OPEN ACCESS
Saudi Journal of Pathology and Microbiology
Abbreviated Key Title: Saudi J Pathol Microbiol
ISSN 2518-3362 (Print) |ISSN 2518-3370 (Online)
Scholars Middle East Publishers, Dubai, United Arab Emirates
Journal homepage: https://saudijournals.com

Original Research Article

Mycosis Fungoides in UAE

Hamodat Mowafak M[1*], Al Maashari Raghda S[2], Al Zaabi Eiman[3]

[1]Consultantdermatopathologist. Pathology and Laboratory Medicine Department, Sheikh Shakbout Medical City (SSMC), Abu Dhabi, U.A.E
[2]Dermatology Consultant, Dermatology Department, Sheikh Khalifa medical city (SKMC), Abu Dhabi, U.A.E
[3]Chair of Pathology and Laboratory Medicine Department, Sheikh Shakbout Medical City (SSMC), Abu Dhabi, U.A.E

DOI: 10.36348/sjpm.2021.v06i07.006 | **Received:** 12.06.2021 | **Accepted:** 23.07.2021 | **Published:** 27.07.2021

***Corresponding author:** Hamodat Mowafak M

INTRODUCTION

Primary cutaneous lymphomas (PCL) are defined as extranodal non-Hodgkin Lymphomas [1]. Among the cutaneous lymphomas, nearly two-thirds are of the T cell type with Mycosis Fungoides (MF) being the most common T cell lymphoma (CTCL) [1]. We will be reviewing the incidence, risk factors, prognostic factors, radiology findings, staging and treatment of the disease. To our knowledge this is the first study of its kind to be conducted in the United Arab Emirates (UAE).

METHOD

We conducted a retrospective review of the electronic medical records for all patients documented to have biopsy proven mycosis fungoides following between 2013 -2019 in Sheikh Khalifa Medical City (SKMC) one of the health care facilities operated by Abu Dhabi Heath Care Company (SEHA) in the Emirate of Abu Dhabi. An IRB (Institution Research Board) approval been obtained by SKMC.

A total number of 40 patients found, 22 were female and 18 were male. Their ages ranged between 18-76 years. All were UAE nationals except for three patients.

Hematoxylin and eosin (H & E) biopsies were reviewed by a dermato-pathologist with several cases requiring further immune-histochemical stains to reach the final diagnosis. Stains used included CD 3, CD 4, CD 7, CD 8, CD 30.

Biopsy results were classified accordingly into: patch, plaque, tumor stage, large cell transformation or Sezary syndrome.

Additional data from the charts were recorded by a dermatologist. This included patient's clinical and demographic information such as age, gender, nationality, time of diagnosis, clinical presentation, radiology findings, staging and treatment.

RESULT

Out of the 40 patients included in this study, 22 were female (55%) and 18 were male (45%). Age range affected by mycosis fungoides in our population fell down between 60-69 years (25%), 50-59 years (20%), 40-49 years (20%) and (5%) less than 20 years old. The median age at diagnosis is 47 years (age range between 18-76). Three patients were non-UAE nationals while the remaining were UAE nationals (92.5%).

With regards to the histological diagnosis, majority of cases were patch stage mycosis fungoides 42.5% followed by Folliculotropic and Large transforming variant.

Clinically majority of patient population 17 patients (42.5%) presented with erythematous eczematous patches at the time of diagnosis. 8 patients (20%) with hypo pigmented patches, 5 patients (12.5%) with hyper-pigmented patches, polikodermatous and erythroderma (10%) for each, and (5%) with nonspecific presentations.

According to the TNM classification, 45% of our patient population presented with stage 1A.

Topical potent steroids and phototherapy (NB-UVB) were the most commonly used therapies, other treatment options included systemic methotrexate, acitretin and chemotherapy.

DISCUSSION

Mycosis fungoides (MF), is the most common cutaneous lymphoma. It is a low-grade CTCL characterized by skin-homing CD4+ T cells [2]. Mycosis fungoides has an incidence of around 6 cases per million per year in Europe and the United States [1].

The median age in our study is 47 years, which is comparable to Middle Eastern countries: Jordan (45 years) [3], Saudi Arabia (41 years) [4] and Kuwait (35.20 ± 14.37 years) [5].Moreover, hypopigmented variant was the commonest M.F subtype in the younger age group in Kuwait [5] and Israel [6] with a mean age at diagnosis of (27.60±12.42 years) and (36 years) respectively. Figure-1 illustrates the number of patients in each age group, in our population.

