Case 14
A 9 year-old Thai boy from Pathumtani

Chief complaint: brownish warty skin lesion starting from the angle of right side of the mouth and extending to the right cheek, neck and back which was present since birth

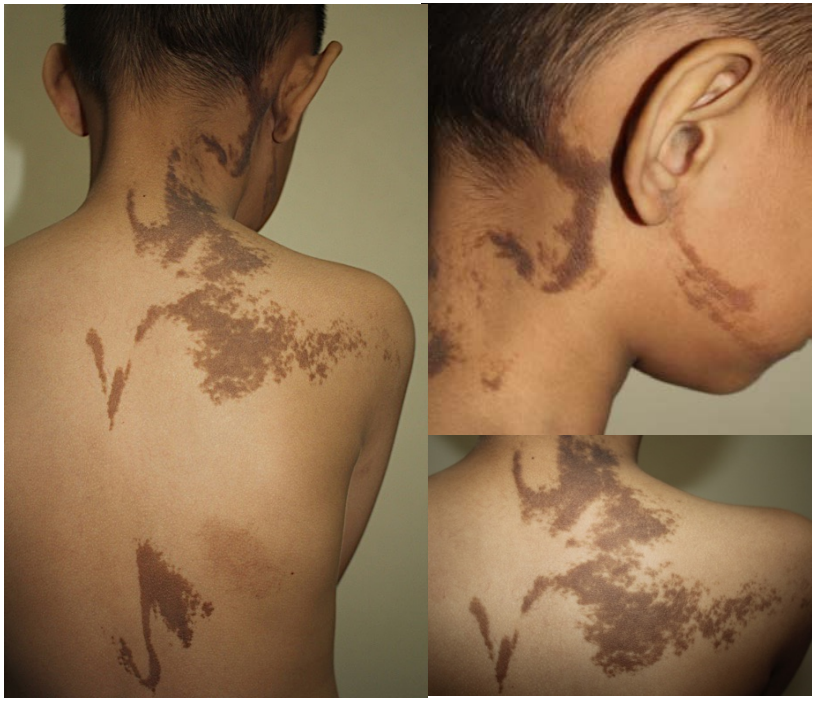

Fig. 14.1

Present illness:
The skin lesions became darker and progressed gradually with age. It was not associated with any itching and he had no history of seizures or difficulty learning, hearing or visual problems.

Past history: no prenatal complications and normal developmental milestones

Underlying disease: no other underlying diseases

Family history: no family history of similar condition

Physical examination:
Other systemic examination revealed no abnormality

Dermatologic examination: (Fig.14.1)

- Multiple, brownish verrucous papules coalescing to form well-demarcated streaks and plaques starting from the angle of the right side of mouth and extending to right cheek, neck and back in a blaschkoid pattern
- Hair, nails and mucosa were normal

Investigations:
No further investigations were done

Histopathology: (S18-005564A, skin, back)

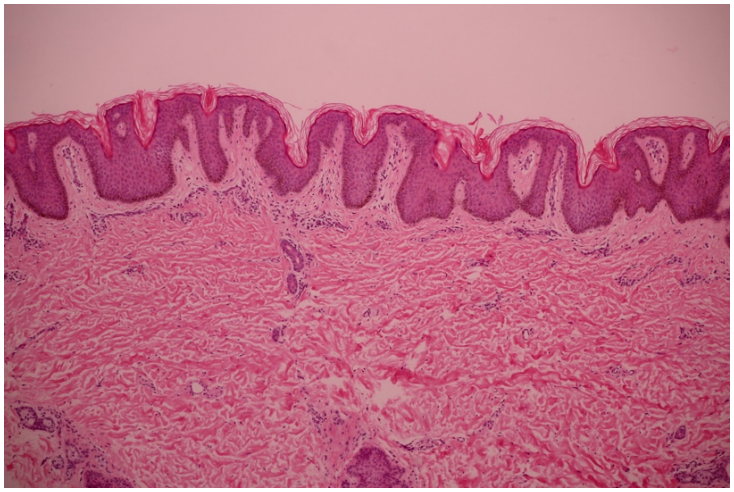

Fig. 14.2

- Mild hyperkeratosis, papillomatosis and epidermal hyperplasia
- Sparse inflammatory cells infiltrate composed of lymphocytes in the papillary dermis

