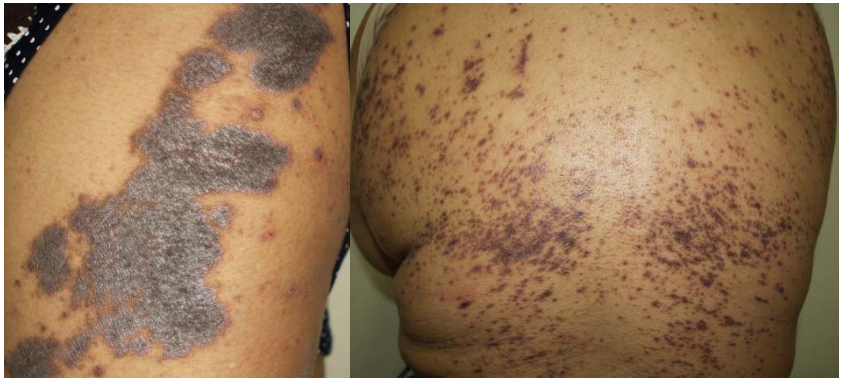
Case 11
A 48 year-old Thai woman from Bangkok
Chief complaint: Generalized pruritic violaceous rash for 3 months



Present illness:
Three months PTA, the patinet developed erythematous to violaceous rash on left thigh that progressed to her whole body.The rash was very itchy and had no clinical relation to sun exposure. She had previous treatment with topical steroid but showed no improvement.

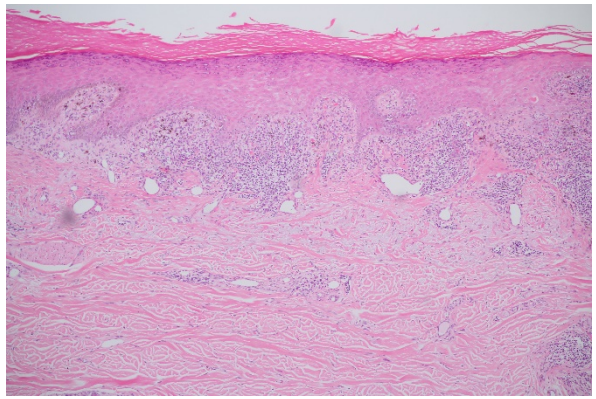
Past history:
None

Physical examination:
HEENT: Not pale conjunctivae, anicteric sclerae
Lymph node: Not palpable
Heart&lung: Normal
Abdomen: Soft, not tender, no hepatosplenomegaly

Dermatologic examination:

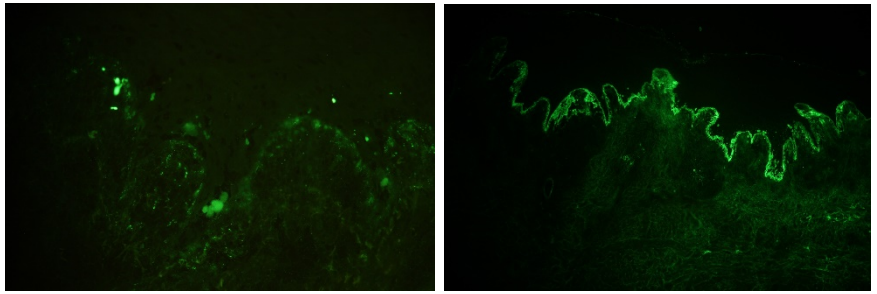
- Generalized discrete erythematous to violaceous flat-topped papules and plaques on trunk and both extremities.
- Large well-defined violaceous flat-topped plaques on left thigh.

Histopathology: (S16-16112A, left thigh)



- Compact hyperkeratosis, hypergranulosis, and saw-tooth epidermal dermal hyperplasia
- Dense band-like lichenoid inflammatory cell infiltration of lymphocytes with some melanophages obscuring the dermoepidermal junction

Direct immunofluorescense test:



- Positive cytoid body of IgG, IgM, C3

- Positive fibrin deposition at DEJ in shaggy pattern

Diagnosis: Generalized lichen planus

Investigations:
- CBC: Hb 13 g/dl, Hct 39%, WBC 9,680/cumm (N 57%, L 32%, Mo 5%, Eos 5%, Bas 1%), platelet 380,000/cumm
- LFT: AST 40 U/L, ALT 43 U/L, ALP 80 U/L, GGT 60 U/L, TP 72.1 g/L, Alb 40.8 g/L, TB 0.3 mg/dl, DB 0.1 mg/dl
- Anti-HCV negative

