Mycosis fungoides in a Sri Lankan context

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Introduction

Cutaneous lymphomas comprise of T and B cell lymphomas with majority being T cell types. Mycosis fungoides (MF) is the commonest cutaneous T cell lymphoma which usually has an indolent course. A variety of topical and systemic treatment modalities are being used to treat MF with variable success rates. PUVA therapy is one of the commonly used treatment modalities where the effect is known to be stage dependant. Since most available data on this topic are from studies done on Caucasian patients with very limited information on Sri Lankan patients we identified this as an area worthy to do a study. Since the effects of UV radiation varies considerably according to the skin type a very important purpose of our study was to evaluate the effects of PUVA therapy on Sri Lankan patients with MF.

Objectives

1. To study the demographic data of patients with cutaneous lymphoma in order to identify the local trends of the disease.

2. To identify disease characteristics among mycosis fungoides patients such as symptoms and duration of illness at presentation.

3. To assess the effectiveness of PUVA therapy in inducing remission in Sri Lankan patients with mycosis fungoides with special attention to PUVA (SOL) [solar exposure].

Method

This is an ongoing study being conducted at the Dermatology Unit, Teaching Hospital, Kandy. All patients diagnosed with cutaneous lymphoma from 2005 January to December 2008 were included in the study. After obtaining informed written consent all relevant data were recorded using an interviewer administered questionnaire and the monthly clinic records. Initial evaluation was done with a comprehensive history and examination. Full blood count with a blood picture, metabolic panel and a CXR. Clinically suspicious lymph nodes were biopsied and imaging studies and bone marrow biopsies were performed when indicated. All patients were staged according to the North American staging system.

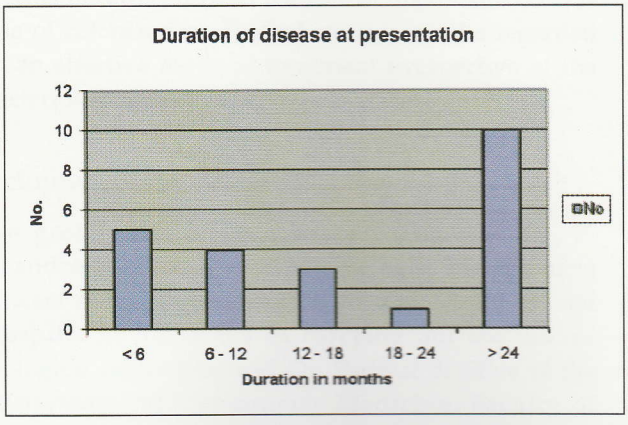
Skin biopsies for histology (and immunocytology when necessary) were performed at the onset of treatment and at 6 monthly intervals. Patients were followed up for a mean period of 22 months (3 months-48 months) with monthly evaluations. PUVA therapy was given twice a week starting at 0.5 J/cm-2 with 8 methoxsalen 0.06 mg/kg-1 and PUVA (SOL) was used for patients who had practical difficulties in coming to the hospital frequently. Remission was defined as histological clearance of the lesions while relapse was defined as appearance of new lesions or reactivation of previous lesions with histological features of the disease. Data were analyzed using SPSS statistical package.

Results

23 patients with cutaneous lymphoma were diagnosed during the 4 year study period and all were included in the study. Out of the study population 69.56% (16) were males and 30.43% (07) were females. Mean age of the study group was 49.60 years (range 9-80 yrs).

21.73% of the population (i.e. 5 patients) had pre existing skin disease out of which 80% had endogenous eczema. Results of the duration of disease at presentation are given in Graph 1.

Graph 1



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Reason for their presentation to the clinic was analyzed which showed symptoms of the disease as being the commonest 52.17% (n=12), cosmetic in 43.47% (n=10) and a medical referral in 01 patient. Out of the symptomatic patients pruritus was the commonest symptom present in 75% (n=9) followed by ulceration in 16% (n=2) and scaling in 33% (n=1).

Type of lymphoma diagnosed after histological analysis of skin biopsies is given in Table 1.

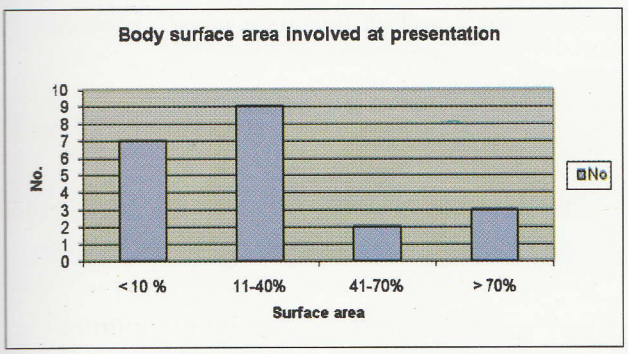
Table 1. Type of lymphoma

Type of lymphoma	*No.*
Mycosis fungoides	18
Mycosis fungoides variants	
Folliculotrophic MF	01
Polkiloderma atrophicans vasculare	02
B cell lymphoma	01
Large cell anaplastic lymphoma	01

Of the 21 patients with mycosis fungoides (MF) 09 patients (42.85%) had patch stage lesions, 08 plaque stage (38.09%), 03 tumour stage (14.28%) and 01 (4.76%) patient with erythrodermic skin.

