

Mycosis Fungoides at a Moroccan Referral Hospital: A Retrospective Study of 33 Cases

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Abstract

Background: Mycosis fungoides (MF) is the most frequent cutaneous T cell lymphoma with slow disease progression. The aim of this study was to describe demographics, clinical, treatment outcomes and progression in patients with MF within Moroccan population.

Materials and Methods: A retrospective descriptive study conducted on MF patients admitted to the Military hospital of instruction Mohamed V Rabat Morocco. We included 33 patients. Data were collected from medical records.

Results: Thirty-three patients with clinically and histopathologically confirmed MF were enrolled. Median age at initial diagnosis was 57years+/- 19,6 with a with a male to female ratio of 1,75. The majority of patients (60%, n=20) had limited-stage (IA-IB) disease at the time of diagnosis. Disease progression from early stages of MF (IA-IB) to advanced stages (II-IV) was 21%. 46% patients received topical treatment. Overall mortality was 15%

Keywords: *Mycosis fungoide; Progression; Mortality*

1. Introduction

Mycosis fungoides (MF) is the most frequent cutaneous T-cell lymphoma, representing nearly 50% of all primary cutaneous lymphoma [1]. It is an extranodal non-Hodgkin lymphoma that is presented initially with erythematous skin patches and plaques that may progress to tumors with extracutaneous involvement [2]. Even if MF is considered as an indolent disease, it may lend a higher mortality in advanced stage [3]. Several treatments have been proposed in terms of stages of MF but it is still considered as an incurable disease. This retrospective study aims to study the demographics, clinical, treatment outcomes and progression in patients with MF within Moroccan population.

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2. Materials and Methods

We retrospectively conducted a chart review of MF patients that were treated in our Military hospital of instruction Mohamed V Rabat Morocco from March 2012 to January 2020. Data collected from medical records included: Age at diagnosis, gender, diagnosis, stages, comorbidities, treatments, mortality probability. We included in this study all the patients with confirmed diagnoses of MF. We excluded patients without available medical records for clinical courses and relevant laboratory data. Data collection and statistical analysis were done using JAMOVI 2.2.5 current.

3. Results

From March 2012 to January 2020, we enrolled 33 MF patients. Of the thirty-four cases, 63% were male and 37% were female, with a male to female ratio of 1,75. The median age at diagnosis was 57 years+/- 19,6. The average age of male was 60 years+/- 17,9 and for female 51,7 years +/-22. The median time from onset of symptoms until established diagnosis was 53mois [4].

Location of the lesions was variable: Trunk (47%), Inferior member (33%), Upper limb (31%), Disseminated (18%), Face (10%), Neck (5%). At the time of diagnosis, 20 patients (60%) had limited stage of MF. 42% were in stage IA and 18% were in stage IB. 13 patients (40%) with advanced stage. 27% were in stage II, 10% were in stage IIIA and 3% were in stage IVA. Among of the thirty-four cases, 26 patients (78%) had classic MF, 3 patients (9%) had folliculotropic MF, 3 patients (9%) had hypopigmented MF and 1 patient (3%) had poikilodermatous MF.

Out of the 44 patients with MF, 6 patients (18%) had an increased LDH level, 8 patients (24%) had hypereosinophilia, 3 patients (9%) had hyperlymphocytosis and 9 patients (27%) had an increased sedimentation rate. The histopathological pattern comprised immunohistochemically confirmed CD3+/CD4+ positive cells, with visual atypia and epidermotropism. True Pautrier's abscesses were seen in 75% (n=25) of cases. Disease progression from early stages of MF (IA-IB) to advanced stages (II-IV) was 21% (FIG. 1). 15 patients (46%) were treated with topical corticosteroids only or in combination with phototherapy in 6 patients (18%). Seven patients (21%) were treated with méthotrexate, 2 patients (6%) with retinoid and 3 patients (9%) with chemotherapy. 67% were still under follow up and Overall mortality was 15% (TABLE 1).

