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Case Report

Rapidly progressive tumor stage mycosis fungoides: A case report from Syria

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ABSTRACT

Introduction and importance: Mycosis Fungoides, the most common type of cutaneous T-cell lymphomas (CTCLs), has typically an indolent course over years or decades. Patches grow into infiltrated plaques which may turn eventually into tumors. The tumor stage represents advanced lymphoma, but this occurs in a minority (~10%) of cases.

Case presentation: we present a rare case of mycosis fungoides in a 30-year-old woman showing rapid progression to tumor stage, unlike the indolent clinical course seen classically.

Clinical discussion: Mycosis Fungoides (MF) is the most common primary cutaneous T-cell lymphoma, representing less than 1% of the total number of non-Hodgkin lymphoma. Patients with a classical type of MF progress from patch stage to plaque stage and finally to tumor stage disease, and they have a protracted clinical course over years or even decades.

Conclusion: Although mycosis fungoides is a rare disease, it requires a high degree of suspicion clinically. The disease can have an excellent prognosis when identified and treated promptly.

1. Introduction

It was the French physician Jean-Louis-Alibert who described Mycosis Fungoides (MF) for the first time in 1806, when he suggested the name because of the appearance of the skin lesions that looked like mushrooms [1]. MF is the most common primary cutaneous T-cell lymphoma (CTCL) and can be defined as a primary cutaneous non-Hodgkin lymphoma (NHL) of T-cell origin. It is a tumor composed of small/medium-sized, epidermotropic T-helper lymphocytes. This disease accounts for ~50% of all PCLs and ~60% of cutaneous T-cell lymphomas (CTCLs) [2]. Genetic background, environmental factors, chronic antigenic stimulation, and exposure to cancerogenic agents have all been considered risk factors for MF, yet its etiology is far to be known [3]. Most patients are adults with a median age at diagnosis between 55 and 60 years, but it may also be seen in children and adolescents [4]. MF is twice as common in males as in females. The clinical course is typically indolent, extending over years or even decades. Patches grow into infiltrated plaques which turn eventually into tumors [5]. Although this is a cutaneous lymphoma, lymph nodes and visceral organs may be involved at the terminal stages of the disease. The initial skin lesions have a predilection for the buttocks and covered sites of the trunk and limbs [4]. Most case reports described the challenges and difficulties in diagnosis at early stages because the symptoms and skin biopsy findings are similar to many other conditions, but in this case, we present a patient with rapid progression to the tumor stage.

This case report has been reported in line with the SCARE criteria 2020 [6].

2. Case presentation

A 30-year-old woman presented to the dermatology department at Tishreen University Hospital in 2021 suffering from nodular and tumorous skin lesions for two months. She had erythematous itchy maculopapular eruption four years before tumors and was treated with

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topical steroids and anti-histamine medications without complete improvement. In the last two months, she noticed a rapid increase in the size of the lesions which also changed their appearance. The patient does not smoke or drink alcohol. She had no history of allergies. There was no tumor history in her family. On examination, there were multiple nodules and tumors symmetrically distributed on the axillary region, upper arm, and abdomen. The tumors were 0.5–4 cm in size, itchy and mobile over underlying structures, and some of them had started to ulcerate. There were only a few scaly infiltrated plaques [Fig. 1]. No enlargement was found in the lymph nodes, liver, and spleen. A clinical impression of MF was made and followed by a skin biopsy sent to the pathology department for histopathological study. Microscopic examination revealed a massive infiltration of malignant T-cells in the epidermis and dermis [Fig. 2A] with dermal reticular fibroplasia [Fig. 2B]. Atypical lymphoid cells surrounded by a clear halo in the epidermis (epidermotropism) were seen [Fig. 2C]. Malignant cells were pleomorphic with cerebriform irregular nuclei [Fig. 2D]. Immunohistochemical studies demonstrated, that tumor cells were positive for CD3 and CD4. On the other hand, CD8 and CD20 stains were negative [Fig. 3]. The patient was referred to the oncology department for initiation of chemotherapy.

