Mycosis fungoides: A report of two cases

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ABSTRACT

Introduction: Mycosis fungoides is one of the rare malignant skin neoplasms also one of the most common primary cutaneous T cell lymphomas. The etiology and risk factors of the disease are poorly realized, nevertheless, in Nigeria, the disease seems to be under reported. Mycosis fungoides resembles many skin lesions, particularly during its early clinical course, creating diagnostic challenges, especially in growing countries as it requires tissue biopsy, histological diagnosis with hematoxylin and eosin and immunohistochemistry. Case Series: Two females of 20 years and 60 years, were diagnosed with mycosis fungoides in our archival (2003–2016). Conclusion: Mycosis fungoides is one of the non-Hodgkin lymphoma that is presented with cutaneous lesions and it has an indolent clinical course. However, the disease was under-diagnosed in our environments. Thus, the clinician should often regard it as unitary of the differential diagnoses of the skin lesion.

Keywords: Diagnosis, Mycosis fungoides, Rare disease, Skin neoplasms

INTRODUCTION

Mycosis fungoides (MF), likewise known as granuloma fungoides, was first reported in 1806 by French dermatologist Jean-Louis-Marc Alibert [1]. The name mycosis fungoides depicts mushroom-like fungal disease because of its structural appearance [2]. It is the most usual variety of cutaneous T-cell lymphoma that generally involves the skin, which may shape up to affect the viscera. The classical form of MF includes folliculotropic, pagetoid reticuloid and granulomatous cuti laxa that are assorted by the World Health Organization [3, 4]. The lesions form as a consequence of uncontrolled peripheral T cell epidermotropic lymphocyte cell division [3, 5]. More than 65% of primary cutaneous lymphoma is the T cell type, where 25% are formed by B cell and 10% are unspecified [5]. The disease has an indolent clinical course but patients commonly present with a nodular skin rash, patches, and sometimes with associated skin itching. In the early stages, the skin patches can present as eczema or psoriasis-like lesion [2]. Mycosis fungoides is a rare disease and its risk factors are not well elucidated worldwide [6]. We, thus, present two cases of MF, with one of them masqueraded a giant acne and review of literature.
CASE SERIES

Case 1

A 20-year-old female presented with one-year history of the face and left arm swelling that (Figures 1, 2 and 3) was subtle in onset and continue to increase in size. The swellings are painless with no associated itching or ulceration and no preceding history of trauma and no family history of similar illness.

On examination, the patient was clinically stable. She had two swellings on left cheek extending to the maxillary region measuring 8x6 cm and 6x4 cm respectively; it was diffuse, soft, nontender, no differential warmness and did not transilluminate (Figure 1 and 2).

Clinical diagnosis of neurofibromatosis was made and excision biopsy of the left arm mass was taken for histology. Verbal consent was obtained before the pictures were trailed.

Grossly, we received a rubbery grayish white tissue that was partly covered by an ellipse of skin and it measured 4.5x2x2 cm. The cut surface appeared solid grayish white.

Histological sections showed skin tissue displaying monomorphic lymphoid cells focally arranged in a nodular pattern invading the dermis with numerous foci of epidermal infiltrations (Figure 4 and 5). The cells are predominantly small and medium sizes with an occasional blast and they have round to oval vesicular nuclei that have a cerebroid configuration. Focal pautriers microabscesses and areas of necrosis are present. The features are consistent with mycosis fungoides.

Case 2

A 60-year-old female presented with generalized hypo and hyper pigmented skin lesions associated with itching, which were noticed 20 years prior to presentation. The lesions were painless and slowly increased in size and there was no family history reported with similar ailments.

On examination, there were multiple skin lesions predominantly maculopapular, more at the extremities resembling neurofibromatosis. Clinical impression of Aspergillosis was made.

