Folliculotrophic mycosis fungoides masquerading as lepromatous leprosy

J Liyanage[1], M Dissanayake[2], R M P Rathnayake[3]

Sri Lanka Journal of Dermatology, 2015, **17**, 14-16

Abstract

Folliculotrophic lymphoma is a variant of cutaneous T-cell lymphoma with distinct clinical and pathological features. Its clinical presentation is variable although it shows predilections for head and neck area. We describe a patient with an infiltrated rash resembling a leonine face that was initially treated as lepromatous leprosy. Poor response to multi drug therapy prompted further investigations and subsequently, the diagnosis was made as folliculotrophic mycosis fungoides.

Introduction

Leonine face is an extreme manifestation of the infiltration of the facial skin. Possible etiologies for leonine face are lepromatous leprosy, actinic reticuoid, folliculotrophic lymphoma and sarcoidosis. Differentiation among above is a challenge to the physician. Slit skin smear, repeated skin biopsies and immunohistochemistry are the key investigations to arrive at a diagnosis.

Case report

A 73 year old male presented with severely itchy skin lesions which initially appeared over the face and then extended to upper torso over an 18 months period. In the early stages of the disease, facial lesions were discrete and later they had coalesced to involve the entire face.

Patient was initially diagnosed at a peripheral unit as lepromatous leprosy and treated with multi bacillary treatment for 10 months period with a poor response.

On clinical examination, facial skin was erythematous and indurated resembling leonine face and both ear lobes involvement was apparent. Skin over the upper trunk showed induration (Figure 1). Discrete enlarged lymph nodes were found, which were less than 1 cm in diameter over both axillae.

Slit skin smear was negative for AFB. Skin biopsy revealed infiltration of hair follicles with atypical lymphocytes. Intervening epidermis showed epidermotrophism with Pautrier's microabcesses (Figure 3).

Full blood count revealed leucocytosis (67×10^3/μL) lymphocytosis (44%). 8% were eosinophils.

In the blood picture atypical lymphocytes were noted.

Bone marrow biopsy revealed increased number of atypical cell infiltration.

Patient's LDH level was in the normal range while ultra sound of the thorax and abdomen were normal.

Therefore patient was diagnosed ultimately as folliculotrophic mycosis fungoides with bone marrow infiltration thus stage IV disease.

Patient was treated with CHOP chemotherapy regimen which includes cyclophosphamide, adriamicin, vincristine and prednisolone.

Initial response was dramatic with reduction of the skin induration (Figure 2).

Later following 4 cycles of chemotherapy patient developed severe skin sepsis with multiple abscesses and within few days he passed away.

Discussion

Folliculotrophic mycosis fungoides (FMF) is a rare variant of cutaneous T-cell lymphoma in which the neoplastic T lymphocytes have affinity for the follicular epithelium. Atypical T cells invade hair follicular epithelium leading to selective or predominantly follicular involvement. FMF has been classified as a distinct variant of CTCL in the new World Health Organization—European Organization for Research and Treatment of Cancer classification system for cutaneous lymphomas.

[1]Senior Registrar in Dermatology, [2]Consultant Dermatologist, [3]Consultant Pathologist, Teaching Hospital Kandy, Sri Lanka.

