

Bilateral Inflammatory Verrucous Epidermal Nevus in Association with Unilateral Lobster Claw Hand Deformity: An Unusual Association

Sir,

Inflammatory linear verrucous epidermal nevus (ILVEN) is a type of epidermal nevus characterized by linear, pruritic, erythematous, and hyperkeratotic papules that often coalesce into plaques, with a raised scaly surface, which may be unilateral or bilateral and usually follows the lines of Blaschko. The age of onset may vary from birth to early infancy. Histology shows psoriasiform hyperplasia with hyperkeratosis, parakeratosis, alternating hypergranulosis, and hypogranulosis. The affected patients usually seem normal but sometimes there may be associated musculoskeletal or neurological abnormalities prompting the classification of ILVEN as a part of epidermal nevus syndrome (ENS). We hereby report a case of a young girl with bilateral ILVEN with a co-existent lobster claw hand on the right side.

A 7-year-old girl born of non-consanguineous parentage presented to us with multiple linear streaks of itchy elevated lesions over the right and left upper limb, present since infancy. To start with, there were a few pea-sized itchy erythematous lesions in the same distribution, which gradually increased in number and morphology to attain the present status. There was no history of bullous lesions prior to the development of the keratotic plaques. She had sought treatment for this condition from multiple physicians. Unfortunately, the lesions were refractory to treatment. The family history was unremarkable. Cutaneous examination revealed multiple streaks of verrucous papules coalescing to form plaques in a Blaschkoid pattern, distributed over the upper extremity of both sides, on the right side the lesions were extending from the elbow to the tip of the fingers, whereas the lesions were present only over the dorsum of the middle finger on the left side [Figure 1]. There were no similar lesions elsewhere in the body. Hairs, nails, and mucosae were normal. Besides, there was absence of the index finger and middle finger

of the right hand, which was diagnosed as lobster claw hand [Figure 2]. There was no history suggestive of any neurological deficit. Her dentitions were also normal. There was no history of any infection or teratogenic drug usage by her mother during pregnancy. Histopathological examination from the verrucous plaque showed psoriasiform epidermal hyperplasia with hyperkeratosis, parakeratosis, alternating hypergranulosis and hypogranulosis, acanthosis, widened rete ridges, superficial perivascular infiltrate of lymphocytes and neutrophils, with focal extension towards the epidermis [Figure 3]. Based on the history, clinical features, and histology, a diagnosis of the inflammatory verrucous epidermal nevus with lobster claw hand was done. ILVEN is clinically manifested with pruritic erythematous and verrucous papules and plaques, in a linear distribution over the extremities, with infants to children being the most affected group. Altman and Mehregan described six characteristic features of ILVEN: (1) early age of onset, (2) predominance in females (4:1 female-male ratio), (3) frequent involvement of the left leg, (4) pruritus, (5) marked refractoriness to therapy, and (6) a distinctive psoriasiform and inflammatory histologic appearance, and our case had almost all the characteristics.^[1] Rarely, ILVEN has been found to co-exist with musculoskeletal abnormalities, like supernumerary digits, congenital bony anomalies of the ipsilateral extremities, and congenital dislocation of the ipsilateral hip.^[2] In our case, it was associated with lobster claw hand which is quite unusual. Other rare associations of inflammatory verrucous epidermal nevus include arthritis, basal cell carcinoma,^[3] porokeratotic eccrine ostial and dermal duct nevus,^[4] neurofibromatosis,^[5] lichen amyloidosis,^[6] and woolly hair nevus.



Figure 1: Multiple streaks of verrucous papules coalescing to form plaques in a Blaschkoid pattern bilaterally distributed over the extremities



Figure 2: Lobster claw hand deformity on the right side

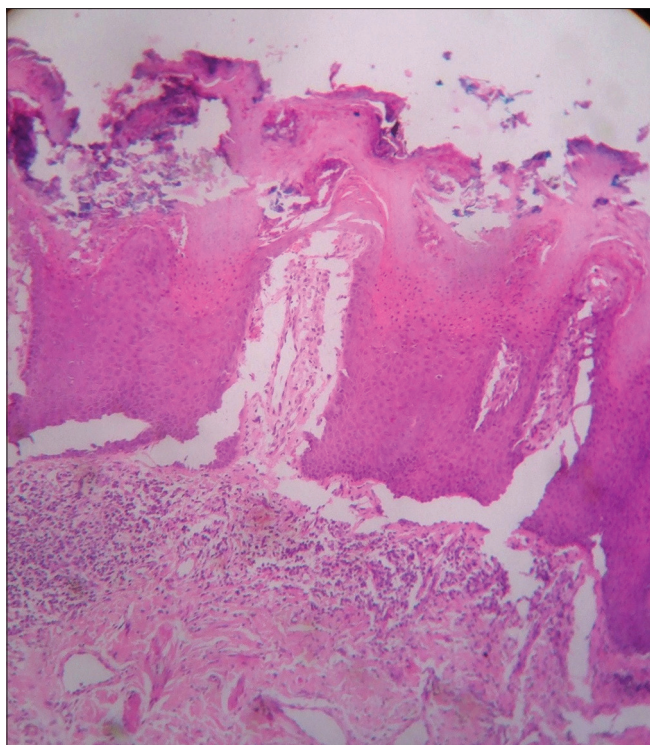


Figure 3: Photomicrograph showing hyperkeratosis, parakeratosis, alternating hypergranulosis and hypogranulosis, acanthosis, widened rete ridges, superficial perivascular infiltrate of lymphocytes, and neutrophils with focal extension towards the epidermis (H & E, 100X)

Lobster claw hand, a rare congenital anomaly, also known as ectrodactyly or split hand foot malformation refers to central deficiencies of the hand, developing as a result of the longitudinal failure of the formation of 2nd, 3rd, or 4th ray.^[7] There are two expressions of ectrodactyly, one is the non-syndromic form with isolated limb involvement and the other is the syndromic form which is associated with other anomalies like tibial aplasia, mental retardation, orofacial clefting, enamel hypoplasia, learning abnormalities, renal abnormalities, and deafness.^[8] In our case, no other associated congenital anomaly was detected. The closest clinical differentials of ILVEN include linear psoriasis and lichen striatus. The condition is difficult to treat, options being topical and intralesional glucocorticoids, excision, cryotherapy, and laser therapy (carbon dioxide laser and flash lamp-pumped pulsed-dye laser).

ILVEN in association with unilateral lobster claw hand is quite unusual, which prompted the present report.

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Conflicts of interest

There are no conflicts of interest.

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