

Extensive linear verrucous epidermal nevus involving the right upper limb and palm

Keywords: asymptomatic hyperkeratotic, hyper pigmented, warty plaques, verrucous plaques, abnormalities seen, Histopathological diagnosis, epidermal nevus, Microscopic picture, papillomatosis, acanthosis, Pseudohorn cysts, psoriasis, Epidermal nevi, Neutrophilic pustulation

Abbreviations: ILVEN, Inflammatory linear verrucous epidermal nevus; LVEN, linear verrucous epidermal nevus

Case presentation

A 13 year old female, born of non-consanguineous marriage, presented with asymptomatic hyperkeratotic, hyper pigmented warty plaques distributed in a linear fashion over the right side of the body. The patient's mother complained that these lesions were present since infancy. They were stable for a long period of time, but had progressed in size to involve the whole of right upper extremity over the past 3 years. On physical examination, patient had well-defined, hyper pigmented verrucous plaques in a linear blaschkoid distribution extending from just above the supramammary area, shoulder, right arm and forearm extending in a contiguous fashion to involve the thinner eminence of right palm and till the tip of index finger with some areas of intervening normal skin. There was no evidence of any other skin lesions and hair, nail, and mucosae were normal (Figure 1). The child was developmentally normal and there were no gross abnormalities seen Figure 2. A detailed systemic examination revealed no abnormality. Ophthalmologic, neurological and orthopaedic specialist consultations were sought to rule out an epidermal nevus syndrome and there was no abnormality detected in any of the systems. Microscopic picture of the biopsy showed an epidermal proliferative lesion (Figure 3). There was hyperkeratosis, papillomatosis, acanthosis and increased basal layer pigmentation. Dermal collagen and a few telangiectatic vessels were seen within the papillary projections along with mild inflammatory infiltrate. Pseudohorn cysts were also seen. Histopathological diagnosis was consistent with linear verrucous epidermal nevus (LVEN). Inflammatory linear verrucous epidermal nevus (ILVEN) shows a characteristic histological feature of regular alteration of slightly raised parakeratotic areas without a granular layer and a slightly depressed orthokeratotic area with prominent granular layer and hence was ruled out.^{1,2} LVEN resembles seborrheic keratosis which generally lacks the inflammatory changes and characteristic changes in the stratum corneum seen in LVEN.

In psoriasis, distinction can be difficult without clinical correlation. Neutrophilic pustulation is generally not a feature of LVEN. Epidermal nevi represent Blaschkoid hamartomas of the skin that result from mosaic post-zygotic mutations.³ LVEN are the most common amongst these epidermal nevi that include nevus comedonicus, ILVEN and nevus sebaceous.⁴ LVEN can exist as single or multiple lesions, may be of any size and may occur on any site.⁵ Clinically, they appear as brownish to skin coloured, well demarcated, verrucous papules that coalesce into plaques. The majority of epidermal nevi are either present at birth or occur within the first year of life.⁴ LVEN pose a therapeutic challenge because superficial destruction may result in recurrence, whereas deeper modalities are limited by the extent of affected areas

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and a higher risk of post-procedural scars.^{6,7} Various light-based treatments, which include Argon,⁸ PDT,⁹ ruby laser,¹⁰ Erbium:YAG,¹¹ and CO₂ laser⁸⁻¹⁴ have been reported to be variably effective in LVEN. Extent of lesional hyperkeratosis may be a predictor of response to laser therapy. For hard or keratotic LVEN, CO₂ laser was shown to be more effective than the Argon laser.⁸ In a study of 25 patients with epidermal nevi (24 patients with LVEN), CO₂ laser vaporization led to complete or almost complete resolution in 92% of patients with soft flattened nevi but only in 33% of patients with keratotic nevi. The majority of recurrent lesions occurred in patients with keratotic nevi. Of those treated, 12.5% had hypertrophic scars.¹⁴



Figure 1 There was no evidence of any other skin lesions and hair, nail, and mucosae were normal.



Figure 2 Shows involvement of the entire arm and chest and the extent of involvement.

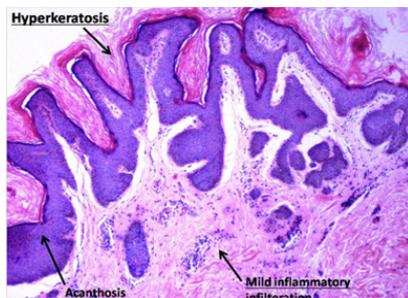


Figure 3 Microscopic picture of the biopsy showed an epidermal proliferative lesion.

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Conflict of interest

The authors declare there is no conflict of interest.

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