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Pityriasis Versicolor-like Acquired Epidermodysplasia Verruciformis; How to distinguish: A Case Report

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Abstract

Epidermodysplasia Verruciformis (EV) is known as an uncommon inherited disease which has high genetic susceptibility to specific types of Human Papilloma Virus (HPV) infection. The clinical presentation of multiple hypopigmented scaly macules and papules may be mistaken for or difficult to distinguish from pityriasis versicolor. In this case report, we described an acquired EV associated with acquired immunodeficiency syndrome who presented with expanded pityriasis versicolor-like hypopigmented scaly macules and papules all over the body. The histological and dermoscopic examinations were compatible with EV.

Introduction

Epidermodysplasia verruciformis is a rare autosomal recessive genetic disease which associated with EVER1 and EVER2 gene mutation. It is commonly related with a high risk of skin carcinoma that results from an abnormal susceptibility to infection by specific types of Human Papillomaviruses (HPV).

We presented a rare case of acquired Epidermodysplasia verruciformis, an otherwise previously healthy 21 years old male presented with extended in numbers of generalized hypopigmented macules and papules spreading from face and neck to trunk and extremities for 1 year. Informed consent was obtained to the participant prior to the study.

Case Presentation

A 21 years old Thai male from Chonburi, came to the hospital with increase in number of hypopigmented macules and papules on face and neck which had slowly spread to trunk, abdomen and both arms during one year period (Figure 1,2). There were no other abnormal systemic symptoms. He was an otherwise previously healthy young man with no past history of any underlying diseases, neither drug nor food allergy. He also denied similar complaints or previously known genetic disease in his family members. He had never consulted a doctor for this complaint before, and hence, had not received any treatment for the skin lesions. He had a history of intravenous drug use with his friends but denied history of unsafe sexual intercourse.

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Figure 1

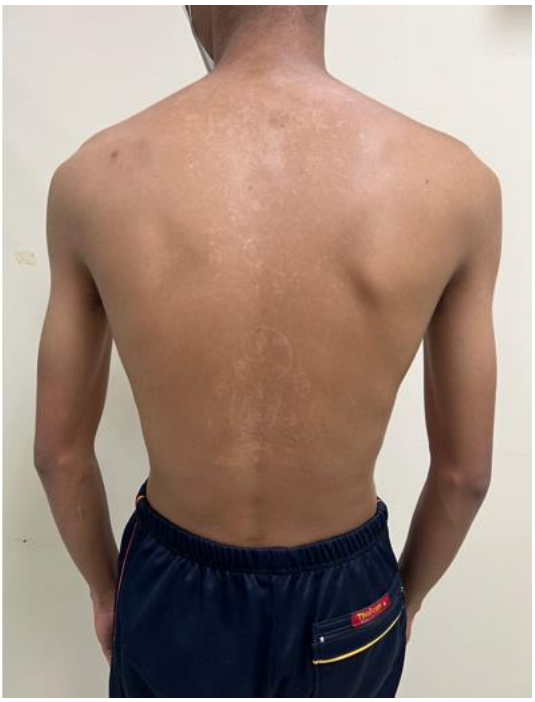

Figure 2

Physical examination revealed multiple hypopigmented macules and papules on face, neck, trunk and both arms. Some lesions appeared in linear arrangement compatible with koebner phenomenon. The hair, nails, mucous membrane and other systemic examination showed no abnormalities. Histological examination revealed focal hypergranulosis with enlarged keratinocytes with blue-gray cytoplasmic pallor without significant keratinocytic atypia, pleomorphism or increased mitotic figures (Figure 3). Dermoscopic examination showed unfocused dotted vessels on an erythematous to hypopigmented background (Figure 4,5).