Diagnosis: Linear verrucous epidermal nevus

Treatment:

- CO_2 laser for skin lesions
- Close follow to monitor for late onset systemic involvement

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Discussion:

Epidermal nevi are organoid nevi arising from the embryonic ectoderm. These cells give rise not only to keratinocytes but also to skin appendages. They are often classified according to the predominant component they originate from e.g. nevus verrucosus (keratinocytes), nevus sebaceous (sebaceous glands), nevus comedonicus (hair follicles) and nevus syringocystadenosus papilliferus (aprocrine glands).[1] However, according to Solomon and Esterly, only few lesions are exclusively of one type. The predominant tissue varies with evolution of the lesions and different areas of the same lesion shows a variety of components at the same time.[2] The most common type of epidermal naevi is the keratinocytic naevi, also called verrucous epidermal naevi.[3] Verrucous epidermal nevus (VEN) is a congenital, non-inflammatory nevus formed by keratinocytes.[4] They are usually present at birth or infancy and may enlarge slowly during childhood as seen in our case. The naevus typically appears as verrucous papules that coalesce to form well-demarcated, skin-colored, brownish or grey-brown, papillomatous plaques[3] or as localized linear lesions distributed along the lines of Blashko.[5]

Based on the extent, distribution and presence or absence of inflammation, VEN can be divided into different subtypes: localized VEN, nevus unius lateralis, systematized VEN or ichthyosis hystrix and inflammatory linear verrucous epidermal nevus (ILVEN).[4] They are said to be systematized when they occur as more than one linear lesion according to Lever's classification of epidermal naevi.[6] In our case, a 9 year old boy presented with naevus unius lateralis which usually presents with unilateral or nearly unilateral verrucous lesions arranged in linear streaks or bands. The first photo

documented case of linear verrucous nevus involving half of the body was in 1915, published by Corbet.[7]

The exact mechanism underlying the development of epidermal naevi is not yet understood clearly but it appears that the disease is a mosaic disorder resulting from a post-zygotic mutation.[8] It has been hypothesized that the lesions with features of epidermolytic hyperkeratosis may indicate a mosaic disorder of suprabasal keratin gene mutation, in particular, the keratin 1 or keratin 10 genes. These patients may transmit the mutation to an offspring and it is more likely to occur if the epidermal nevus is extensive.[9]

Histopathologically, at least 10 different patterns of epidermal nevi have been described, with any lesion possibly revealing more than one histological pattern. Common epidermal changes include hyperkeratosis, papillomatosis, hypergranulosis, and acanthosis[9] which is similar to our case. Depending on histology, it can be classified into two groups: epidermolytic and non-epidermolytic. Perinuclear vacuolization of keratinocytes and premature excessive formation of kerato-hyaline granules characterize epidermolytic VEN[4] unlike our case. Non-epidermolytic verrucous epidermal naevi is usually present at birth. Extra-cutaneous features and malignant degeneration has been reported to occur in non-epidermolytic VEN.[4,5] Hence, there is a need to differentiate between the two variants.

Epidermal nevi can be a part of the epidermal nevus syndrome, which is a combination of epidermal nevi with abnormalities in other organs, commonly the central nervous system, the eyes, and the musculoskeletal system[9] but there are isolated examples of involvement cardiovascular and renal systems.[2,10] In our case, none of these associations were observed even though the skin lesions were quite extensive. Only a few cases of extensive verrucous epidermal naevus with no involvement of other organs similar to our case has been reported in the literature.[4–6,11] A thorough examination and observation for the entire lifetime of patient is necessary as sometimes neurological manifestations may present late in adulthood.[5,11]

Treatment of epidermal nevi is challenging. Destructive therapies like surgical excision, cryotherapy, carbon dioxide laser are the mainstay of treatment but surgical excision of widespread lesion is not feasible. Topical therapies that have been used include retinoids,5-fluorouracil,steroids,podophyllin,vitaminD analogues [11,12] and topical calcipotriol/betamethasone dipropionate combination.[9] The successful use of oral retinoids has also been described.[9] Various studies have reported good clinical result with CO_2 laser in treatment of epidermal naevi.[13,14] Our patient, is also undergoing CO_2 laser therapy and is being followed up regularly to monitor for late onset systemic involvement.

Reference:

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