Treatment:
- Desoximetasone cream apply lesion bid.
- Acitretin 25 mg/day

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Discussion:

Lichen planus is an inflammatory mucocutaneous dermatosis that commonly affects the skin and oral mucosa. Specifically, cutaneous lichen planus (CLP) affects 0.2–1 % of the adult population[1]

Classic LP lesions commonly present with the four P's: purple, pruritis, polygonal, and papules/plaques. The papules often have dry, shiny surfaces with branny scale that forms fine, whitish streaks known as Wickham's striae. Many variants in morphology and location also exist, including oral, nail, linear, annular, atrophic, hypertrophic, inverse, eruptive, bullous, ulcerative, LP pigmentosus, lichen planopilaris, vulvovaginal, actinic, lichen planus-lupus erythematosus overlap syndrome, and lichen planus pemphigoides. Many of the variants occur much more infrequently than classic LP.

LP lesions are typically symmetric distribution and can affect any area of the body, but LP tends to favor dorsal surface of hand, shin, trunk and sacral region and especially flexural surfaces such as the forearms, wrists, and ankles.[2]

Associated factors and disease conditions seen in LP include but are not limited to stress/anxiety, hepatitis C virus (HCV), autoimmune diseases, internal malignancies, dyslipidemia, and viral infections.[3] A meta-analysis to find the potential association found LP patients have 5.58 times the odds of having concurrent HCV infection than the control population (95% CI: 3.72–8.38, $P < 0.05$) and were significantly more likely to have dyslipidemia, with a pooled odds ratio of 1.74 (95% confidence interval [CI]: 1.19–2.54, P = 0.004).[4-6]

Pathogenicity of LP lesions involves the autoimmune mediated lysis of basal keratinocytes by CD8+lymphocytes, though definitive etiological triggers are still unknown.[2]The diagnosis of lichen planus is usually clinical. However, microscopic examination of a lesion may be used for confirmation.

Pathologic changes typically shows circumscribed, wedge shaped hypergranulosis in the epidermis, marked hyperkeratosis, and irregular saw-tooth like acanthosis of rete ridges. The dermal–epidermal junction typically shows signs of vacuolar degeneration with apoptotic keratinocytes, while the upper dermis characteristically contains a dense, band-like lymphocytic infiltrate that can obscure the dermal–epidermal junction. Civatte bodies, hypothesized to be apoptotic keratinocytes ready for phagocytosis, can be seen in the epithelium and upper dermis. Direct immunofluorescence often reveals a large number of IgM-staining cytoid bodies in the dermal papillae or peribasilar areas.[7,8]

For the management of CLP, no single treatment has emerged as the standard of care. Therapies such as topical and systemic corticosteroids, retinoids, calcineurin inhibitors, immunosuppressive agents, phototherapy, and biologics are used

to decrease the time to lesion resolution, alleviate patient discomfort, and enhance quality of life.[9]

CLP may resolve spontaneously, the goals of therapy are to shorten the time between onset and lesion resolution and to reduce itching. In a long-term follow-up study of patients with CLP, Irvine et al[10] found that the duration of the inflammatory eruption was 1 year or less in 68 % of patients and 6 months or less in 42 %.

The decision on which CLP treatment to use for a specific patient should be based on disease severity as well as the patient's medical background, the safety profile of the treatment, and patient preferences. Although some treatments were found to be less successful than others in achieving a specific efficacy outcome, they may be considered first-line options in specific circumstances.

From recent meta-analysis[11]; systemic treatments with acitretin, sulfasalazine, and griseofulvin[12-15] were associated with increased overall response rates compared with placebo. Hydroxychloroquine was more effective than griseofulvin in achieving an overall response.[16] NBUVB was more effective than 6 weeks low-dose prednisolone in achieving a complete response[17], and prednisolone was more effective than enoxaparin.[18] Methotrexate was effective, with a nonsignificant difference in the complete response rate in comparison with oral betamethasone.[19] Topical treatment with betamethasone valerate 0.1 % ointment had comparable efficacy to calcipotriol ointment.[20]

In this case, We chose acitretin for treatment in generalized LP.The patients must be monitored closely for potentially serious side effects. Acitretin is highly teratogenic, and can cause serious skeletal and metabolic defects in the fetus. Therefore, women must strictly avoid pregnancy for at least 3 years after the discontinuation of the medication. Moreover, it is recommended that patients undergo testing of liver enzymes and lipid levels every 3 months to monitor for liver side effects and hyperlipidemia.[9,21,22] The safety/efficacy ratio of acitretin should be thoroughly discussed with the patient.

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