Grossly, we received three fragments of gray, white tissue partly covered by skin aggregating 1.8 cm. Histological sections showed ulcerated skin tissue exhibiting intradermal monomorphic lymphoid cells infiltrating the epidermis and the adipose tissue. The cells have round to oval nuclei with the moderate amphophilic cytoplasm. Quite a few cells that have vesicular nuclei with the cerebroid nuclear pattern were noted (Figure 6, 7 and 8). Focal areas showing pautriers microabscesses were also present (Figure 7) with low mitotic activity. The overall features are consistent with mycosis fungoides.
Mycosis fungoides is one of the primary cutaneous non-Hodgkin lymphomas. The WHO and the European Organization for Research and Treatment of Cancer have classified primary cutaneous lymphoma into; cutaneous T cell lymphoma, cutaneous B cell lymphoma, cutaneous lymphoma from the natural killer cell and unclassified cutaneous lymphoma. These classifications were based on phenotype and molecular characterization [1, 2]. Mycosis fungoides expresses the T cell receptor with αβ+ subunit and CD4+ immunophenotyping [2]. The disease also represents less than 1% of the total number of non-Hodgkin lymphoma, it has an indolent clinical course and better prognosis when detected early [1, 5]. An established association between MF and industrial exposure to oil has been observed [6]. However, the risk factors of the
Primary cutaneous lymphomas are the second most frequent extranodal non-Hodgkin lymphoma in Europe an average incidence rate of 1/100,000 inhabitants have been observed, [6] however, in our environment only two cases were experiential in four decades, perhaps the disease was under diagnosed, looking at its wide clinical presentation that resembles other skin lesions, as a result of this it is possible that many cases might have missed the definitive histological diagnosis. In the United State, from 1973 to 1992 the incidence of MF was 0.36/100,000 inhabitant [6, 7] with the ratio between blacks to Caucasian put at 1.7:1. The proportion of the disease between Asians and the white was 0.6:1 [8].

In a survey in Switzerland, MF constitutes 60% of the total 263 primary cutaneous lymphomas (PCL) that were analyzed within nine-year period and they establish a mean age of 59 years with a slight female preponderance in the proportion of 1:1.4. In our case series, the two patients were female, age 20 and 60 years respectively.

In Brazil, an estimated incidence of 9,640 cases of extranodal non-Hodgkin lymphoma in 2012 including PCL was observed [1].

Mycosis fungoides occurs as patch-like lesions with slight erythematous, pink or brownish coloration which can be a single or multiple lesion of different diameter [9]. The lesion is often seen in covered areas usually gluteal region and thigh [9]. In this case series both patients presented with lesions on sun-exposed areas (cheek, limbs and upper back). Parapsoriasis, neurofibromatosis Aspergillosis and giant acne were differential diagnoses. Sézery syndrome was ruled out based on clinical basis, which shares similar histochemical markers with MF, even though MUM-1 is always positive in Sézery and negative in MF. The provisional diagnosis of parapsoriasis, neurofibromatosis, aspergillosis and giant acne were made. In our case reports the diagnosis was made based on morphological ground immunohistochemistry were not done because the tissue has lost its receptor integrity at the time of the request.

One of four cases that masqueraded giant acne a 20-year-old female and it is reckoned that just 5% of all MF cases occur in childhood and adolescence [10]. The progression of MF in such age group is supposed to be firmer and more aggressive with frequent extracutaneous involvement [9]. Mycosis fungoides is characterized by epidermotropic peripheral T lymphocytes whose phenotype is CD2+, CD3+, CD4+, CD5+, CD8-, CD45RO+, CD20 + and CD30-, with few exceptional cases. Both Ki-67 and CD 30 are negative in the early stage of MF. Recent surveys have indicated that regular T cell MF correlates well with the phase of the disease and prognosis particularly FOXp3 Tregs positive [11, 12]. Hence early detection includes monoclonal T cell lymphocytes, TCR gene rearrangement analysis using PCR is necessary. Knowledge about the molecular mechanism involved in the pathogenesis of the tumour progression in MF is poorly understood [13].

In MF, alteration of p16INK4a gene and p15INK4b gene were discovered in 7 (18%) and 2 (5%) out of 39 cases examined [13].

There are many treatment modalities of MF; such as sunlight and ultraviolet light exposure, steroid, topical and systemic chemotherapies, local superficial radiotherapy, histone deacetylase inhibitors vorinostat, total skin electron beam radiation, photopheresis and systemic therapist with interferon, retinoids, rexinoid or biological therapist [14]. These treatments were used in combination and the treatment modalities depends a tumour, node, metastasis and blood staging system (TNMB) which also prognosticates the tumor [15]. Combined therapy was administered in these patients. However, follow-up was lost after the initial dose.

CONCLUSION

Mycosis fungoides (MF) is a rare disease in our environment even though it is common among the black race with male preponderance and generally it has an indolent clinical course. The diagnosis of the disease is still challenging, especially in developing countries because of its confusing clinical presentation. A few skin specialists, especially in underdeveloped countries have made the disease sub-optimally detected most of the time biopsies are not done. Diagnostic tools such as immunohistochemistry can make some early lesion to be detected. Hence, clinicians should always consider MF as one of their differential diagnosis whenever patients presented with skin lesions and biopsy should be encouraged for histology.

REFERENCES


Author Contributions
Bukar Abba Zarami – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

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The corresponding author is the guarantor of submission.

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Conflict of Interest
Author declares no conflict of interest.

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