Poikilodermatous Mycosis Fungoides (MF)

Dr. Samia Abu Shaikha
Dr. Hussain Alsada
Prof. Hassan Riad

Summary

Mycosis fungoides (MF) is a lymphoma of low-grade malignancy with prolonged survival. The progression from the clinical stage of patches to those characterized by plaques, tumors, and extra-cutaneous spread usually takes place over many years or decades. One of the problems in describing this disease is that it doesn't look the same for all patients. Patches, plaques and tumors are the clinical names of the different presentations. Patches are usually flat, possibly scaly. Patches are often mistaken for eczema, psoriasis or "non-specific " dermatitis until an exact diagnosis of Mycosis fungoides is made.

We reported an unusual variant of two cases of poikilodermic MF.

Introduction

Mycosis Fungoides (MF) is the most common subtype of cutaneous lymphomas. [1] Clinically, it is characterized by flat, scaly, red, variably pruritic patches to indurated, annular plaques progressing to frank tumors. Microscopically, patch-stage MF exhibits a sparse papillary dermal lymphocytic infiltrate with epidermotropism and atypical lymphocytes, either in collections or singly within the epidermis, and these are probably the most specific findings in early MF. Immunohistochemical studies are essential to confirm the diagnosis. There are many clinical variants including follicular, vesicular, poikilodermic, hypo- and hyperpigmented, ichthyosiform, and pigmented purpura-like lesions. In the following patients, an unusual variant of MF was diagnosed after years of non-remitting disease.

Case report

Case1 (figure 1)

A29 year old female presented with Poikilodermatous skin lesion since childhood. The lesions are bilateral and symmetrical involving mainly flexors and trunk with some lesion showed follicular accentuations on the sun exposed area.

Case 2 (figure 3)

A 20 year old female presented with Poikilodermatous skin lesion since childhood. The lesions are mainly on the trunk and limbs.

Both cases have features in common: since childhood, and both females.

The lesions are bilateral and symmetrical involving mainly on flexor and trunk.

The lesions are ill-defined pigmented macules on background of erythematic taking deferent pattern form wavy and liner. Atrophy and telangatesia are present uniformly among the affected area especially on the lower part of the body instigative hair and nail, lymph nodes few scattered lymph node enlargement.

Immunopheotyping was done for all the biopsy was taken from patients **(CD3+,CD4+,CD7+,CD8+,CD 15+,CD20+,CD30+,LCA,UCHL(CD45+R))**

It showed mainly positive for CD4+, CD8+, UCHL (CD45+R) in both cases and positive CD30+ in the cases 2 and negative for the other in both cases.

Histopathology showed Fig 2&4 H&E section, x40 magnification. The pathologic picture was showing atypical lymphocytes in both dermis and epidermis.

Discussion

Lymphoma accounts for 3.3% of all cancers worldwide. It ranks the 8th most common cancer site for males and 9th for females (figure 5). In Qatar, Non-Hodgkin lymphoma is the 5th most common cancer in male and the 4th in females (figure 6). There is no data about the incidence of cutaneous lymphoma in Qatar until now.[9-11]

MF is a peripheral non-Hodgkin T-cell lymphoma initially presenting in the skin. [2] Clinically, the disease is typified by the gradual progression from patches and plaques mostly on photoprotected sites to tumors. Erythroderma may intervene at any time, and its distinction from Sezary syndrome (SS) depends on the findings in peripheral blood and other clinical features. Unusual clinical and histopathologic variants often coexist with typical patches or plaques of MF. The common denominator among all variants is the presence of diagnostic histologic findings.[3]

Poikilodermic MF is characterized by the presence of hypo- and hyperpigmentation, dryness, atrophy, and telangiectasia (poikiloderma atrophicans vasculare) developing slowly at the sites of preexisting patches, usually in the areas where the skin is chronically

rubbed by clothes and are accompanied by otherwise typical patches and plaques of MF elsewhere.
The initial treatment is stage related. Early aggressive therapy does not improve survival.[4]
The standard treatment for early lesions includes topical steroids, topical chemotherapy, PUVA, narrow-band UV-B, interferonα-2a, or oral retinoids.[5,6] Combination therapies, which may reduce the toxicity of the single agents, are increasingly being used.[7] Systemic chemotherapies and new immunologic agents, (alemtuzumab), and interleukin-12, are used in advanced stages withnodal or visceral involvement.[8]

Conclusion,

We reported two cases of Poikilodermatous CTCL which is a variant type of primary cutaneous T cell lymphoma, it presented in females and started in both patients in childhood. Our point of report is to shed some light on this special sub-entity of mycosis fungoides which affect mainly young female in the pre and post adolescent period.

References

1. *Willemze R, Kerl H, Sterry W, et al. EORTC classification for primary cutaneous lymphomas: a proposal from the Cutaneous Lymphoma Study Group of the European Organization for Research and Treatment of Cancer. Blood. 1997;90:354-371.*
2. *Isaacson PG, Norton AJ. Cutaneous lymphomas. In: Isaacson PG, Norton AJ, eds. Extranodal Lymphomas. London: Churchill Livingstone; 1994:131-191.*
3. *Fung M, Murphy M, Hoss D, Grant-Kels J. Practical evaluation and management of cutaneous lymphoma. J Am Acad Dermatol. 2004;46:325-57.*
4. *Kaye FJ, Bunn PA, Steinberg SM,et al. A randomized trial comparing combination electron-beam radiation and chemotherapy with topical therapy in the initial treatment of mycosis fungoides. N Engl J Med1989;321:1784–1790.*
5. *Kim YH, Martinez G, Varghese A, et al., Topical nitrogenmustard in the management of mycosis fungoides: updateof the Stanford experience.Arch Dermatol 2003;139:165–173.*
6. *Baron ED, Stevens SR. Phototherapy for cutaneous T-cell lymphoma. Dermatol Ther 2003;16: 303–310.*
7. *Duvic M, Lemak NA, Redman JR,et al.Combined modality therapy for cutaneous T-cell lymphoma. J Am AcadDermatol1996;34: 1022–1029.*
8. *Cerroni L, Gatter K, Kerl H, eds.An Illustrated Guide to Skin Lymphoma, 2nd edn. Oxford: Blackwell PublishingLtd, 2004: 31–33.*
9. *Parkin DM, Bray F, Ferlay J, Pisani P. Global cancer statistics, 2002. CA Cancer J Clin. 2005 Mar-Apr;55(2):74-108. 10-El-Attar IA. Cancer databases in the Arab world. Ethn Dis. 2005 Winter;15(1 Suppl 1):S1-3-4.*
11. *Cancer Incidence in Five Continents GLOBOCAN 2002 database: summary table by population http://www-dep.iarc.fr/*