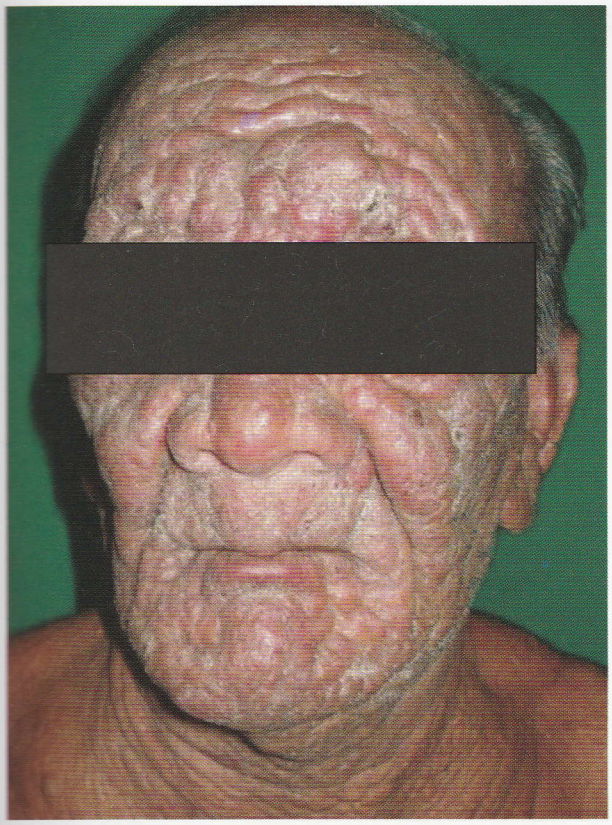

Figure 1. *Leonine face with infiltrated skin.*

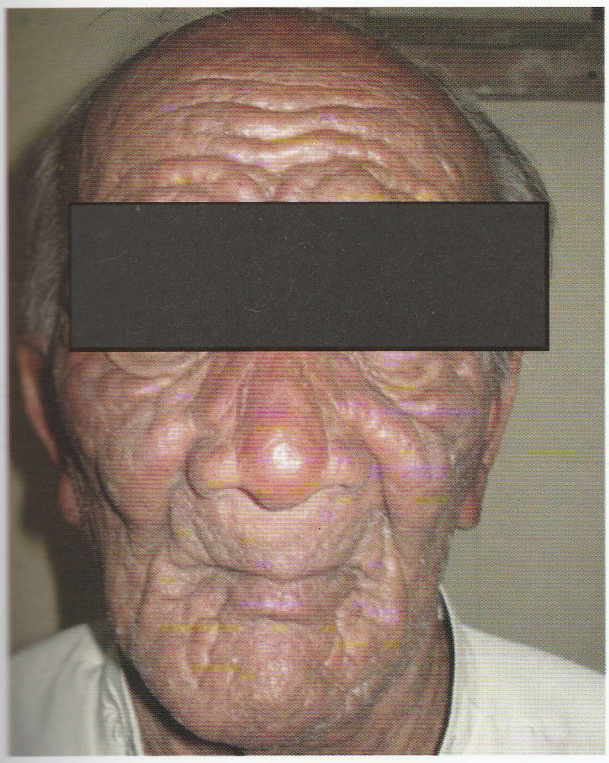

Figure 2. *Following chemotherapy.*

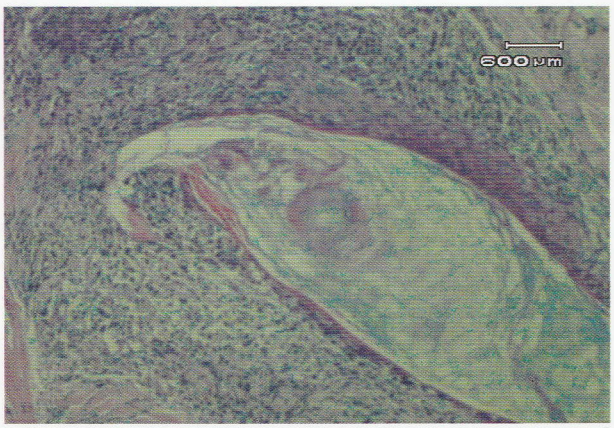

Figure 3. *Infiltration of hair follicle with atypical lymphocytes.*

Follicular papules and plaques with a predilection for the face and scalp is the commonest presentation of FMF[1,2,3] whereas in classical MF these areas are generally spared[4]. Other uncommon presentations are cystic lesions, psoriasiform plaques and leonine face[2,3]. Skin lesions are often associated with alopecia. Our patient who presented with the leonine face was first treated for lepromatous leprosy. As leprosy being the most common clinical entity we encounter with a similar clinical presentation in this part of the world, high degree of clinical suspicion and other unusual features like pruritus are useful clues to arrive at a diagnosis of FMF. Apart from skin biopsy, positive immunohistochemistry with CD3, CD4 and negative CD8 make folliculotrophic lymphoma more likely. T cell receptor (TCR) gene rearrangement of peripheral blood lymphocytes will also support the diagnosis. Furthermore, positive slit skin smear for AFB and a Grenz zone separating epidermis from dermis with dense infiltration of foamy macropharges in the skin biopsy confirm the diagnosis of lepromatous leprosy. Naked granulomas with asteroid bodies or Schaumann bodies would support the diagnosis of sarcoidosis.

Pruritus is a common symptom among FMF patients. This in turn, results in xerosis, excoriations and prurigonodularis like lesions. This can get aggravated by superinfection by *Staphylococcus aureus*. A study done by the Northwestern Cutaneous Lymphoma Group showed 68% FMF patients had pruritus[3]. Successful treatment of lymphoma has subsided this symptom. Although there is a marked difference in outcome of early MF and FMF patients (Stage IIA above) advanced disease has more homogenous course[2].

Our patient who had bone marrow infiltration at presentation with advanced disease (stage IV)

