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SYSTEMATIZED INFLAMMATORY LINEAR VERRUCOUS EPIDERMAL NEVUS WITH EPIDERMOLYTIC HYPERKERATOSIS- A RARE CASE REPORT

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Abstract

The inflammatory linear verrucous epidermal nevus (ILVEN) is a rare, keratinocytic type of epidermal nevus characterized by erythematous, hyperkeratotic, warty, psoriasiform or lichenoid patches with a typical linear arrangement along the lines of blaschko. This condition typically affects neonates, but may occur in children older than 5 years of age and rarely in adults. Females are commonly affected than males. We report a rare case of inflammatory linear verrucous epidermal nevus in a 13-year-old male who presented to the department of dermatology with complaints of bilaterally symmetrical dark and raised lesions all over the body. Routine blood tests showed no signs of systemic involvement. Histopathology showed Epidermolytic hyperkeratosis, acanthosis, papillomatosis with perivascular lymphocytic infiltrate. Epidermolytic hyperkeratosis (EHK) in histopathology may indicate the risk of bullous icthyosiform erythroderma (BIE) in the offspring necessitating routine biopsy and histopathological examination of all epidermal nevi.

Keywords: Inflammatory Linear Verrucous Epidermal Nevus, Epidermolytic Hyperkeratosis, Acanthosis, Papillomatosis.

INTRODUCTION

Inflammatory linear verrucous epidermal nevus (ILVEN) is a rare keratinocytic type of epidermal nevus characterized by pruritic erythematous scaly plaques arranged in a linear configuration, along the lines of Blaschko (1). Children and females are more commonly affected (2). Epidermal nevi are hamartomatous growths which occur due to hyperplasia of epidermis or adnexa (3). The various subtypes include verrucous epidermal nevus, nevus sebaceous, nevus comedonicus, eccrine nevus and apocrine nevus (3). Epidermal nevi may be associated with developmental abnormalities of skin, eyes, nervous system, skeletal, urogenital and cardiovascular systems known as 'epidermal nevus syndromes' (4). The various syndromes include keratitis, erythrokeratoderma and sensorineural deafness (KID syndrome), congenital hemidysplasia, ichthyosiform erythroderma and limb defects (CHILD syndrome) and Gardner's syndrome (5). Hence, we report a rare case of inflammatory linear verrucous epidermal nevus (ILVEN) in a 13- year-old male who presented to the department of dermatology with complaints of bilaterally symmetrical dark and raised lesions all over the body.

ILVEN constitutes 6% of the epidermal nevi (6). The clinical criteria for ILVEN was first described in 1971 by Altman and Mehregan and later modified by Morag and Metzker in 1985 (6). It is refractive to treatment (6).It occurs due to postzygotic mutation by genetic mosaicism (7).

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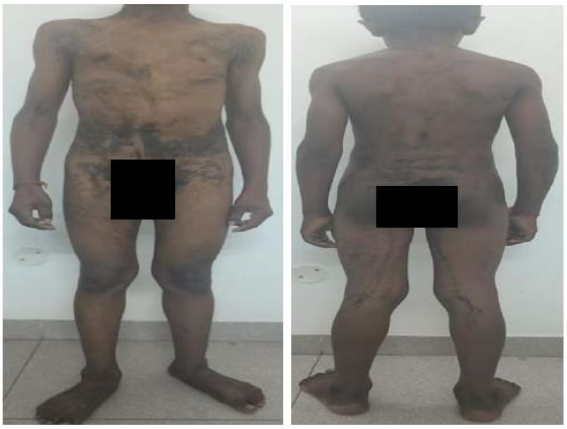
Case Report:

History – A 13 year old male who presented to the department of dermatology with complaints of bilaterally symmetrical dark and raised lesions all over the body. The parents initially noticed dark coloured skin lesions on the left forearm at 45 days of birth which gradually increased in size within 2 years of age. History of second-degree consanguinity and mild speech delay was present.

Physical Examination - On examination, bilateral symmetrical multiple hyperpigmented macules and hyperkeratotic papules and plaques arranged in streaks and whorls was noted on the face, chest, abdomen, upper and lower limbs with involvement of palms and soles. There were no other systemic abnormalities.



Figures A AND B- showing bilateral symmetrical multiple hyperpigmented macules and hyperkeratotic papules and plaques arranged in streaks and whorls

Laboratory data - Routine blood tests showed no signs of systemic involvement. **Histopathology** – Epidermis showed Epidermolytic hyperkeratosis, acanthosis, papillomatosis with perivascular lymphocytic infiltrate in the papillary dermis.