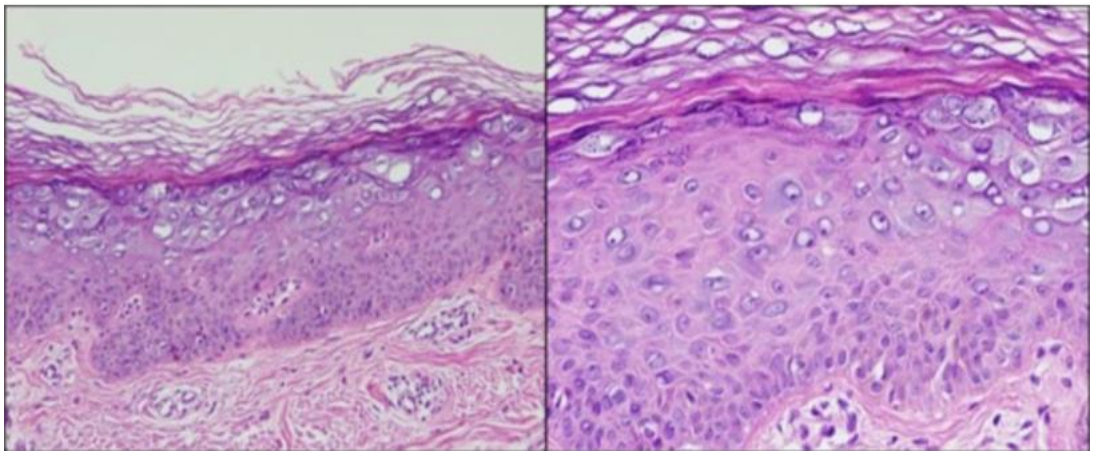

Figure 3

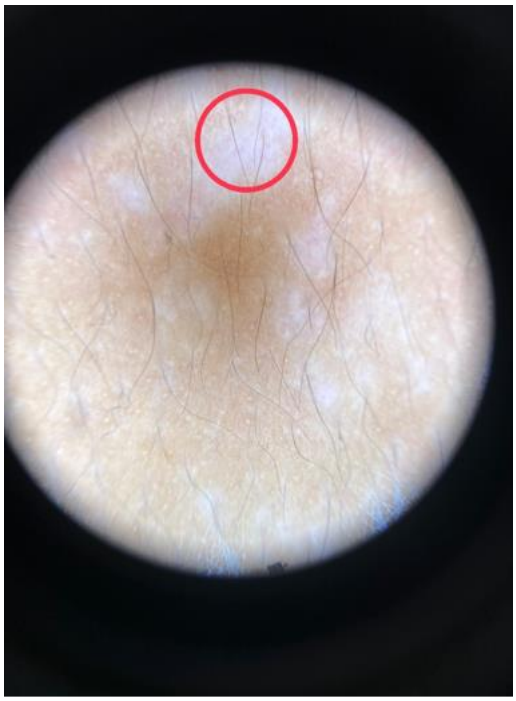

Figure 4

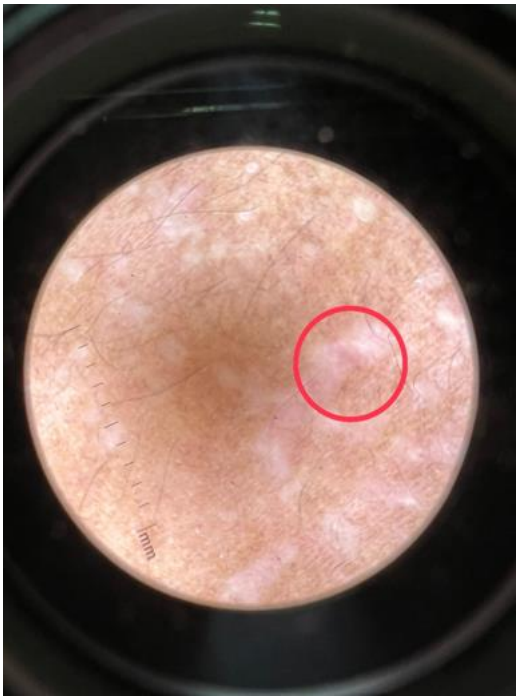

Figure 5

Laboratory tests revealed normal complete blood count, renal function test and liver function test. KOH preparation of the scales from the skin lesions was negative for fungus. On the other hand, anti-HIV test showed positive result with number of CD4 count of 349.9 cells.

After the diagnosis was made, the 50 milligrams per day of oral acitretin was prescribed, along with topical 10% urea cream. After 3 months of oral acitretin and anti-retroviral medications, there was a significant improvement of skin lesions, and, hence, acitretin was discontinued.