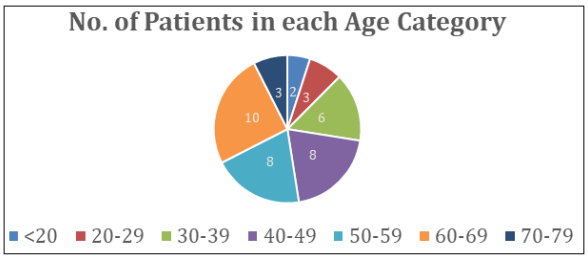

Figure-1: Number of patients in each age group, in our population.

With regards to the histological diagnosis, majority of cases were patch stage mycosis fungoides followed by Folliculotropic and Large transforming variant as shown in Table-1.

Table 1: Histological stage at time of diagnosis

Diagnosis	No. Of Patients
Patch stage	27
Folliculotropic	3
Large cell transforming	3
MF+ Follicular Mucinosis	1
Tumour stage	1
Plaque	1
After phototherapy	1
Lichenoid	1
L.N – Extra cutaneous	2

Upon reviewing consults, the final diagnosis of five cases were changed. One was excluded from the study after the diagnosis was changed from Mycosis Fungoides to Vitiligo. The other four, were initially diagnosed as spongiotic dermatitis, lichenoid dermatitis, Pityriasis Lichenoides Chronica, and Drug reaction, however, the final diagnosis was Mycosis Fungoides.

Time to diagnosis varied, with the longest duration being 192 months (16 years), and the shortest was 4 months, few cases had no specified time of diagnosis from the chart. Time to diagnosis for the majority of patients ranged between 60- 180 months (5-15) years. This is shown in Table-2. The long duration reflects the indolent nature of the disease in those patients.

Table 2: Months elapsed to receive the diagnosis

No. of Months	No. of Patients
192	1
180	4
120	4
84	4
72	1
60	4
48	3
36	2
24	2
18	1
12	2
6	1
4	1
NA	10

17 patients (42.5%) presented with erythematous eczematous patches, 8 patients (20%) with hypo pigmented patches, 5 patients (12.5%) with hyper-pigmented patches, polikodermatous and erythroderma (10%) for each, and (5%) with nonspecific presentations. In comparison with a study from Kuwait, 22% of patients presented with hypo pigmented M. F [5]; given hypo pigmented variant was the second commonest presentation.

In view of staging, the majority of our patient population presented with stage 1A (45%). This is depicted in Table-3.

Table 3: Stage of disease

Stage of Disease	No. Of Patients
IA	18
ND	1
IB	3
IIB	1
IIIA	1
IIIB	3
Patch transformed to large	2
Tumour transformed to large	3
Stage identification unknown	4
Stage identification not complete	4

More than half of the patients had within normal radiological findings, while one quarter of them showed lymphadenopathy. Of those, 3% evolved to Sezary syndrome, 3% evolved to large transforming lymphoma, and 2% evolved to Folliculotropic M.F and patch stage 1B.

Treatment options ranged from topic steroids, phototherapy, systemic methotrexate and acitretin to chemotherapy.

19 patients lost follow up as they completed their treatment outside the country. 22.5% of patients had complete resolution of their lesions, 20% made an excellent response where lesions subsided gradually and 10% are still under active treatment.

(Quaglino *et al*., 2017) [7], 853 patient treated in different centers in US, Australia, Japan, Europe and Brazil, receiving more than 24 different treatment modalities, found that treatment type did not lead to statistically impactful survival when compared US and non US centers.

Poor prognostic factors in an American study (Su *et al*., 2017) [8], included males, old age, African American race, government insurance, higher stage of disease and history of receiving chemotherapy or radiotherapy.

CCR3 and CCR4 expression suggests poor prognosis as shown by (Shono *et al*., 2019) [9]. Moreover, early stage at presentation had a better prognosis with a survival of 10-35 years as shown by (Scarisbrick *et al*., 2018) [10]. Of those, 25% of patients might progress to advanced disease with less than 4 years survival. The same study showed that patients presenting with nodal involvement had a median survival of 13 months, while those with plaque stage (IIB) folliculotropic M.F might have worse prognosis than those with tumor stage (IIB). Furthermore, those with tumor stage (IIB) have worse prognosis than those with erythrodermic stage (IIII).

