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Case Study

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CASE REPORT: ILVEN V/S LINEAR POROKERATOSIS: A DIAGNOSTIC DILEMMA

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ABSTRACT

ILVEN or inflammatory linear verrucous epidermal neavus is a rare form of epidermal naevus which manifests due to somatic mutations, reflecting genetic mosaicism. It consists of hyperplasia of the normal components of the epidermis. Porokeratosis is a group of genetic disorders characterized by clonal proliferation of keratinocytes. Linear porokeratosis is a heterogeneous disorder of keratinization which may have sporadic or AD inheritance. The following case report sheds light on how the patient was diagnosed as ILVEN on the first visit, but after HPE, it was proved that the disease was linear porokeratosis. Of all variants, linear porokeratosis is especially prone to malignant degeneration with an increased risk of transformation to squamous cell carcinoma. Thus, it is essential to diagnose and treat such cases at the earliest to prevent further complications.

KEYWORDS: Porokeratosis is a group of genetic disorders characterized by clonal proliferation of keratinocytes.

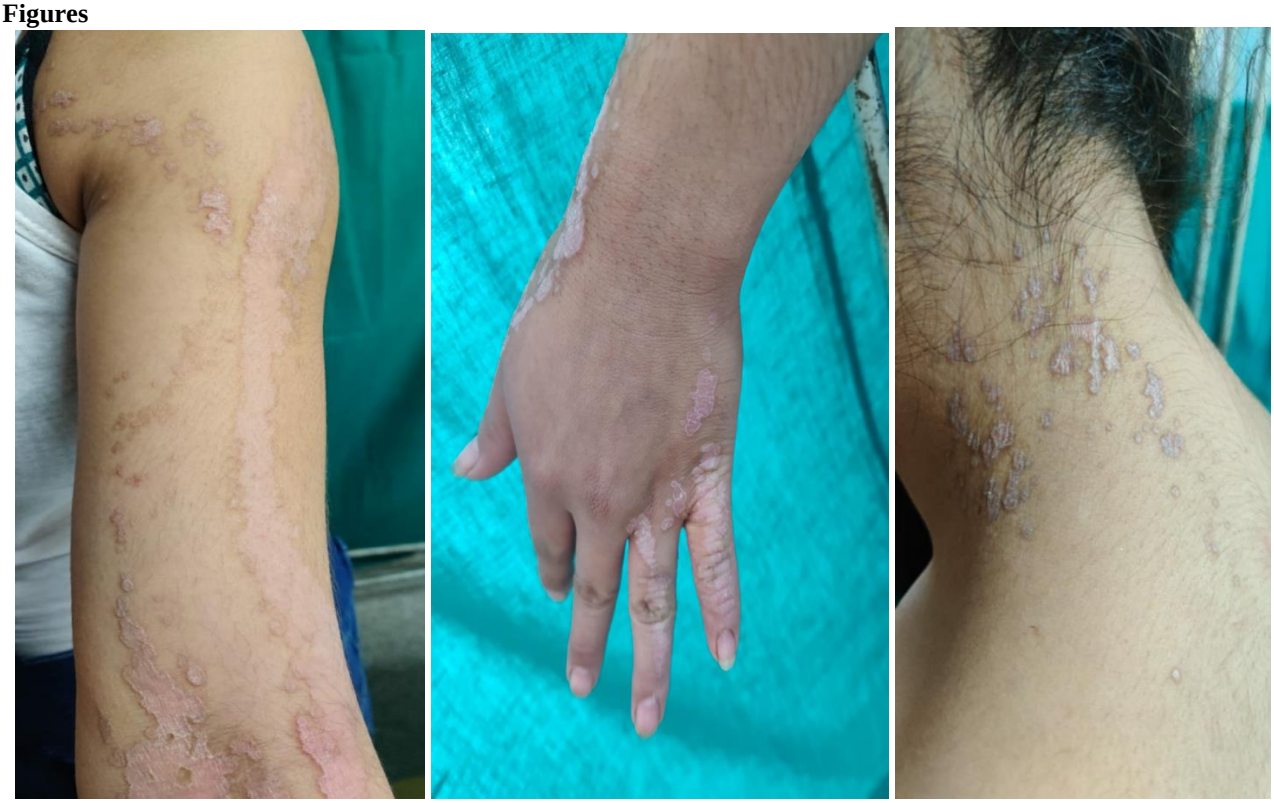
INTRODUCTION

ILVEN or inflammatory linear verrucous epidermal neavus is a rare form of epidermal naevus which manifests due to somatic mutations, reflecting genetic mosaicism. It consists of hyperplasia of the normal components of the epidermis. Its clinical features include erythematous and hyperkeratosic, warty, sometimes psoriasiform or lichenoid patches with a typical linear arrangement. The classical presentation of the same is the presence of pruritic, unilateral, erythematous scaly lesions along the Blaschko's lines.[1] Porokeratosis is a group of genetic disorders characterized by clonal proliferation of keratinocytes. Five variants have been identified to date. Linear porokeratosis is a rare clinical variant that usually arises in childhood. Linear porokeratosis is a heterogeneous disorder of keratinization which may have sporadic or AD inheritance. It is further classified as localised and generalised. It commonly appears on the extremities as unilateral, linear hyperkeratotic papules and annular plaques. Lesions may coalesce into larger plaques with central atrophy and a raised border along the lines of Blaschko.[2]

CASE REPORT

A 24 year old female presented to the dermatology OPD with the chief complaints of red raised itchy lesions on the left upper limb for the past 15 years. The lesions were insidious in onset and started from the left upper limb. They progressed to involve the neck and upper trunk over a period of 8 years after which the lesions did not progress further. The patient had a negative history for seizures, joint pains and did not demonstrate koebnerisation. The patient also had a history of multiple visits to the hospital. On examination, there was presence of multiple, well defined erythematous to violaceous papules coalescing to form plaques of size ranging from 0.3 X 0.3 cms to 5 x 3 cms with elevated margins arranged in a linear fashion extending from left nape of neck to upper limb and trunk. The lesions were non tender and non indurated. A provisional diagnosis of ILVEN and linear porokeratosis was considered based on history and clinical examination. Further, to establish diagnosis, a biopsy was done for histopathological examination. On HPE, in the epidermis, there was characteristic presence of invagination of floor which lacked granular layer and rising from its centre was a column of parakeratotic cells (cornoid lamella). The dermis contained moderately dense superficial perivascular patchy lichenoid lymphocytic infiltrate with focal interface vacuolar change. The diagnosis of linear porokeratosis was established and the patient was started on appropriate therapy in the form of topical 5-flourouracil. A significant reduction was seen in the lesions over a period of 8 weeks.

Figures



Figures 1-3: multiple, well defined erythematous to violaceous papules coalescing to form plaques of size ranging from 0.3 X 0.3 cms to 5 x 3 cms with elevated margins arranged in a linear fashion extending from left nape of neck to upper limb and trunk.

DISCUSSION

The resemblance between clinical presentations of various dermatological diseases poses a difficulty in diagnosis thus, delaying the treatment. It is important to establish a diagnosis timely with the help of necessary investigations. The above case report sheds light on how the patient was diagnosed as ILVEN on the first visit, but after HPE, it was proved that the disease was linear porokeratosis. Of all variants, linear porokeratosis is especially prone to malignant degeneration with an increased risk of transformation to squamous cell carcinoma.[3] Thus, it is essential to diagnose and treat such cases at the earliest to prevent further complications.

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