underwent chemotherapy. Despite good initial response patient later developed skin sepsis with multiple abscesses, most probably due to the immunosuppression caused by the disease as well as by the therapy. On the other hand, skin infection is known to exacerbate mycosis fungoides[7]. In the presence of *Staphylococcus aureus* superantigens, nonmalignant T lymphocytes stimulate the proliferation of malignant T lymphocytes by molecular cross-talk[8] This emphasize the importance of skin barrier protection with prophylactic topical antibiotic application to prevent superficial skin infections in patients who undergo chemotherapy according to the NCCN (National Comprehensive Cancer Network) guidelines.

References

1. Cristina M, Teresa E, Ramon MP, Fernando G, Pilar G, Climent J, Octavio S. Folliculotropic mycosis fungoides: Clinicopathological features and outcome in a series of 20 cases. *J Am Acad Dermatol* 2010; **62**: 418-26.
2. Doorn RV, Scheffer E, Willemze R. Follicular Mycosis Fungoides, a distinct disease entity with or without associated follicular mucinosis, *Arch Dermatol* 2002; **138**: 191-8.
3. Pedram G, Steve R, Timothy K, Susan LB, Joan G, Folliculotropic Mycosis Fungoides: An Aggressive Variant of Cutaneous T-Cell Lymphoma. *Arch Dermatol* 2008; **144**: 738-46.
4. Panagiota M, Per H, Gjersvik P, Bassarova A, Ole PF. Clausen, Jan D. Clinical and Histopathological Features of Folliculotropic Mycosis Fungoides. A Norwegian Patient Series. *ActaDermVenereol* 2012; **92**: 325-9.
5. Lehman JS, Robert HC, Brent RW, Roger HW, Lawrence EG, Weaver AL Mark RP, Folliculotropic Mycosis Fungoides: Single-Center Study and Systematic Review. *Arch Dermatol* 2010; **146**: 607-13.
6. Nashan D, Faulhaber D, Sta¨nder S, Luger TA, Stadler R Mycosis fungoides: a dermatological masquerader. *British Journal of Dermatology* 2007; **156**: 1-10.
7. Ziemer MB. A eosinophilic pustular folliculitis in infancy: not a distinctive inflammatory disease of the skin. *Am J Dermatopathol* 2005; **27**: 443-55.
8. Shin J, Monti S, Aires DJ, *et al.* Lesional gene expression profiling in cutaneous T-cell lymphoma reveals natural clusters associated with disease outcome. *Blood* 2007; **110**: 3015-27.