On another account, a study by (Vergier *et al*., 2000) [11], found that transformed M.F cases with patient age at least 60 years with extra cutaneous manifestations are associated with poor prognosis. Age, sex, transformation speed, percentage of large cells or CD30 expression have no prognostic value.

While most of the derm-path textbooks agree that folliculotropic M.F has worse prognosis than a classic M.F, (Giberson *et al*., 2017) [12] showed in a Meta analysis of two studies, no significant statistical difference in survival for patients with folliculotropic and classic M.F.

Source(s) of support: nil

Conflictis of Interest: nil

This manuscript has not been published before.

CONCLUSION

In conclusion, to date, this is the first study of mycosis fungoides conducted in UAE highlighting the incidence, risk factors, prognostic factors, radiology findings, staging and treatment of the disease. Further studies with larger sample size are warranted, to follow up the evolution of the disease and monitor response to newer treatment modalities.

REFERENCES

1. Vaidya, T., & Badri, T. (2020). Mycosis Fungoides. 2020 Aug 26. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2021 Jan. PMID: 30137856.
2. Phyo, Z. H., Shanbhag, S., & Rozati, S. (2020). Update on Biology of Cutaneous T-Cell Lymphoma. Frontiers in Oncology, 10, 765.
3. Awad Hasan Al-Tarawneh, M. D., & Board, J. (2018). Clinical and Histopathological Spectrum of Mycosis Fungoides. Bahrain Medical Bulletin, 40(2), 103-107.
4. Binamer, Y. (2017). Cutaneous T-cell lymphoma in Saudi Arabia: retrospective single-center review. Annals of Saudi medicine, 37(3), 212-215.
5. Alsaleh, Q. A., Nanda, A., Al- Ajmi, H., Al- Sabah, H., Elkashlan, M., Al- Shemmari, S., & Demierre, M. F. (2010). Clinicoepidemiological features of mycosis fungoides in Kuwait, 1991–2006. International Journal of Dermatology, 49, 1393-1398.
6. Lambroza, E., Cohen, S. R., Phelps, R., Lebwohl, M., Braverman, I. M., & DiCostanzo, D. (1995). Hypopigmented variant of mycosis fungoides: demography, histopathology, and treatment of seven cases. Journal of the American Academy of Dermatology, 32(6), 987-993.
7. Quaglino, P., Maule, M., Prince, H. M., Porcu, P., Horwitz, S., Duvic, M., ... & Kim, Y. (2017). Global patterns of care in advanced stage mycosis fungoides/Sezary syndrome: a multicenter retrospective follow-up study from the Cutaneous Lymphoma International Consortium. Annals of Oncology, 28(10), 2517-2525.
8. Su, C., Nguyen, K. A., Bai, H. X., Cao, Y., Tao, Y., Xiao, R., ... & Zhang, G. (2017). Racial disparity in mycosis fungoides: an analysis of 4495 cases from the US National Cancer Database. Journal of the American Academy of Dermatology, 77(3), 497-502.
9. Shono, Y., Suga, H., Kamijo, H., Fujii, H., Oka, T., Miyagaki, T., ... & Sato, S. (2019). Expression of CCR3 and CCR4 suggests a poor prognosis in mycosis fungoides and Sézary syndrome. Acta dermato-venereologica, 99(9-10), 809-812.
10. Scarisbrick, J. J., Quaglino, P., Prince, H. M., Papadavid, E., Hodak, E., Bagot, M., ... & Willemze, R. (2019). The PROCLIPI international registry of early- stage mycosis fungoides identifies substantial diagnostic delay in most patients. British Journal of Dermatology, 181(2), 350-357.
11. Vergier, B., de Muret, A., Beylot-Barry, M., Vaillant, L., Ekouevi, D., Chene, G., ... & Wechsler, J. (2000). Transformation of mycosis fungoides: clinicopathological and prognostic features of 45 cases. Blood, The Journal of the American Society of Hematology, 95(7), 2212-2218.
12. Giberson, M., Mourad, A., & Gniadecki, R. (2017). Folliculotropism does not affect overall survival in mycosis fungoides: results from a single-center cohort and meta-analysis. Dermatology, 233(4), 320-325.