CASE SERIES

Common Presentations of Mycosis Fungoides in Pakistani Population: A Clinicopathological Study

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ABSTRACT

Objective: To study the different clinical, pathological and immunophenotypic features in patients of Mycosis fungoides presenting to Civil Hospital Karachi.
Methods: A case series consisting of 10 patients was conducted at the Dermatology department of Civil Hospital Karachi, from January 2013 to December 2016.A Performa was used after written consent from the patients and the head of the department to record the history, physical examination and laboratory findings.
Results: Out of the 10 patients, 3 had stage IIB, 3 had stage III, 1 patient evolved from stage III to IV and 3 had stage IV disease, 2 patients were in erythrodermic stage, while 1 evolved from the erythrodermic stage to the tumor stage. 3 patients showed psoriasiform appearance, 3 showed tumor d emblee stage, while only 1 patient revealed poikilodermatous appearance. Male to female ratio was 5:1. Histopathological and immunophenotypic markers were consistent with those of mycosis fungoides. Five patients were previously treated as eczema or psoriases.
Conclusion: Knowledge about the varied presentations of mycosis fungoides would lead to better understanding, and help us with the diagnostic challenges in our population

Key words: Mycosis fungoides, Cutaneous T cell Lymphoma, Sezary syndrome, Erythroderma, Pautrier's microabscess.

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INTRODUCTION

Mycosis Fungoides is the most common variant of primary cutaneous T-cell lymphoma, that is primarily a tumor of skin [1]. Sezary syndrome is the aggressive form of mycosis fungoides. It has an indolent clinical course and males are affected twice as commonly as females [2]. The clinical manifestations of mycosis fungoides is variable [3] with most of the cases being treated as eczema, psoriasis and other dermatosis for months or years by the general practitioners before they develop the typical lesions or are referred to a consultant dermatologist for further evaluation and management [4]. It is seen in all skin types; however, the presentation is different in dark-colored skin as compared to the white population. Here, we are presenting the various clinical manifestations and pathological findings exhibited in 10 patients from Karachi, Pakistan. All of these patients presented to the Department of Dermatology, Civil Hospital Karachi from Jan 2013 to Dec 2016. Out of the 10 patients, 5 were initially treated as eczema or psoriasis for a couple of years and later on referred with the suspicion of MF. Hence, accurate diagnosis of this condition remains challenging and depends on thorough clinical examination by the experts and confirmed by histological and immunohistochemistry(IHC) findings[6].

Data regarding the status of this condition in Pakistan is insufficient. To date, there is no detailed clinicopathological study or published literature on MF in adults from Pakistan. The following study will help in enhancing our knowledge regarding the variable clinical and histopathological presentations of MF in our population, hence improving the diagnostic and

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management challenges.

METHODS

A total of 10 cases are being reported in this study conducted at the Department of Dermatology, Civil Hospital Karachi, Pakistan, from January 2013 to December 2016. After collecting the detailed history, patients were thoroughly examined by the dermatologist and internist, investigated with full blood count, biochemical testing, peripheral smear, x-ray chest, ultrasound abdomen and computed tomographic scans of the chest and abdomen. A 4mm punch biopsy of the skin was carried out on all the patients after taking an informed consent. Skin sections were examined for histopathology and immunohistochemistry. Lymph node biopsy was performed where required. Staging was carried out according to the TNM classification [7]. The demographic features, clinical presentations and pathological changes were assessed.

RESULTS

Clinical Features:
A total of 10 patients were included in this case series. Table 1 summarizes the demographic and clinical characteristics of the patients.

