A Man with Asymptomatic Annular Lesions

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Date:	12 March, 2001
Venue:	Yaumatei Dermatology Clinic
Organizer:	Social Hygiene Service, DH; Clinico-pathological Seminar

CASE SUMMARY

History

A 37-year-old man presented with a six-month history of erythematous scaly annular lesions on his back. The lesions were asymptomatic but were slowly increasing in size. There was no sensory loss. He enjoyed good past health. The family history was unremarkable.

Physical examination

There were multiple erythematous annular lesions with fine scale on the back. There were hypopigmented areas within the annular lesions (Figure 1). The sensation was intact. There was no palpable lymph node and no organomegaly.

Differential diagnosis

The differential diagnoses include erythema annulare centrifugum, mycosis fungoides, leprosy, granuloma annulare and subacute cutaneous lupus erythematosus.

Investigations

Blood tests including complete blood picture, liver and renal function tests, lactate dehydrogenase and fasting blood sugar were normal. The erythrocyte sedimentation rate was one. Autoimmune markers like ANF and anti-ENA were negative. C3, C4 and immunoglobulin pattern were normal. No Sezary cell was detected in the peripheral blood. Skin scrapping for fungal study was negative.

A skin biopsy was performed for histopathology and direct immunofluorescent study. It showed that the epidermis was thickened with compact orthokeratosis decorated by areas of parakeratosis. There were focal but dense lymphocyte exocytosis, many with pericellular halo, in the epidermis. The lymphocytes lined along the DEJ in one focus. Some of them showed nuclear enlargement and irregularity. There was no definite Pautrier's microabscess. In the dermis, there were palisading granulomas with central collagen degeneration and mucin deposition. There were scattered atypical lymphoid cells in the dermis. The immunostaining confirmed almost exclusive T-cell infiltration. Direct immunofluorescence was negative. T-cell receptor (TCR) gene rearrangement study was negative. The diagnosis was mycosis fungoides associated with granuloma annulare-like changes. A negative TCR gene rearrangement study did not negate the diagnosis of mycosis fungoides.

Staging investigations were performed. Chest X-ray and ultrasonogram of abdomen were normal. CAT scan of thorax and abdomen did not show any enlarged

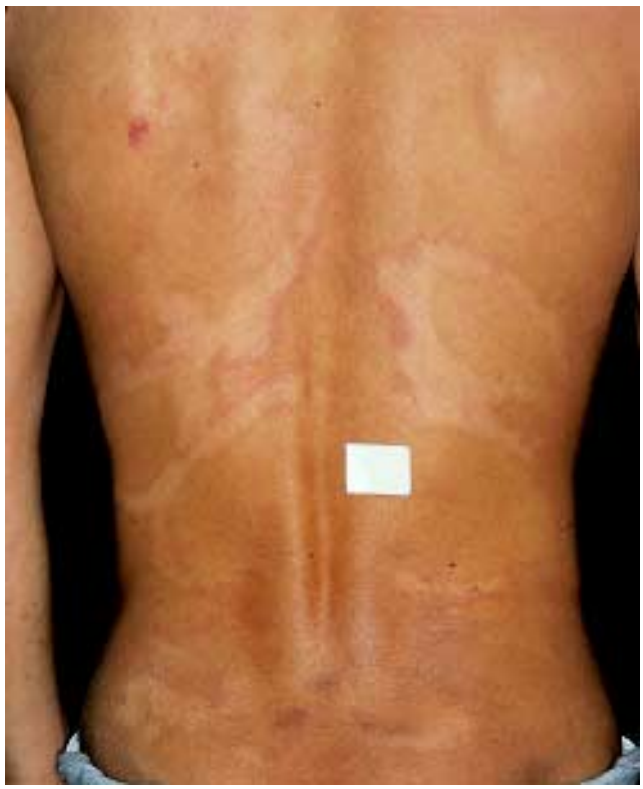

Figure 1: On the back, there are multiple erythematous annular lesions with hypopigmented areas within

lymph nodes. The spleen was normal in size. The liver was enlarged with smooth outline. It measured 16.1 centimetres along the midclavicular line (normal: 15.5 centimetres). There was no focal mass. There were cortical renal cysts up to 1.5 centimetres in the lower poles of both kidneys.

Management

The patient was referred to medical unit for assessment of enlarged liver. He was treated with PUVA therapy and the lesions responded well.

REVIEW ON GRANULOMATOUS MYCOSIS FUNGOIDES

Granulomatous mycosis fungoides (GMF) is a rare histological variant of cutaneous T-cell lymphoma (CTCL). It was first described by Ackerman and Flaxman in 1970.[1] At least 32 cases of GMF had been reported in the English literature.[2-18] Granulomatous changes were present in up to 4% of CTCL.[5] It can occur in all stages of CTCL. The clinical appearance of GMF is identical to classic MF. It can appear as patch, plaque, tumour and annular lesions.

Histopathology

GMF shows features of MF admixed with giant cells and collections of epithelioid histiocytes within the infiltrates. There are three histological patterns, namely, sarcoidal, granuloma annulare-like and granulomatous with multinucleated giant cells.

Pathogenesis

The pathogenesis of GMF is poorly understood. There are a few hypotheses. Some suggested that it might be true sarcoidosis preceding MF. Some proposed that it might be sarcoidal reaction to the underlying MF or granuloma formation in response to cytokines produced by the tumour cells.

Management

GMF is managed with the same protocol for classic MF. Treatment options depend on the stage of the disease.

Prognosis

GMF has a variable prognostic spectrum.[2,7] It was previously thought to be associated with good prognosis and long survival.[17] However, some recent reports showed aggressive nature and poor outcome in GMF.[2,3,7,10,14]

Diagnostic pitfalls

The diagnosis of CTCL can be obscured if the granulomas are more prominent which can mask the neoplastic lymphocytic infiltrate. The clinical information is important. CTCL should be considered in the clinical differential diagnosis to alert the pathologist. The biopsy specimen should be scrutinized for epidermotropism and atypical lymphocytes. TCR gene rearrangement is helpful in identifying monoclonal proliferation of T cells. However, monoclonal bands are present in only 50% to 83% of MF specimens.

GMF has to be distinguished from granulomatous slack skin disease. Granulomatous slack skin disease is a separate entity although it shares similar histological features. Their features were compared in Table 1.[19]

Table 1.

Granulomatous mycosis fungoides	Granulomatous slack skin disease
Clinical features:	
Patch, plaque, tumour, annular	flexural involvement; gradual development of bulky pendulous skin folds
Histopathology:	
3 patterns: sarcoidal pattern; granuloma annulare-like pattern (palisading granuloma and central necrobiosis); granulomatous with multinucleated giant cells	stereotypic appearance; granulomatous inflammation; cutis laxis; permeation of subcutis and dermis by lymphocytes; more marked epidermotropism; more even distribution of granuloma and giants cells within the infiltrate; giant cells have larger number of nuclei; elastolysis involving the full thickness of dermis; wreath-like arrangement of mononuclear cells around the giant cells
Clinical course:	
Variable	Indolent

Learning points:

Granulomatous mycosis fungoides is a rare histological variant of mycosis fungoides. Histological features of mycosis fungoides such as epidermotropism and atypical lymphocytes must be looked for in the presence of granulomatous infiltrate in order not to miss the diagnosis. TCR gene rearrangement may provide additional information.

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