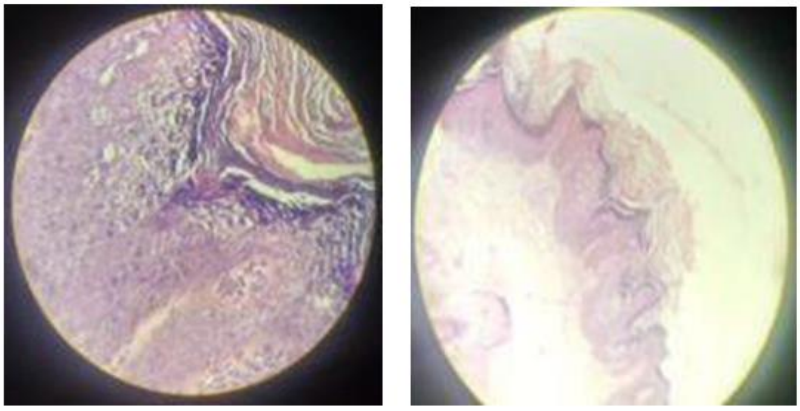


Figure C, D-showing Epidermolytic hyperkeratosis, acanthosis, papillomatosis with perivascular lymphocytic infiltrate in the papillary dermis

Dermoscopic examination - brownish scales with brown glomerular type of structures over a white background were seen.

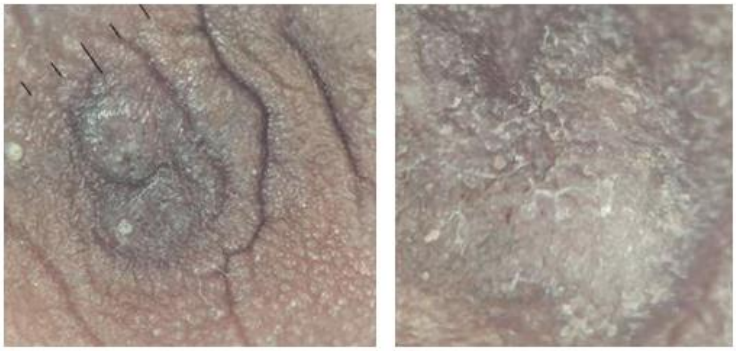


Figure E, F-showing brownish scales with brown glomerular type of structures over a white background

Treatment – He was treated with topical retinoids and topical corticosteroids. Follow up after 3 months showed mild improvement.

DISCUSSION

In 1896, Unna first described the inflammatory linear verrucous epidermal nevus (ILVEN) as congenital malformations or hamartomas derived from embryonic ectoderm (8).Incidence of ILVEN is 1-3/1000 live births (8).

Sucheta Bansal et al (8) reported a case of ILVEN in a young female who presented with unilateral pruritic, erythematous and verrucous papules, in a linear distribution following Blaschko's lines localised to the left side of the body.

Sanjay singh et al (9) reported a case of Systematized Inflammatory Linear Verrucous Epidermal Nevus in a 4-year-old female child who presented with generalized multiple hyperpigmented hyperkeratotic papules and plaques in blaschkoid distribution.

We present a case of 14-year male patient with complaints of bilaterally symmetrical dark and raised lesions all over the body. On examination, the lesions are bilaterally symmetrical and multiple hyperpigmented macules and hyperkeratotic papules and plaques arranged in streaks and whorls was noted on the face, chest, abdomen, upper and lower limbs with involvement of palms and soles. Routine blood tests showed no signs of systemic involvement.

ILVEN is clinically diagnosed and supported by medical history and histological examination. The histopathological examination is characterised by epidermal hyperplasia of normal components, with acanthosis, papillomatosis, hyperkeratosis and parakeratosis. In papillary dermis, inflammatory reactions either diffuse or perivascular (10).

Similarly, in our case, the histological examination showed epidermolytic hyperkeratosis, acanthosis, papillomatosis with perivascular lymphocytic infiltrate.

There is no specific and effective treatment for this condition available. But there may be a wide range of treatment options includes topical corticosteroids, dithranol and retinoids, topical vitamin D analogues, 5-FU and calcineurin inhibitors (11). Other treatment modalities like surgical excision, laser therapy and cryotherapy are used by some dermatologists (11).

CONCLUSION

Epidermal nevus may be associated with developmental abnormalities of skin, eyes, nervous system, skeletal, urogenital and cardiovascular system. A thorough systemic screening is necessary with specific investigations depending on the involved system. If histopathology shows EHK, counselling is necessary regarding risk of developing BIE in offspring.

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