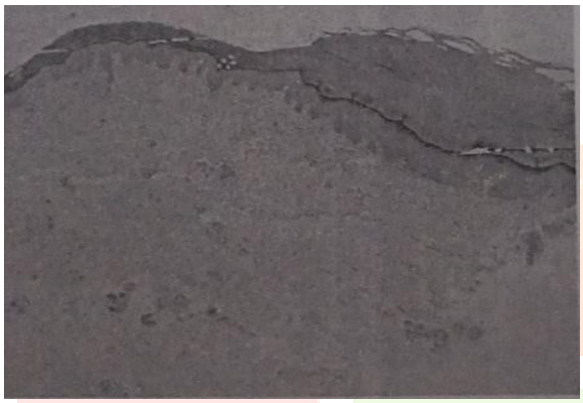
Cutaneous hamartomas with a wide range of clinical variations are known as epidermal nevi (EN). Epidermal nevi typically manifest as well-circumscribed, verrucous papillomatous lesions.(3) Any part of the epidermis, including keratinocytes, hair follicles, sebaceous glands, eccrine glands, and apocrine glands, may be affected by the lesions. The most prevalent kind of epidermal nevi is called verrucous epidermal nevus (VEN).(4)

CASE PRESENTATION

A 26 years old female patient was brought with the chief complaints of dark coloured elevated lesions present over lateral aspect of right hand over palm and right foot and few on medical aspect of left foot in the last 5 years.

The history of the current illness includes gradual development of several papular lesions. Plaques were formed when lesions came together. distribution mostly across the right foot and hand's lateral aspects. Not related to itching or scaling. long-term, gradual course. The patient's past medical history includes topical tretinoin 0.05% cream, Colchicine (which was terminated after about three weeks because of foul-smelling urine), and Lozivate-MF ointment (topical; used for about three months, then stopped). Upon local examination, we found numerous tiny, hyperpigmented papules and plaques that ranged from poorly defined to well characterized. a linear pattern of lesions. Skin has become thicker. The right foot, left elbow joint, and right palm (palmar aspect) all have them. Borders appear asymmetrical, yet the pattern is linear.

Vitamin B12 levels in the lab are 150 pg/ml, which is below normal, and the histopathology report indicates that the histological features are suggestive of callus or keratoderma. Features consistent with callus are listed in the skin biopsy report.



Report of Histopathology

The patient showed progressive clinical improvement after the stepwise multimodal therapy. After starting topical retinoids and systemic isotretinoin, there was a decrease in erythema, scaling, and hyperkeratotic papules. Lesion thickness and inflammation further improved after topical corticosteroid combinations and calcitriol-based therapy were added. The patient's disease progression stabilized with prolonged treatment of tacrolimus, emollients, and photoprotection. Colchicine used orally helped reduce lesion activity and alleviate symptoms. There was a partial remission of the lesions by the most recent follow-up in February 2026, along with improved skin texture, decreased scaling, and the absence of any new, substantial lesion development. Nonetheless, xerosis and some residual hyperpigmentation continued.

Follow-up

The patient was monitored on a monthly or biweekly basis. Lesion size, scaling, erythema, and symptom severity were all assessed clinically during follow-up appointments. Treatment changes were made in response to tolerability and response. Sun protection, consistent use of emollients, and adherence to topical medication were emphasized. Topical immunomodulators, calcitriol-based medications, and moisturizers were recommended as part of long-term maintenance therapy. The patient received counseling regarding the condition's chronicity and the necessity of routine follow-up to track any advancement or relapse.

Adverse Effects Monitoring

The patient was observed during the course of treatment for any possible side effects related to topical and systemic treatments. The patient had minor skin and lip dryness during isotretinoin medication, which was treated with daily lip moisturizers and emollients. There were no notable reports of hepatotoxicity or abnormalities in the lab. The use of topical corticosteroids was monitored to prevent skin atrophy, and

intermittent treatment was recommended. There were no significant side effects from colchicine, calcipotriol, or tacrolimus. Overall, the patient responded well to the treatment plan, and treatment was continued under close observation.

Discussion:

Mutations in the ATP2A2 gene cause Darier's disease, commonly referred to as keratosis follicularis, which results in aberrant keratinization and compromised calcium homeostasis. Greasy, hyperkeratotic papules scattered over seborrheic areas are the disease's clinical manifestation; heat, perspiration, and sun exposure can exacerbate the condition. The persistent relapsing course and inconsistent responsiveness to therapy make management difficult.(5,6)

Normalizing keratinization and decreasing follicular clogging are two functions of topical retinoids like tretinoin. Because systemic retinoids, especially isotretinoin, improve lesion shape and reduce hyperkeratosis, they are thought to be beneficial in moderate to severe disease. (7) In this case, lesion thickness and scaling significantly improved after starting isotretinoin. Tacrolimus was utilized as an immunomodulatory drug that spared steroids, while topical corticosteroids were used sporadically to manage inflammation. An analog of vitamin D called calcipotriol helped control the proliferation and development of keratinocytes. The introduction of oral colchicine as an adjuvant treatment may help lower lesion activity and inflammation.(8,9)

To preserve skin barrier function and avoid exacerbations, supportive interventions such as emollients, photoprotection, and medicated shampoos were crucial. With combined therapy, the patient showed steady progress, underscoring the significance of tailored and incremental care. Because the illness is persistent, long-term maintenance therapy is essential.(10)

Conclusion:

This case highlights Darier's disease's chronic and recurrent character as well as the necessity of customized multimodal therapy. Significant clinical improvement and disease stabilization were achieved by combination treatment using topical medications, immunomodulators, systemic retinoids, and supportive care. For the best possible illness control and enhanced quality of life, early beginning of suitable therapy, frequent follow-up, and patient counselling regarding trigger avoidance are crucial. To develop standardized treatment procedures for long-term management, more research is required.

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