Results of the analysis of area of body surface involvement at presentation among the MF patients are shown in Graph 2.

Graph 2



Final staging of the MF patients at presentation is given in Table 02.

Table 2. Staging of the patients with MF

Stage	*No.*	*%*
Ia	06	28.57
Ib	12	57.14
IIa	-	-
IIb	02	9.52
III	01	4.76
IV	-	-

15 of the 23 patients were managed in the dermatology unit alone with PUVA therapy while 06 patients needed combined management with the oncology unit with various combinations of cytotoxic drugs, interferones and local radiotherapy (excluding the 02 non MF lymphoma patients).

Out of the 15 MF patients treated with PUVA therapy alone 09 (60%) achieved remission. The number of PUVA sessions each of them had received at the time of their first remission is given in Table 3.

Table 3. Number of PUVA sessions received by each patient at the time of first remission

Patient no.	*PUVA sessions*	*PUVA(SOL)*
1	30	-
2	43	56
3	115	-
4	-	50
5	50	25
6	46	-
7	33	-
8	38	-
9	186	-

Out of the 09 patients with remissions 04 had relapses during the follow up period. Time taken to relapse after remission showed a mean of 10 months (range 6 months to 1 year). 03 out of these 04 patients relapsed while being maintained on a once a week PUVA dose. Out of the 06 non responders 03 had defaulted treatment and the other 03 were only treated for a short period at the time of data collection.

Discussion

Cutaneous T cell lymphoma (CTCL) is not as rare as commonly believed among Sri Lankan patients since 23 patients were diagnosed within the 04 years study period.

A male predominance with 69.56% males was noted which was compatible with other published studies[2]. Majority of the patients (52.17%) were middle aged (between 41-70 yrs) as described in most published studies[2].

17.38% of the study population (n=4) had pre existing endogenous eczema but further studies will be necessary to identify whether there is an actual association between the 02 conditions.

Average disease duration at the time of presentation had a mean of 42.34 months (range 2 months-20 yrs) compatible with a study done in USA which showed an average of 4 yrs at presentation[1].

It was interesting to note that a large proportion of patients (i.e. 43.47%) had presented due to the cosmetic effects the disease has produced while 52.17% had presented with symptomatic disease and only one patient had been referred by a general practitioner (GP) giving a clue towards the low level of awareness among GPs regarding cutaneous lymphoma. Of the patients who presented with symptoms pruritus was found to be the commonest (i.e. 75%) a feature described in a study[2] done in USA.

In keeping with published data[3,4] mycosis fungoides was found to be the commonest lymphoma among our study population (91.30%). Majority of the MF patients had early disease with 85.17% patients having stage I disease (28.57% with Ia and 57.14% with Ib). Non of the stage I patients progressed to advanced stages during the follow up period. Of the fifteen MF patients with stage Ia and Ib disease who were treated with PUVA therapy alone, 60% (n=9) went in to remission which is quite a satisfactory value especially considering that 06 non respondents were either defaulters (n=03) or patients who have received treatment for a few months only at the time of date collection (n=03). A considerable proportion of patients (i.e. 55%) had achieved remission with 30-50 PUVA sessions. Although some patients had required as much as 186 PUVA sessions there was no significant correlation between the BSA involved and the number of PUVA sessions needed.

An interesting observation was that PUVA (SOL) also appears to be effective to some degree in controlling MF as one patient with stage Ib disease who was treated with PUVA (SOL) alone had achieved remission after only 50 sessions. This observation needs further investigations and might be a promising option to our patients as PUVA facility is only available in 2 centres in Sri Lanka.

Although the rate of remission was satisfactory remissions were not sustained (mean 9.5 months) and 44.44% patients had gone into relapses during the follow up period. The fact that maintenance PUVA therapy is not effective in sustaining remission was reaffirmed by our study as 3 of these patients (i.e. 75%) had relapses while being on a once a week maintenance regimen. Further studies with larger number of patients with longer follow up periods are needed to evaluate the beneficial effects of maintenance PUVA therapy over risks of long term phototherapy in Sri Lankan patients.

References

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