Table 1. Clinical and demographic characteristics of patients with mycosis fungoides (MF)

PATIENT	AGE ATONSET /DIAGNOSIS	GENDER	INITIAL DIAGNOSIS	CLINICAL FEATURES	STAGE AT DIAGNOSIS
1	50/75	M	Psoriases	Erythematous scaly plaques	IIb
2	30/42	M	Psoriases	Erythematous scaly plaques, some horse-shoe shaped. Palpable axillary lymph nodes.	IIb
3	52/54	F	Psoriases	Annular and horseshoe shaped plaques	IIb
4	20/32	M	MF	Erythrodermic to tumor stage	III evolved to IV
5	55/74	M	MF	Erythroderma and palmoplantar hyperkeratosis	III
6	27/35	M	Psoriases	Erythroderma	III
7	62/80	F	MF	poikiloderma, acquired icthyiosis, alopecia, madarosis	III
8	45/55	M	Eczema	Nodulo-ulcerated lesions. Tumor d emblee.Palpable axillary lymph nodes.	IV
9	50/50	M	Cutaneous Leshmaniases	Nodulo-ulcerated lesions. Tumor d emblee.Axillary lymph nodes palpable	IV
10	50/70	M	MF	Multiple nodules and popular eruptions. Tumor d emblee., Axillary lymph nodes palpable	IV

Disease onset was in the fifth decade in 7/10 patients. The duration between the onset of the disease and confirmed diagnosis was more than a decade (in 8 out 10 patients).
Five patients had lymph node involvement. Four had palpable axillary lymphadenopathy while patient seven had subcentimeter thoracolumber lymph nodes, confirmed on radiological imaging.

Histopathology and Immunohistochemistry: Biopsy results of all of the patients showed common findings with a few variations.

Common features included a dense dermal lymphocytic infiltrate with sheets of atypical lymphoid cells showing irregular hyperchromatic to vesicular, convoluted or cerebriform nuclei. Epidermotropism was seen in five patients, whereas classical Pautrier's microabscess was seen in none of the patients. TCR gene analysis could not be done due to non-availability. Table2 reveals the immunophenotypic characteristics.

Table 2. Immunophenotypic of patient lesions

PATIENT NO:	CD3	CD5	CD45RO	CD20	CD79a	EPIDERMOTROPISM
1	Diffusely +	+	-	-	-	NO
2	Diffusely +	-	-	-	-	YES
3	Diffusely +	-	-	-	-	YES
4	Diffusely +	Diffusely +	-	-	-	YES
5	Diffusely+	Diffusely +	-	Diffusely +	Focally +	NO
6	Diffusely +	+	+	+	+	YES
7	Diffusely +	+	-	+ in background cells	-	NO
8	Diffusely +	-	-	-	-	YES
9	Diffusely +	-	+	-	-	NO
10	Diffusely +	Diffusely +	Diffusely +	+ in background cells	+ in background cells	NO

DISCUSSION

The mean age of patients at the time of diagnosis included in our study was 55.7 years. This was similar to the study of 552 patients of Primary Cutaneous Lymphomas by Regina et al[8]. In a large cohort of study of 1502 patients of MF/SS, the mean ages of the study population was 54 years[9]. There was male preponderance of 5:1 in our study which was close to the gender predisposition seen in a study conducted at Singapore of 131 patients with mycosis fungoides[10]. MF is a slowly progressing tumor. In the majority of our patients (8 out of 10), the duration from onset of disease was about 12 to 20 years. This was similar to the study done by Dummer[11] in 2002 in which the duration between onset and diagnosis was also long. Most of the patients>80% presented at an older age and <20% in the younger age group. If we consider distant metastasis, 5 of our patients presented with

lymph node involvement, however, other organ involvement was not present in any patient. Other relevant investigations including complete blood count,liver and renal function test, ultrasonographic studies and computed tomographic scans were within the normal range.

Histopathological examination in our patients showed characteristic features of mycosis fungoides, though the classic Pautrier microabsecess was not seen in any of the patients. This was also the case in a study of MF- associated follicular mucinosis in young patients by Brian J et al [12].

Immunohistochemistry results were also consistent with those of mycosis fungoides.

In conclusion, the diagnosis of MF can pose great difficulty as its clinical presentation not only varies according to the stage but also mimics a number of other common dermatological conditions. The knowledge of this rare disease entity and its variable presentation is crucial especially in developing countries like ours where delay in referral and timely diagnosis commonly adds to the patient's morbidity.

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