Systematized epidermal nevus: A rare case report and a review of literature.

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ABSTRACT:

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Systematized epidermal nevus is a rare skin condition. It is considered to be a benign epidermal proliferation, which is characterized by well-circumscribed, hyperpigmented verrucous lesions, usually distributed bilaterally along the lines of Blaschko. We report a 14-year-old Thai boy who presented with multiple asymptomatic hyperkeratotic brownish plaques all over the body since birth. The clinical presentation and histopathologic findings support the diagnosis of systematized epidermal nevus.

Key words : Systematized epidermal nevus, Epidermal nevus syndrome

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บทคัดย่อ:

นงศักดิ์ ชนะสุมน, แพรมาลา ฉายาวิจิตรศิลป์. รายงานผู้ป่วยโรค SYSTEMATIZED EPIDERMAL NEVUS วารสารผิวหนัง 2559; 32: 212-216.

สาขาวิชาโรคผิวหนัง คณะแพทยศาสตร์โรงพยาบาลรามาธิบดี มหาวิทยาลัยมหิดล

โรค Systematized epidermal nevus จัดอยู่ในกลุ่มโรคที่เกิดจากการแบ่งตัวผิดปกติของชั้นหนังกำพร้าซึ่งพบได้ไม่บ่อย ผื่นมีลักษณะเฉพาะคือ สีเข้ม ผิวขรุขระ ขอบเขตชัดเจน โดยตำแหน่งที่พบเป็นตามลำตัวและแขนขาทั้งสองด้านเรียงตัวตาม blaschko's lines รายงานฉบับนี้เป็นการนำเสนอผู้ป่วยเด็กชายไทย อายุ 14 ปี ซึ่งมีอาการแสดงและผลการตรวจเข้าได้กับโรค Systematized epidermal nevus

คำสำคัญ : โรค Systematized epidermal nevus, Epidermal nevus syndrome

Case report

A 14-year-old Thai boy presented with multiple asymptomatic hyperkeratotic brownish plaques on face, trunk, and extremities. The lesions first appeared since he was born, and gradually increased in size and thickness with age. His growth and developmental history were normal. He had no history of seizures, learning difficulty, hearing difficulty or altered vision. He had no other underlying disease. He denied having family members with similar findings. Physical examination revealed multiple hyperkeratotic brownish papules, which coalesce into plaques extensively on the face, both sides of trunk (predominantly on the left side), extremities and knuckles of both hands. The distribution was along the lines of Blaschko. There was a sharp midline demarcation on his trunk (Figure 1). The examination of other systems did not reveal any abnormality. Histological examination demonstrated compact hyperkeratosis, epidermal hyperplasia, and papillomatosis with "church-spire" pattern (Figure 2). Clinical presentation and histopathology are compatible with systematized epidermal nevus in the absence of epidermal nevus syndrome. The patient was treated with a topical keratolytic and the lesions appeared to be improving.

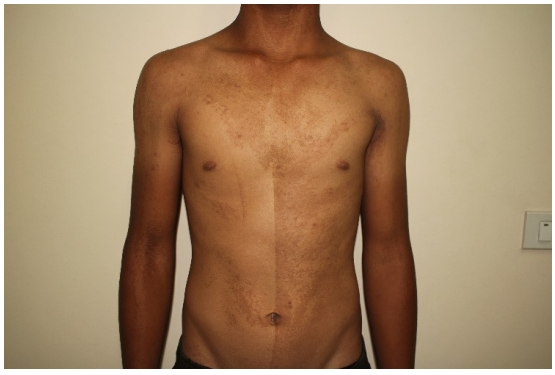

Figure 1 Multiple hyperkeratotic brownish plaques on neck, trunk, and extremities with a sharp midline demarcation