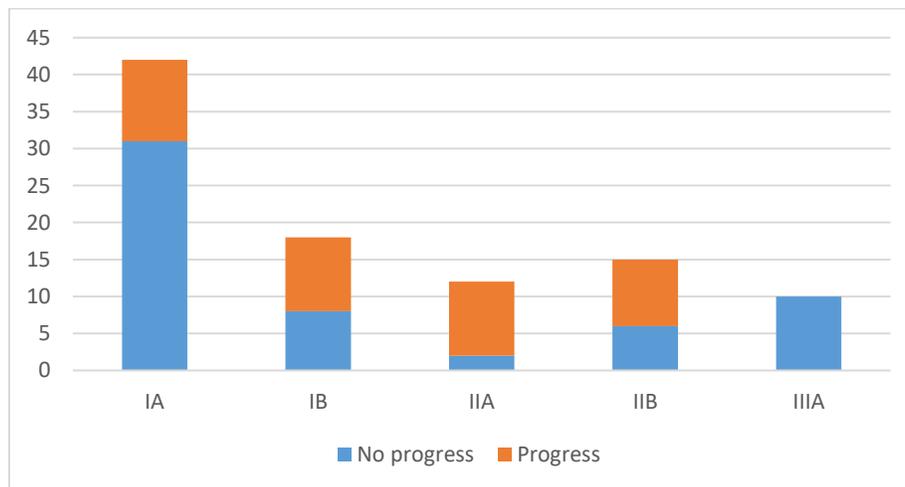


FIG. 1. Staging at initial diagnosis of mycosis fungoides and progression.

TABLE 1. Patient's characteristics (n=33).

Characteristics	Valeurs (N=33)
Age in years, mean (SD)	57 +/- 19,6
Gender	
Male	21(63%)
Females	12(37%)
Clinical phase, n (%)	
Plaque	20(61%)
Tumor	4(12%)
Erythroderma	6(18,8%)
Location, n (%)	
Face	3(10%)
Neck	1(5%)
Trunk	15(47%)
Upper limb	10(31%)
Lower limb	11(33%)
Disseminated form	6(18%)
Extracutaneous involvement, n (%)	
Mucosal involvement	3(8%)
Nail involvement	6(18%)
Visceral involvement	4(11%)
Types of mycosis fongoide, n (%)	
Classical MF	26(78%)
Folliculotropic MF	3(9%)
Hypopigmented MF	3(9%)
Poikilodermatous MF	1(3%)
TNM Stage, n (%)	
IA	14(42%)
IB	6(18%)
IIA	4(12%)
IIB	5(15%)
IIIA	3(10%)
IVA	1(3%)
Treatment, n (%)	
Topical steroid	15(46%)
Methotrexate	7(21%)
Phototherapy	6(18%)
Acitretin	2(6%)
Chemotherapy	3(9%)
Evolution, n (%)	
Alive	22(67%)
Deceased	5(15%)
Out of sight	6(18%)

4. Discussion

MF is an extranodal non-Hodgkin lymphoma and constitutes about 50% of the cutaneous lymphomas [1]. Median age of diagnosis is 55 to 60 years, but it can also be seen in young adults and children [4]. In line with the literature, the mean age in our study was 57years+/- 19,6 and males outnumbered the females. Several hypotheses have been postulated for the

pathogenesis of mycosis fungoides [5,6]. The clinical presentation of MF includes patches, plaques, or tumors [1]. They seat mostly on non-sun exposed areas, but any other area of the skin can be affected [7].

The prognosis of MF depends on the stage, favorable on early stage and worse on advanced stage [8]. In our study, the majority of our patients (60%) were in the early stage, and 40% were in an advanced stage. The progression of MF from patches to tumor may take several years or decade [1]. 21% of our patients progress from early stages to advanced stages.

Multiple risk factors for MF have been demonstrated including demographics, stage and the presence of extracutaneous involvement. Knowledge of this can refine the prognostic workup of patients with higher risk of disease progression. The diagnosis is often difficult to establish on the early stages, therefore the diagnosis is suspected clinically and confirmed by skin biopsy.

The typical histopathological features leading to the diagnosis in early stages of MF include epidermotropism, dermal fibrosis and atypia of dermal lymphocytes, Pautrier's microabscesses, basal alignment of neoplastic lymphocytes [9]. Treatment of MF should follow stage-adapted approach based on the recommendation of the European Organisation for Research and Treatment of Cancer-Cutaneous Lymphoma Task Force (EORTC-CLTF).

In our study, the early-stage MF patients were mainly treated by dermocorticoids only or combined with phototherapy. It has been reported in several studies, that MF patients have an increased mortality within the first 5 years of follow-up [10]. The overall mortality in our study was 15%.

5. Conclusion

MF is an incurable disease with an increased risk of mortality. Despite advances in Dermatology, treatment is often palliative of most patients despite the achievement of major symptomatic improvement.

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