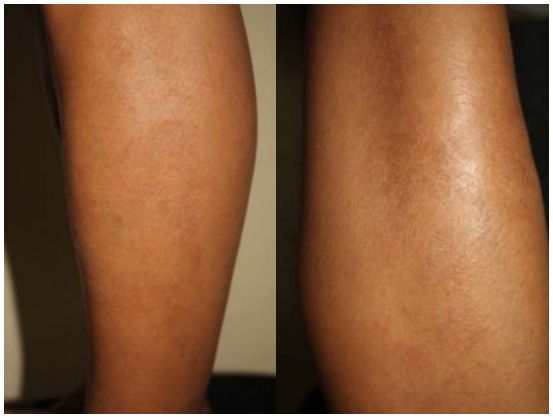
Case 24.1
A 49-year-old Thai woman from Khonkaen

Chief complaint: Pruritic annular erythematous rash on right arm for 2 months


(Fig. 24.1.1)

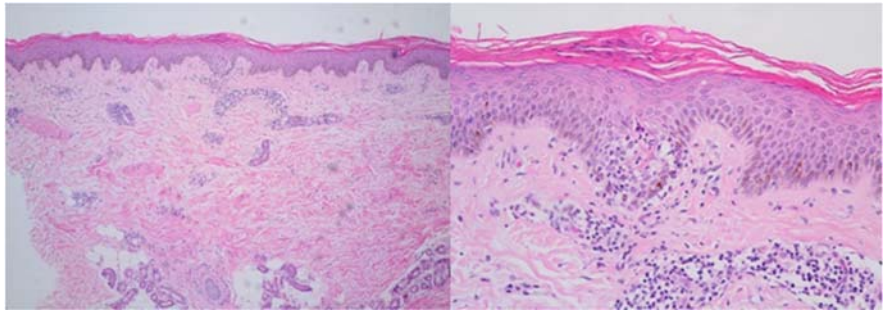
Present illness: 2 months ago, she had pruritic erythematous rash on right arm. The rash gradually expanded peripherally over time and progressed to her left leg. The lesion worsen by exposure warm weather or over sweating. She had tried using topical steroids resulting in marked improvement.

Past history: Multinodular goiter S/P total thyroidectomy

Physical examination: Unremarkable

Dermatological examination: (Fig. 24.1.1)
Few ill-defined annular erythematous patches with peripheral scales on right arm and left leg

Histopathology: (S17-22956, Right arm) (Fig. 24.1.2)


(Fig. 24.1.2)

- Focal epidermal spongiosis with neutrophils and necrotic cells at acrosyringium
- Superficial and deep perivascular and periductal inflammatory cell infiltrates of lymphocytes in the dermis

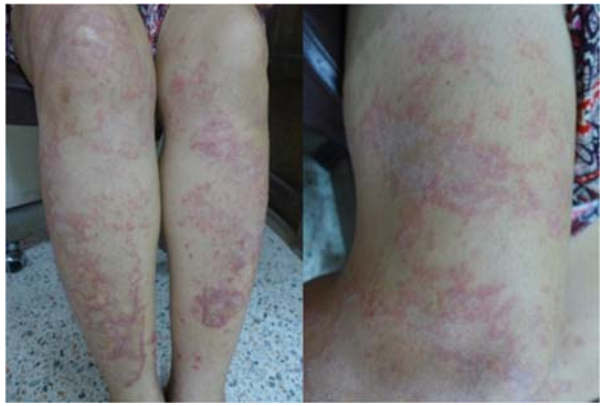
Diagnosis: Erythema papulatum centrifugum

Investigation: KOH examination left arm: Negative

Treatment: 0.1% betamethasone valerate cream apply lesions twice daily

Case 24.2
A 33-year-old Thai woman from Phuket

Chief complaint: Pruritic annular erythematous rash on both legs for 2 months


(Fig. 24.2.1)

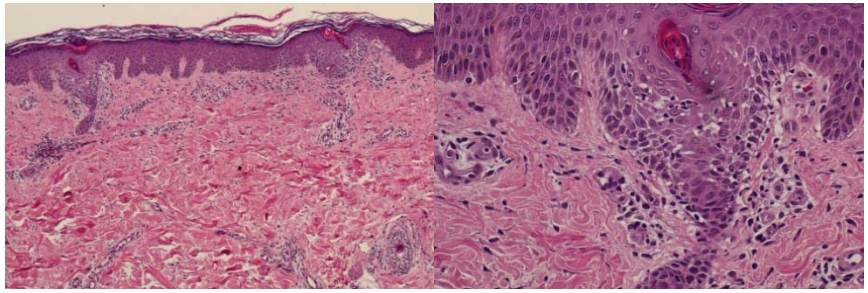
Present illness: 2 months ago, she had pruritic rash on both legs. The lesion progressed to both thighs, arms and back. The lesions worsened by warm weather such or over sweating.