Discussion

Epidermodysplasia Verruciformis (EV) was first described in 1922, by Lewandowski and Lutz It is a rare autosomal recessive disorder of cell mediated immunity which increased susceptibility to specific HPV genotypes. This inherited disease is caused from the deficiency of EVER protein, result from EVER 1 and EVER 2 genes, located on chromosome 17q25, which plays an important role in maintaining zinc homeostasis which also known as the barrier of HPV.[2] Moreover, EVER genes cause defective in CD4 T-cell proliferation and increase in production of natural killer cells.[3] Even though it is claimed that Epidermodysplasia verruciformis is the result from genetic transmission but there are also acquired forms of the disease observed in immunocompromised patients such as those with HIV infection.[1] Defective CD4 function increases susceptibility to HPV infection. HPV-5 and HPV-8 are more commonly,[4] than other HPV subtypes, responsible for EV and malignant transformation of infected keratinocytes to squamous cell carcinoma which predominantly occur in sun exposed area.[5] The histopathological examination of EV patients showed hyperkeratosis, acanthosis and enlarged keratinocytes,[1] with light blue pale cytoplasm and karyopyknosis nucleus.[4]

The skin lesions of EV can be misidentified due to its similarity with pityriasis versicolor. The benign lesions are erythematous to flesh colored scaly macules or as flat wart-like papules while the malignant are verrucous

and seborrheic keratosis-like, which appeared more frequently on sun-exposed area.[1,2] The non-follicular origin and the presence of koebner phenomenon might help distinguish acquired EV from pityriasis versicolor which, caused by Malassezia furfur, usually presents with confluent follicular scaly hypopigmented or hyperpigmented macules or papules with sharp margin, occurred in various shapes on rich sebum area such as face, scalp, chest and back.[6] The dermoscopic findings of pityriasis versicolor include pigmentary network in a hyperpigmented lesion surrounded by hypopigmentation ring,[7] while the dermoscopic findings of EV reveal unfocused dotted vessels in a hypopigmented or erythematous background (Figure 4).[8]

There is no definitive treatment of EV, however the aim of EV management is to prevent malignant transformation of the benign lesions, using preventive methods including genetic counselling, photo protection combination with topical and oral medication such as oral retinoid, imiquimod, interferon and 5-fluorouracil,[2] along with close observation and monitoring for early detection of premalignant and malignant transformation. Oral retinoid, due to its anti-proliferative effect of epithelial cells,[9] and reducing progression of EV lesions to dysplasia by maintaining normal epithelial differentiation,[2] should be used in early premalignant lesion. For malignant transformation, surgical procedures such as complete excision with split-thickness or full-thickness skin grafts are the most effective treatment.[6] Preoperative chemotherapy with or without radiotherapy may decreased size of tumor before surgery. In spite of various treatment, early identification of new lesions along with strictly sun protection are crucial for improving overall survival.

Limitation

However, there are some limitations in this study such as the lack of ability to identify HPV subtype due to patient's financial problem.

Summary

EV is not only an inherited disease from autosomal recessive trait but can also be acquired in immunocompromised patients such as those infected with HIV or on immunosuppressive drugs. More susceptibility to HPV infection plays an important role in the pathogenesis and transformation of the infected premalignant cells to squamous cell carcinoma. The clinical presentation of EV could be mistaken for pityriasis versicolor due to their similarities, however dermoscopic findings might help distinguish these two diseases. Even though, there are several treatment options and preventive methods for EV, such as genetic counseling, photoprotection combined with oral and topical medication, however there is currently no definitive or curative treatment for the disease.

Statement of Ethics

Written informed consent was obtained from the patient for publication of this case report and accompanying images. This paper is exempt from ethical committee approval since it's a single case study.

This retrospective review of patient data did not require ethical approval in accordance to local guideline.

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Author Contributions

Punyawee Ongsri contributed to diagnosis/treatment of the patient, writing-review and editing the case report. Vanisha Manorattanawong contributed to chart review and writing the case report. All authors read and approved the final manuscript.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further enquiries can be directed to the corresponding author.

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