INFLAMMATORY LINEAR VERRUCOUS EPIDERMAL NEVUS

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ABSTRACT
Background: Inflammatory linear verrucous epidermal nevus (ILVEN) is a distinctive and rare type of epidermal nevus which the inflammatory is prominent. The lesions are pruritic, erythematous and scaling verrucous papules in linear arrays, if coalescence forms linear plaque. The incidence is unknown, but it was said 1 in 1.2 million people. Since 2000-2006, it is the first case in our Department.
Case: We reported a 10-year old Indonesian boy with pruritic reddish brown linear thickened of the skin along the right buttock to the right ankle and to the anterior right tight since he was 3 months old. On examination there were unilateral erythematous and brown papules, some are coalescence and erythematous brown plaque in linear arrangement along the Blaschko's line. There were no organs involvement. Histopathology examination was hyperkeratosis, parakeratosis, orthokeratosis focal on epidermis. Dermis was infiltrated by lymphocyte and hystiocyte cells around the capillary. The patient was treated with combination salicylic acid 5% in desoxymethason 0.25% ointment twice a day.
Discussion: The difficulty to treat was the problem in this case. Although many therapy had been mentioned as the treatment modalities for ILVEN, but none are satisfied. After 5 weeks, our therapy decreased the pruritic, moderate improvement on the knee and slightly improvement on other lesions. We change the therapy with retinoic acid 0.1% cream and clobetasol propionate 0.05% ointment.

Key word : *inflammatory linear verrucous epidermal nevus*

INTRODUCTION

Inflammatory linear verrucous epidermal nevus (ILVEN) is a distinctive type of epidermal nevus which also called dermatitic epidermal nevus. This type of epidermal nevus has prominent inflammation. It is a relatively rare linear psoriasiform plaque that usually present during childhood.[1,2,3]

In about 75% of cases, ILVEN have their onset during the first 5 year of life, most often in the first 6 months, although later onset has been reported. Females have been affected more frequently than males, its about 4:1 in ratio. Although generally sporadic, there has been reported familial cases.[1,3,4]

The lesions consist of pruritic, erythematous and scaling verrucous papules in linear array. The scaly erythematous papules coalescence to form linear plaque. The lesions most commonly on a limb, anywhere from buttock to the foot. The left leg is more frequently affected than the right. But occasionally the trunk may be involved. The

lesions are almost always unilateral. Although generally the lesions persistent, there is a a tendency for at least some of these lesion to resolve spontaneously at some stage.[1,2,4,5]

Epidermal nevus syndrome refers to a disease complex consisting of the abnormalities of the skin, eyes, nervous, skeletal and miscellaneous system.[5,6] It had been reported a case of ILVEN syndrome with bilateral vertebral artery occlusion.[7]

The histological appearance of ILVEN consist of acanthosis, hyperkeratosis and papilomatosis with elongated rete ridges. May be present alternating areas of hyperkeratosis with thickened of granular layer and parakeratosis without granular area. At dermis shows a dermal chronic inflammatory infiltrate consist of limphocyte and histiocyte.[5]

ILVEN is difficult to treat. Surgical excision, cryosurgery and laser have been reported successful to treat ILVEN. Topical application such as corticosteroid, anthralin, and calcipotriol have been attempted to the lesion but the lesion have not responded consistently to the therapy. Those topical application only provide temporary symptomatic relief and disminished the inflmmatory reaction.[1,2,4]

CASE REPORT :

A 10-year Indonesian old boy came to DermatoVenereology outpatient clinic of Dr M Djamil Hospital on May 4, 2006 with :

Chief complaint :

Appeared brown reddish linear thickened skin and itchy along the right buttock to the right ankle and along the right buttock to the anterior upper right tight since he was 3 months old.

Present illness history :

Appeared brown reddish linear thickened skin and itchy along the right buttock to the right ankle and along the right buttock to the anterior upper right tight since he was 3 months old. Initially, his mother noticed a brown patches like hair line along the right buttock to knee and brown patches on upper right tight when he was born and then when her son was 3 months old the pacthes became reddish, raised and wider. At this time the baby was able to scracth the patches because icthy. As he was grown older the patches became more icthy and wider. Sometimes the icthy made him difficult to sleep.

When he was 1 year old, his mother brought him to Dermato Venereology outpatient clinic of Dr. M Djamil Hospital and the doctor suggested to biopsy but his mother refused it. His mother tried some traditional medicine like powder of leaves but there was not improvement then his mother stopped the traditional medicine.

Since he was 2 years old, he went to the health centre because of itchy and got hidrocortison cream 2,5% and chlortrimeton unregularly untill now.

There had not been appeared blister on that thickened skin before.

He never had seizure nor mentally under development.

He never had difficulty to walk nor weakness on his foot.

He never had any difficulty/problem on eyes.

Family illness history :

There is no family having this disease.

