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CASE REPORT **OPEN ACCESS**

Inflammatory linear verrucous epidermal nevus (ILVEN) with limb atrophy and nail involvement

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ABSTRACT
Report of a sporadic case of inflammatory linear verrucous epidermal nevus (ILVEN) in a linear blaschkoid pattern from left axilla to tip of ring finger including the nail bed with hemiatrophy of left forearm.
***Key words**: Inflammatory linear verrucous epidermal nevus (ILVEN), Hemiatrophy, Nail involvement.*

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INTRODUCTION

Epidermal nevi are the benign hamartomas of skin derived from ectoderm[1]. They usually present at birth or appear in early childhood, in a distinctive pattern[1].The prevalence of epidermal nevi is one in 1000 live birth with female preponderance [1].Inflammatory linear verrucous epidermal nevus (ILVEN) is one of the rare form of epidermal nevus. They are not hereditary but arise due to mosaicism resulting from postzygotic mutation [2]. Clinically appear as unilateral, linear erythematous, verrucous, pruritic, papules and plaque following lines of Blaschko[3]. ILVEN mostly present as an isolated cutaneous lesion but extracutaneous manifestations can occur involving brain, eye and skeletal system [3].

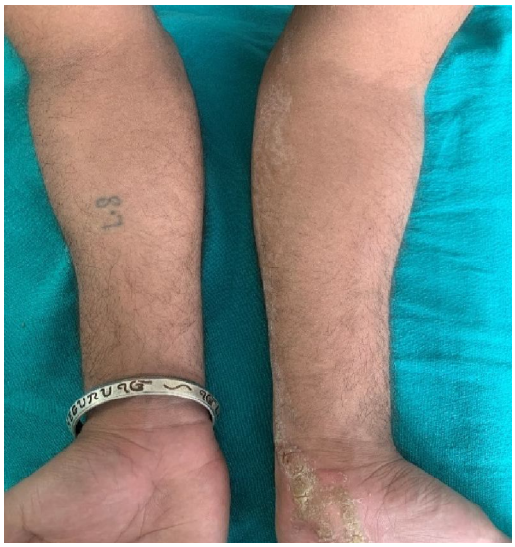
CASE REPORT

A 26 years old male presented with pruritic, hyperpigmented, warty plaque in a linear distribution present on left side of body. The lesions were present since infancy. It initially presented as hyperpigmented papules, which gradually increased in size and number in a linear fashion becoming more thickened and verrucous. Physical examination revealed well defined hyperpigmented, hyperkeratotic, scaly, crusted, verrucous plaque, 2-3 cm in diameter and in a linear blaschkoid pattern extending from left axilla to tip of the ring finger involving the nail bed with nail dystrophy [Figure 1, 2]. No history of similar lesion in the family. No ophthalmologic, neurological and other congenital abnormality was present. There was also atrophy of muscle of left forearm 2 cm less than right forearm at the level of mid forearm (7cm below the olecranon process) [Figure 1]. Clinically ILVEN and linear psoriasis lie in the differential diagnosis. Skin biopsy from the lesion showed hyperkeratosis with foci of parakeratosis, moderate acanthosis, papillomatosis, elongation of rete ridges with patchy exocytosis of lymphocytes and perivascular mononuclear inflammatory infiltrate. Histopathological diagnosis consistent with inflammatory linear verrucous epidermal nevus. X-ray forearm AP view showed no underlying bony abnormality.

DISCUSSION

Inflammatory linear verrucous epidermal nevus is a rare variant of congenital epidermal nevus presented as a linear, erythematous, eczematous or psoriasiform papules and plaque in blaschkoid pattern which are intensely pruritic and do not cross the mid line [3]. The lesions are present mostly on buttock and lower limbs and appear within 5 years of age [3]. Most cases are sporadic but few familial cases have been observed [3]. Our case was sporadic in nature. Pathophysiology of ILVEN is poorly understood but it is thought to be due to somatic mosaicism [1]. ILVEN can clinically and histologically mimics linear psoriasis [4]. As was also seen in our case. Association of developmental defect of skin with involvement of eyes, CNS, skeletal, renal and CVS constitute epidermal nevus syndrome [6].Limb atrophy with underlying muscular atrophy without any bony involvement was seen in our case.

Landwehr and Starink [5] report a case of inflammatory linear verrucous epidermal naevus widespread with bilateral distribution with nail involvement as was seen in our case. Baptista and Cortesao [6] reported a case of two sisters with left hemiatrophy with bilateral verrucous nevus with spontaneous resolution. Sawhney [7] reported a case of crossed systematized epidermal nevus with crossed hemihypertrophy. We report a case of extensive inflammatory linear verrucous epidermal nevus with nail involvement and hemiatrophy of left forearm in a 26 year old male.



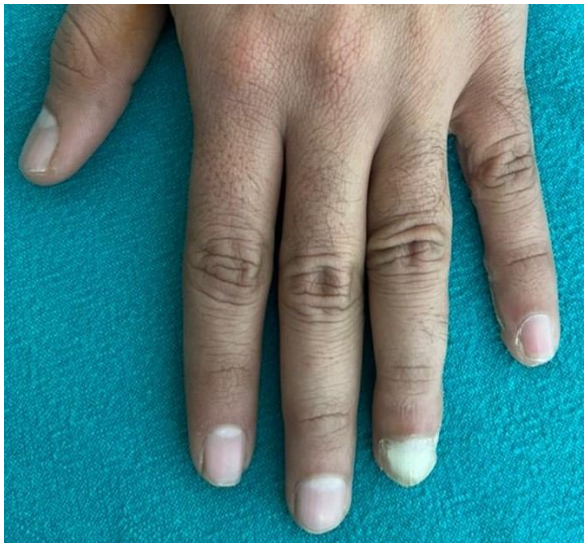


Fig. 1. Inflammatory linear verrucous epidermal nevus involving left hand and left upper limb with atrophy of underlying muscles.
Fig. 2. Nail bed involvement of ring finger with nail dystrophy in a case of ILVEN

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