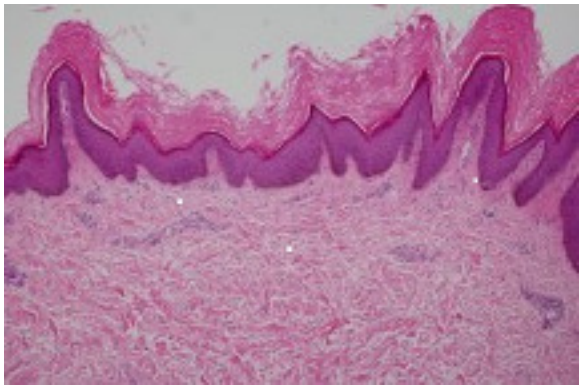

Figure 2 Compact hyperkeratosis and epidermal hyperplasia, papillomatosis with "church-spire" pattern

Discussion

Systematized epidermal nevus is a rare skin disease with extensive verrucous epidermal nevus which is defined as congenital, non inflammatory cutaneous harmatomas composed of keratinocytes. The age of onset is usually at birth or within the first year of life, however, late-onset lesions have also been reported.[1]

Systematized epidermal nevus usually presents with multiple well-circumscribed, hyperpigmented, papillomatous papules or plaques with extensive distribution, most commonly on the trunk, extremities and neck; with transverse configuration on the trunk and linear configuration on the limbs.[2]

The lesions can be limited to one half of the body (Nevus unius lateris) or bilateral (Ichthyosis hystrix).[3] Common distribution is along Blaschko's lines suggesting that they result from postzygotic somatic mutation in the skin and may have an abrupt midline demarcation.

Epidermal nevus rarely affects more than one member of the family. Somatic mosaicism for mutation has been detected in *FGFR3*, *PIK3CA* [4] and *RAS* genes, including KRAS, HRAS, and NRAS [5], as well as the gene mutation in keratin 1 and keratin 10[6]. The same keratin gene abnormalities have also been found in parents who have epidermolytic epidermal nevus and in their offspring who have bullous ichthyosiform erythroderma. So the epidermolytic epidermal nevus is thought to be a mosaic form of this type of ichthyosis[6].

Systematized epidermal nevus is often associated with multisystem abnormalities especially of the central nervous system, eyes and skeletal system, called "Epidermal nevus syndrome".[1] It is also reported to be associated with precocious puberty.[7]

In our patient there was no systemic abnormality, however, the patient should be followed up throughout his lifetime as neurological manifestations may sometimes present late in adulthood.[1]

Histologic patterns are variable and mostly demonstrate hyperkeratosis, acanthosis and papillomatosis. Histologic findings in this case reveal compact hyperkeratosis and papillomatosis which help to confirm the diagnosis of systematized epidermal nevus.

Table 1 Summary of previous case reports of systematized epidermal nevus

Reference	Age(years)/ gender	Age of onset (years)	Disease association	Familial involvement	Treatment	Outcome
Loff et al., 1994[13]	19/female	N/A	Trichiasis	N/A	Four-lid blepharoplasty	Resolution of ocular symptoms
Koh MJ et al., 2013[11]	5/male	4 months	-	-	Topical calcipotriol/ betamethasone dipropionate combination ointment once daily	Marked improvement after 2 months treatment
Dhanaraj M et al., 2015[1]	20/female	Since birth	-	-	CO_2 laser	N/A
Garg T et al., 2015[7]	3/male	3 months	-precocious puberty -spastic cerebral palsy	-	-topical tretinoin 0.025%	No response after 3 months treatment
Mishra V et al., 2015[3]	3/male	3 weeks	-	-	N/A	N/A

Treatment of systematized epidermal nevus is based on size and distribution of the lesions. Full-thickness surgical excision is curative but, in large lesions, can lead to hypertrophic scar or keloid formation.[8] Others destructive therapies include cryotherapy and carbon dioxide laser therapy.[9] Continuous-wave carbon dioxide laser vaporization has been used with benefits for extensive VEN.[10] Topical therapies including corticosteroids, tar, 5-fluorouracil, retinoic acid, salicylic acid, lactic acid, podophyllin, calcipotriol and calcitriol have all been used but showed limited benefits.[3] Topical calcipotriol/betamethasone dipropionate combination ointment has been successfully used in one case of extensive epidermal nevus[11] as well as oral retinoids.[12] Our patient was treated with 10% urea cream twice daily all over

body and 3% lactic acid cream once daily on the face. After three months of treatment, the response was slightly improved. The patient was also advised to be under surveillance for late onset neurological symptoms.

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