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Dermato-venereological Quiz

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A 77-year-old gentleman presented with recurrent itchy patches and plaques on four limbs on and off for few years. He was treated by his general practitioner with emollients and topical steroids for xerosis and discoid eczema. Recently the skin lesions had become more persistent. He was referred to the dermatology clinic for further management. There were no systemic symptoms and his past health was unremarkable.

On examination there were a few erythematous patches and plaques on both lower limbs (Figure 1). Mild skin atrophy was noted. The scalp, nails, genitals and oral mucosa were unremarkable. There was no palpable nerve thickening, no sensory loss or neurological signs and there was no hepatosplenomegaly, no cervical, axillary or groin lymphadenopathy. Skin scraping for microscopy and fungal culture were negative. An incisional skin biopsy was obtained from an arcuate lesion on the left calf (Figure 1) for histopathological examination (Figures 2 and 3).

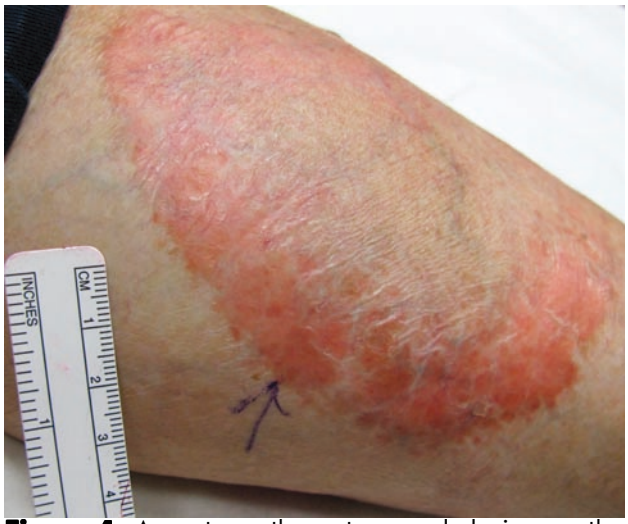

Figure 1. Arcuate erythematous scaly lesion on the left calf.

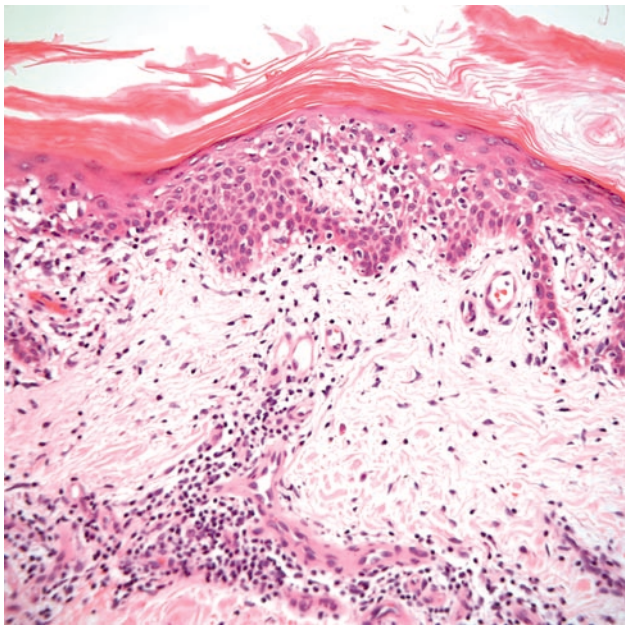

Figure 2. Inflammatory infiltrate of atypical lymphocytes in the superficial dermis and epidermis (H&E stain, Original magnification x 200).

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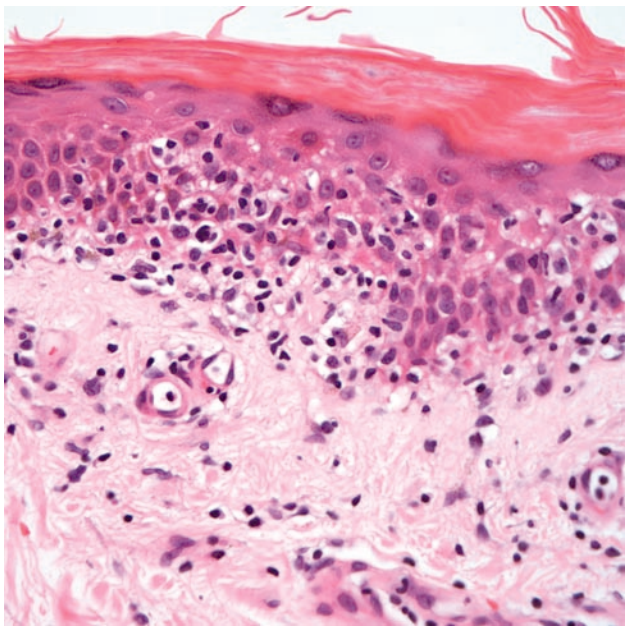

Figure 3. Convoluted lymphocytes noted along the dermo-epidermal junction with prominent epidermotropism (H&E stain, Original magnification x 400).

Questions

1) What are the clinical differential diagnoses?
2) What are the histopathological findings?
3) What is the diagnosis?
4) What is the treatment for this disease?

(Answers on page 107)

Answers to Dermato-venereological Quiz on pages 94-95

1. Differential diagnosis includes mycosis fungoides, tinea corporis, psoriasis, erythema annulare centrifugum and leprosy.

2. Histopathological examination shows linear array of convoluted lymphocytes along the dermo-epidermal junction with prominent epidermotropism. The papillary dermis shows moderate perivascular and interstitial infiltration of atypical lymphoid cells. Immunohistochemical staining shows that the majority of the lymphoid infiltrate belongs to T-cell lineage (CD3-positive). T-cell receptor γ-chain gene rearrangement is positive which is consistent with the presence of a clonal cell population. The result of molecular clonality tests should be interpreted in the context of clinical, histological and immunophenotypic data.

3. The diagnosis is consistent with mycosis fungoides.

4. Management is based on the stage of the disease, the patient's medical condition, available facilities and expertise. Treatment may improve symptoms and quality of life but may not alter the prognosis or long-term survival. Localised patches may be treated with emollients, topical steroids or topical chemotherapy (Stage 1a). Phototherapy with PUVA or Re-PUVA may control widespread patches and plaques (Stage 1 and 2a). Localised nodules are treated with localised radiotherapy; widespread nodules require total skin electron beam therapy (Stage 2b). Cases in the erythrodermic stage may be treated with PUVA plus interferon-alpha-2a or methotrexate (Stage 3). The prognosis is poor for lymph node and visceral involvement despite localised radiotherapy and combination chemotherapy (Stage 4). Research is ongoing on experimental therapies.