Case 7
A 66 year-old Thai woman from Bangkok
Chief complaint: Widespread pruritic red plaques on the trunk for 1 month

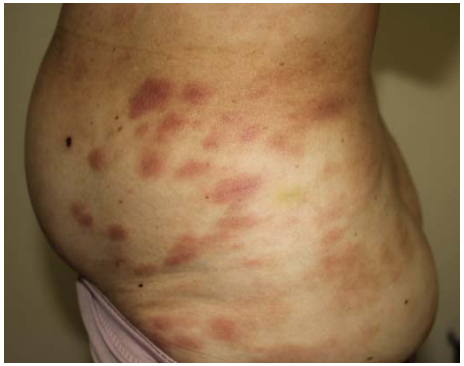
Fig. 7.1

Present illness:
One month ago, the patient developed widespread pruritic erythematous plaques on her trunk and back. One week later, rashes erupted on her thighs and legs. She denied changing of any skin care products or medications, recently.

Past history: Hypertension, dyslipidemia

Physical examination:
GA: A Thai elderly female, good consciousness
VS: T 37.2°C, R 18 /min, PR 70 /min, BP 114/71 mmHg
HEENT: No pale conjunctivae, anicteric sclerae
CVS&RS: WNL
Lymph node: Not palpable
Abdomen: Soft, not tender, no hepatosplenomegaly

Skin examination: (Fig. 7.1)
Multiple well-defined discrete, partially blanchable, erythematous plaques, varying in size from 1-4 centimeters, scattering on trunk, back, buttock and both legs

Histopathology: (S16-035971, right flank) (Fig. 7.2, 7.3)
- Dense superficial and deep perivascular and interstitial infiltrate of lymphocytes in the dermis
- Thicked papillary dermis, composed of coarse collagen bundles and lichenoid infiltrate of lymphocytes with subtle epidermotropism

Immunohistochemistry:

- CD3/4/8: Positive
  CD20: Scatter positive
  CD30: Negative

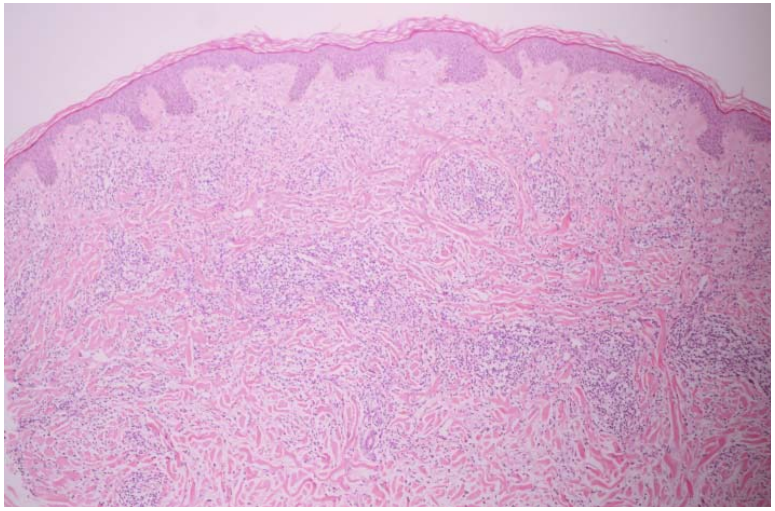
Fig. 7.2

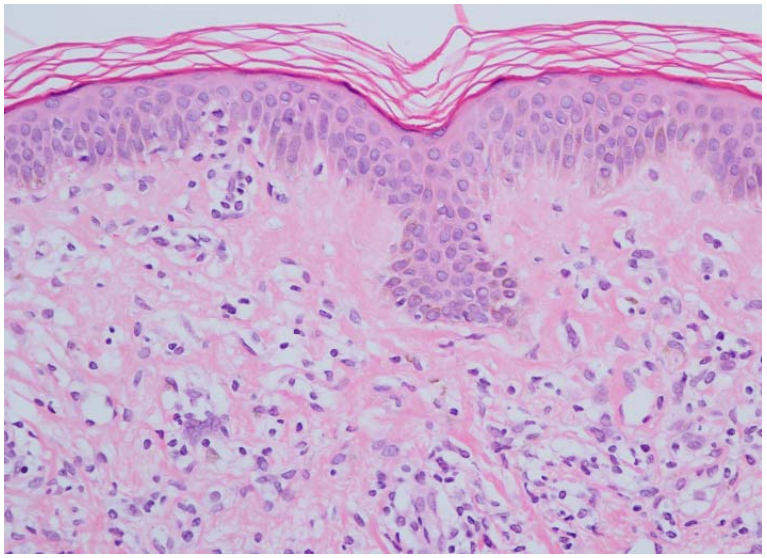

Fig. 7.3

Investigations:

CBC: WBC 8,510/uL (P59%, L29%, Mo5%, Eo6%, Ba1%), Hb 12.1g/dL, Hct 38.6%, Platelet 246,000/uL
LFT/Cr: WNL
LDH 194 U/L (125-220 U/L)
CT chest & abdomen: Normal, no lymphadenopathy

Diagnosis: Interstitial mycosis fungoides

Treatment: Psoralen plus UVA (PUVA)

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Discussion:

Mycosis fungoides (MF) is the most common cutaneous T-cell lymphoma, comprising of almost half of all cases.[1] Several clinicopathologic variants have been reported.[1-3] Interstitial mycosis fungoides (IMF) is a rare histological variant of MF that may mimic other inflammatory dermatoses, mainly interstitial granuloma annulare (GA), inflammatory morphea, and interstitial granulomatous dermatitis (IGD).[4] IMF was first described by Ackerman AB (unpublished observation) and subsequently was mentioned by Shapiro and Pinto.[4] Few small series and case reports of IMF have been published to date.[4-9] Cases of IMF have been observed in early and advanced stages, but are predominantly found in early stages.[5] Lesions with features of IMF correspond clinically most commonly to patches and plaques.[4-5] Buruk et al. reported a unique variant of interstitial MF presenting with lichen sclerosus-like lesions. IMF is observed either before, concomitant with or after conventional MF and rarely represents the only histologic presentation.[5]

Histopathologically, variably long, linear aggregates of dermal lymphocytes splaying the collagen fibers with few histiocytes infiltration, involving predominantly the superficial and mid-dermis or the entire dermis. Atypia is often minimal or even absent. A band-like infiltrate, epidermotropism, Pautrier's collection were also observed in some cases.[4-6]

Immunohistochemical studies revealed a cytotoxic phenotype (50%) which defined as CD8+ (one case reported CD8- but TIA+).[5] Although a cytotoxic phenotype is not uncommon in MF, it seems more frequent in IMF than in conventional MF.[5]

The main differential diagnoses of IMF are inflammatory morphea, granuloma annulare, and IGD (including cases associated with autoimmuneor rheumatic conditions, drug reactions, and infections by Borrelia).[4-5,8-9] Lack of sclerosis of the collagen fibers as well as lack of clusters of plasma cells allows distinction from

inflammatory morphea. Lymphocytes may be seen along the dermal–epidermal interface in both conditions, but slight vacuolar interface alteration, slight squamotization of basal keratinocytes, and effacement of the rete ridge pattern favor inflammatory morphea. Moreover, inflammatory morphea shows focal loss of elastic fibers but no significant alteration in IMF.[4-5,9]

An interstitial expression of GA can sometimes be confused with IMF, as both share an interstitial dermal mononuclear infiltrate, sometimes accompanied by increased interstitial dermal mucins. The presence of interstitial lymphocytes predominating over histiocytes is a strong clue to diagnose IMF, allowing differentiation from the interstitial type of GA.[5] GA also lacks of the epidermotropism and Pautrier's collections seen focally in IMF.[4,5]

Previous study reported the presence of collagen fibers surrounded by lymphocytes (rosetting) in IMF, which is similar to what can be observed in IGD.[5] Interestingly, a similar finding was published as "floating" collagen fibers by Ferrara et al.[6] However, the distinction in IGA is the predominant of histiocytes instead of lymphocytes.[5]

The clinical behavior, prognosis and treatment for IMF was not well documented, however, previous studies did not demonstrate an aggressive course.[4-5]

References

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