Past history: Hypertension

Physical examination: Unremarkable

Dermatological examination: (Fig. 24.2.1)
Multiple confluent annular erythematous plaques with peripheral tiny scaly erythematous papules on both legs, thighs, arms, and back

Histopathology: (60-0508, Right thigh)


(Fig. 24.2.2)

- Focal epidermal spongiosis and mounds of parakeratosis predominately around acrosyringium
- Superficial perivascular and mid perivascular and periductal inflammatory cell infiltrate of lymphocytes in the dermis

Diagnosis: Erythema papulatum centrifugum
Laboratory investigations:
- LFT: AST 23 U/L, ALT 34 U/L
- BUN 12 mg/dl, Cr 0.58 mg/dl
- Stool examination: No parasite found
- CXR: No active chest disease

Treatment: 0.05% betamethasone dipropionate cream apply twice daily

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Consultant: Vipawee Ounsakul, MD

Discussion:

Erythema papulatum centrifugum (EPC) was first described in 1962 by Japanese dermatologist Watanabe as eczematous lesions extending centrifugally. To date, there are more than 150 cases have been reported, mostly in Japanese literature.[1,2,3]

EPC is predominantly found among middle-aged, male patients. Age of onset in more than 90% of cases was from 41-60 years, and the male:female ratio was 9:1.[3]

The skin lesion usually begins with a group of erythematous papule, macules or papulovesicles, which gradually expanding peripherally and forming an annular configuration with central clearing in the center. The lesions continuously enlarge centrifugally and sometimes reach a diameter from 10 to more than 50 cm.[3]

Ohmori R et al. reviewed 152 Japanese cases, the largest EPC case series. The result showed male dominance, middle-age onset (approximately 46 years). The patients usually had single or <5 lesions at the same time. The sites of predilection were trunk and extremities.[3]

The exact etiology and pathogenesis of EPC is still unknown. There were reports of many exacerbating factors, such as warming, summertime, sweating, and hyperhidrosis.[3,4]

Most of the reported EPC cases were not associated with other systemic diseases, though 2 from 152 cases were accompanied by polymyositis with interstitial pneumonia, and advance gastric cancer.[3] There was a single case report of EPC developing around melanoma, suggested EPC as a warning sign or paraneoplastic process in patient with melanoma.[4,5]Ueda C et al.[6] proposed the clinical criteria for a diagnosis of EPC including

1) Onset mostly in elderly males
2) An increase of symptoms during the period from spring to summer
3) Distribution of skin lesion on the trunk and proximal extremities
4) A clinical appearance characteristic of incomplete annular erythema and peripheral papules, with intense pruritus and outward expansion
5) Exacerbation due to external irritation, sweating, warming, and drinking alcohol
6) Multiple relapses of symptoms in the same regions.

The histopathology of EPC shows minimal epidermal changes, such as parakeratosis and spongiosis. In addition, there is a superficial and mid-dermal perivascular mononuclear cell infiltration. Moreover, mononuclear cells also infiltrate around intraepidermal or dermal eccrine ducts, which are the characteristic findings of EPC.

Differential diagnosis of EPC includes other figurate erythema such as erythema annulare centrifugum (EAC), and erythema gyratum repen (EGR). The significant clinical difference of superficial EAC and EPC is the features of annular borders. Superficial EAC shows superficial cord-like or trailing scale, while EPC shows grouped papule or maculopapules in the annular border. Histopathologically, although EPC and EAC share several histological findings such as spongiosis, parakeratosis and superficial perivascular mononuclear cell infiltrate, inflammation around intraepidermal and dermal eccrine ducts is reported only in EPC.[3,7]

EPC usually self-limited within a few months but the symptoms frequently relapse. There is no standard treatment in EPC. Response to treatment are varies and inconclusive, although some reports suggested the efficacy of topical potent corticosteroid, sulfonamide or griseofulvin. Removal of exacerbating factors such as reducing hyperhidrosis by using topical aluminium chloride and discontinuing the use of sauna were also marked effective.[6]

Our patient had annular erythematous rash with peripheral

tiny scaly erythematous papules on both legs, arms and back. Histopathology showed superficial and mid perivascular and periductal inflammatory cell infiltrate of lymphocytes in the dermis. Based on clinical and pathological findings, the diagnosis of EPC was confirmed in our patient. Treatment was effective with potent topical corticosteroid.

In conclusion, the diagnosis of EPC is mainly base on the typical clinical features together with the histopathological findings.

References:

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7. Mir A, Terushkin V, Fischer M, Meehan S. Erythema annulare centrifugum. Dermatol Online J. 2012;15:18:21.