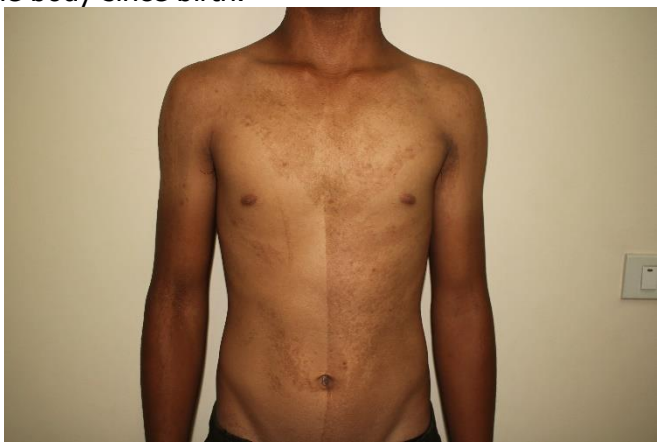
Case 20

A 14-year-old Thai boy from Kalasin province

Chief complaint: Multiple hyperkeratotic brownish plaques all over the body since birth.



Present illness: The patient developed multiple hyperkeratotic hyperpigmented papules coalesce to plaques on face, neck, trunk (predominant on left side) and extremities since birth. His lesions have gradually increased in size and thickness with age. The growth and developmental history were normal. There was no history of seizures, learning difficulty, hearing difficulty or altered vision. There was no other systemic symptoms.

Past history : No underlying disease

Family history : Nil

Physical examination

GA: alert, well co-operated Thai boy

HEENT: normal oral mucosa

Hair and Nails: normal appearance

Neurological system: no abnormality detected

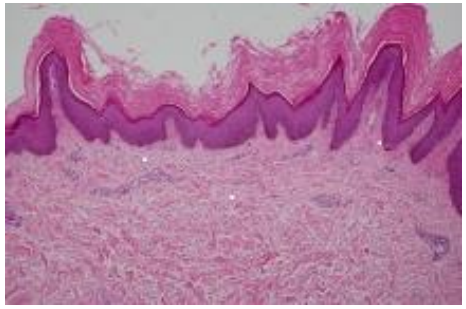
Skeletal system: no abnormality detected

Skin examination

Multiple hyperkeratotic brownish papules coalesce to plaques extensively involving face, both sides of trunk, extremities and knuckles of both hands. The distribution is along the lines of Blaschko. There was a sharp midline demarcation on his trunk.

Histopathology (S15-11264A, Left knee)

- Compact hyperkeratosis
- Epidermal hyperplasia, papillomatosis with "church-spire" pattern



Diagnosis: Systematized epidermal nevus

Treatment:

10% urea cream apply twice daily all over body

3 % lactic acid cream apply once daily on face

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Discussion:

Epidermal nevus are harmatomatous proliferations of the epithelium, divided into five subtypes according to the distribution and the predominant histologic cell type: keratinocyte (verrucous epidermal nevus), sebaceous gland (nevus sebaceous), pilosebaseous unit (nevus comedonicus), eccrine gland (eccrine nevus, and apocrine gland (apocrine nevus)

Systematized epidermal nevus are defined as the extensive disease of verrucous epidermal nevus, which are

congenital non inflammatory cutaneous harmatomas composed of keratinocytes. The age of onset usually at birth or within the first year of life, however late onset lesions have also been reported.[1]

Presentation of systematized epidermal nevus are characterized by multiple well-circumscribed, hyperpigmented, papillomatous papules or plaques with extensive distribution, commonly transverse configuration on the trunk and linear configuration on the limbs. The common sites are on the trunk, extremities or neck.[2]

The lesions can be limited on one half of the body (nevus unius lateris) or extended bilaterally (ichthyosis hystrix).[3]
Distribution is along Blaschko's lines and may have an abrupt midline demarcation.

Systematized epidermal nevus are often associated with multisystem abnormalities especially of the central nervous system, eyes and skeletal system, called epidermal nevus syndrome.[1]It is also reported the association with precocious puberty.[4]In our patient there was no other system abnormalities, however the patient should be followed up for the entire life time as neurological manifestations may sometimes present late in adulthood.[1]

Histologic patterns are variable, mostly demonstrate hyperkeratosis, acanthosis and papillomatosis. Histologic findings in this case reveal compact hyperkeratosis and papillomatosis which help to confirm diagnosis of systematized epidermal nevus. Epidermolytic hyperkeratosis is also one of the rare histologic patterns of epidermal nevus, which is identical as in linear bullous icthyosis erythroderma(BIE). Epidermolytic VIE and BIE had been shown to be due to genetic mosaicism (mutations in keratin1 or keratin10).Hence a parent with epidermal VIE can produce offspring with classical BIE[5]. Genetic counselling should be performed.

Treatment of systematized epidermal nevus based on size and distribution of the lesions. Full-thickness surgical excision is curative but in large lesions can lead to hypertrophic scars or keloid formation.[6] Others destructive therapies including cryotherapy and carbon dioxide laser therapy. Continuous-wave carbon dioxide laser vaporization has been used with benefit for extensive VEN.[7] Topical therapies including corticosteroids, tars, 5-fluorouracil, retinoic acid, salicylic acid, lactic acid, podophyllin, calcipotriol and calcitriol have all been used but are limited in benefits.[3]
Topical calcipotriol/betamethasone dipropionate combination ointment has been successfully used in a case of extensive epidermal nevus.[8] The successful use of oral retinoids have also been described.[9]

References

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