Physical examination :

Within normal limit

Dermatology state :

Location	:	along the right buttock to the right ankle, along the right buttock to the anterior upper right tight (along the Blaschko's line)
Distribution	:	unilateral
Shape and arrangement	:	linear
Border	:	well defined
Size	:	plaque
Efflorescene	:	erythematous and brown papules, some are coalescene, erythematous and brown verrucous plaque, scales

Mucous membrane : normal

Nails : normal

Inguinal lateral limphe nodes : normal

Working diagnosis :

Inflammatory linear verrucous epidermal nevus

Differential diagnosis :

Nevus unius lateris

Lichen striatus

Incontinentia pigmentii (verrucous type)

Linear lichen planus

Suggestion :

- Histopathology examination
- Consult to Ophthalmology Department for koloboma, atrophy N. opticus, astigmat

- Consult to Pediatric Neurology Department for abnormalities N.cranial, mental retardation and seizure
- Consult to Orthopaedy Department for kyphosis, scoliosis, abnormality of limb musculoskeletal.

Therapy :

General therapy :

Do not scratch and manipulate the lesions

Explain to him and his mother that the lesions were difficult to treat

Stop hidrocortison 2,5% ointment

Systemic therapy :

Chlortrimeton 3 x ½ tablet (if itchy)

Topical therapy:

Lanolin 10%

Routine blood and urine were in normal limit.

May 10, 2006 :

A : the plaque were more itchy dan red

Eff : the papules and plaque were more erythematous, the scales were decreased

May 13, 2006 :

Histopathology result :

There was hyperkeratosis, parakeratosis, othokeratosis focal on epidermis.

Dermis are infiltrated by lymphocyte and hystiocyte cells around the capillary.

It is suitable for inflammatory linear verrucous epidermal nevus

Diagnosis :

Inflammatory linear verrucous epidermal nevus

Therapy :

Sistemic :

Chlortrimeton 3 x ½ tablet (if icthy)

Topical :

Combination salisilic acid 5% and desoxymetason 0,25% ointment (2x/day)

Result from Ophthalmology Department, Orthopaedy Department, Pediatric Neurology Department: there was no abnormality

Prognosis:

Quo ad vitam : bonam

Quo ad sanam : dubia ad malam

Quo ad cosmeticum : dubia ad malam

Follow up:

May 23, 2006 :

Anamnesis : itchy was decreased

Efflorescene : erythematous papules became brownish, some were still erythema, some of the plaque were thinner, the scales are decreased.

Therapy :

Chlortrimeton 3 x ½ tablet (if itchy)

Combination salisilic acid 5% and desoxymethason ointment 0,25%

June 20, 2006 :

Anamnesis :

Itchy was decreased

Efflorescene : thinner brownish papules and plagues on the knee, remained erythematous papules and plagues on anterior and posterior thigh.

Interpretation : there was slight improvement

Therapy :

- Retinoic acid 0,1% cream in the evening
- Clobetasol propionate ointment in the morning
- Chlortrimeton ½ tablet if itchy

Discussion

Diagnosis ILVEN was based on anamnesis, dermatology state and histopathology examination. In Indonesia the exact incidence of ILVEN is unknown. The literature said that the incidence 1 in 1,2 million people.[8] This case is the first case in our department.

There are several differential diagnosis for ILVEN including nevus unius lateris, lichen striatus, incontinetia pigmentii, linear lichen planus. Each lesion has characterized feature. Nevus unius lateris is rarely puritic and the histopathology feature show hyperkeratosis, acanthosis and papillomatosis. Lichen striatus is rarely pruritic, resolve spontaneously and the histopathology there is no or little acanthosis. Incontinentia pigmentii was preceded by bullous phase before verrucous phase. Linear lichen planus was marked by purple, polygonal papules, pruritic and the histopathology shows hyperkeratosis, elongation of the rete ridges that resembles saw tooth pattern and band like lymphocytic infiltate on dermis.[5]

We tried to find some abnormalities in epidermal nevus syndrome on this patient and so far we did not find any abnormalities on eyes, neurological and musculoskeletal system.

The problem in this case is ILVEN is difficult to treat. Treatment with potent corticosteroid is partially effective. Taru G et al.,(New Delhi, 2001) reported that betamethason valerat 0,12% ointment for five months gave initial improvement in reducing erythema and pruritic but subsequently the lesion is resistant to the treatment. Then he combined betamethason valerat 0,05% with silisic acid 3% for 4 weeks with no improvement.[9] Kagauchi et al.,(Yokohama,1999) treated ILVEN with topical corticosteroid and vaseline containing salisilic acid, there was improvement in pruritus and slightly decreased the eruption.[10] Succesfull treatment with calcipotriol has been reported, but still in limited effect. Taru G et al .,(New Delhi, 2001) reported that after application calcipotriol 0,05% ointment after 16 weeks there was complete flattening. But after the calcipotriol was stopped the lesion reappeared.[9] Topical tretinoin and fluorouracil cream had been beneficial result, but need long term continued use. Surgical removal carries the highest success and effective for some cases. It required excision particularly for small lesions and the procedures need deep excision to underlying dermis. This surgical have limitations including potential for scarring and increased potential for

adverse effect. This option is impractical if the lesion is extensive or located in anatomical site where the procedures can be difficult to perform. Superficial procedures tend to followed by rapid recurrence.[1] Laser therapy has been successfully treated ILVEN. The only notable side effect of laser therapy was a pale discoloration limited to the treated site.[11] Combination silisilic acid 5% and desoxymethason 0,25% cream in this patient did not give a good result. After 1 month treatment, there was improvement on the knee, but slight improvement on the thight and buttock. We changed the therapy with retinoic acid 0,1% in the evening and clobetasol propionate 0,05% ointment in the morning.

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