3. Discussion

Mycosis Fungoides (MF) is the most common primary cutaneous T-cell lymphoma, representing less than 1% of the total number of non-Hodgkin lymphoma. Patients with a classical type of MF (also known as Alibert – Bazin) progress from patch stage to plaque stage and finally to tumor stage disease, and they have a protracted clinical course over years or even decades [5]. Our patient had a rapid progression to the tumor stage within 4 years. Many patients never progress beyond the plaque stage. However, a minority (~10%) of patients may develop nodules or tumors [3]. The growth rate of tumors in MF is variable, in some cases, they grow rapidly in a matter of a week, and in others, they are relatively stable for months [3]. Tumors may occur anywhere on the body, but have a predilection for the face and body folds: axillae, groin, antecubital fossae, and inframammary area in women [7]. They may be solitary, localized, or generalized and are often (but not always) observed in combination with patches and plaques, ulceration is common [3]. In our case, many tumors rapidly grew within two months and were predominantly located in the axilla and abdomen with variable diameters, and only a few infiltrated plaques were found. Lymph nodes, lungs, spleen, and liver are the most common frequent sites of extracutaneous involvement [3,8]. Fortunately, our patient had no palpable lymph nodes, and no hepatomegaly or splenomegaly. Thus, the possibility of extracutaneous disease is less likely. The neoplastic T cells in tumor stage MF are derived from $CD4^{+}$ T cells with a Th2 cytokine profile (IL4, IL5, IL6, IL10) which may result in impairment of the Th1 cell-mediated antitumor response and contribute to the immunosuppression seen in patients with advanced MF [3]. The development of tumors or large cell transformation usually heralds the terminal stages of the disease [3]. However, many patients in the tumor stage even in the cases of large ulcerated tumors, are in excellent general condition but this is seen only in the absence of extracutaneous disease. The most common complications seen in these patients are infections, particularly sepsis, caused by the presence of large ulcerative tumors in the background of an impaired immune system [3]. Many patients with advanced disease may require immunotherapy or chemotherapy, anyway, treatment is frequently palliative and decided on an individual patient's basis [9]. The major challenge in treating such patients would be to target the skin lesions without impairing the immune system, which is paramount for control of both the neoplastic progression and the infective complications. Unfortunately, this medical need has not been met yet [1].

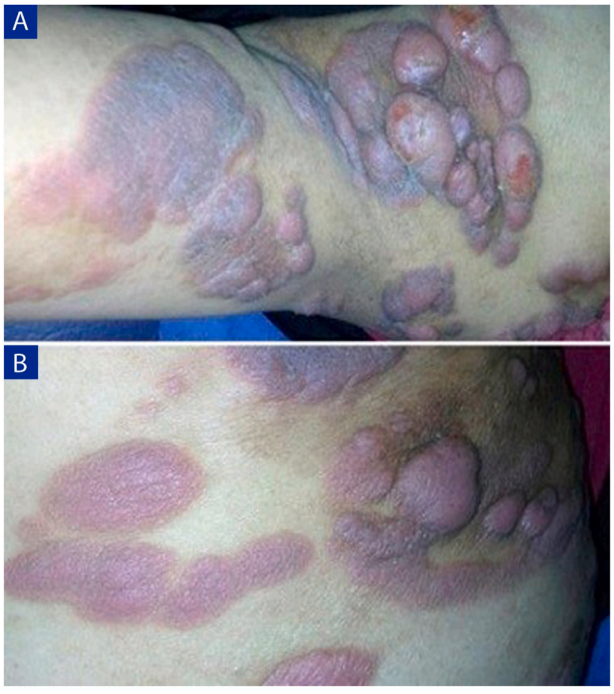

Fig. 1. Clinical image shows large nodules and tumors with scaly infiltrated plaques on axilla (A) and abdomen (B).

4. Conclusion

Although mycosis fungoides is a rare disease, it requires a high degree of suspicion clinically. The disease can have an excellent prognosis when identified and treated promptly. Our case highlights an uncommon progression of MF as it reached the tumor stage rapidly, which signifies the importance of follow-up for such lesions while keeping the most common CTCL in mind.

Ethical approval

No ethical approval was needed for this case report.

Sources of funding

None declared.

Author contribution

Yara Melhem: study design, data collection, data analysis, and writing.

Moatasem Hussein Al-janabi: study design, and writing.

Maha Mansour: data analysis and writing.

Zuheir Al-shehabi and Hameed Suleman: in reviewing the manuscript.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Registration of research studies

Not applicable.

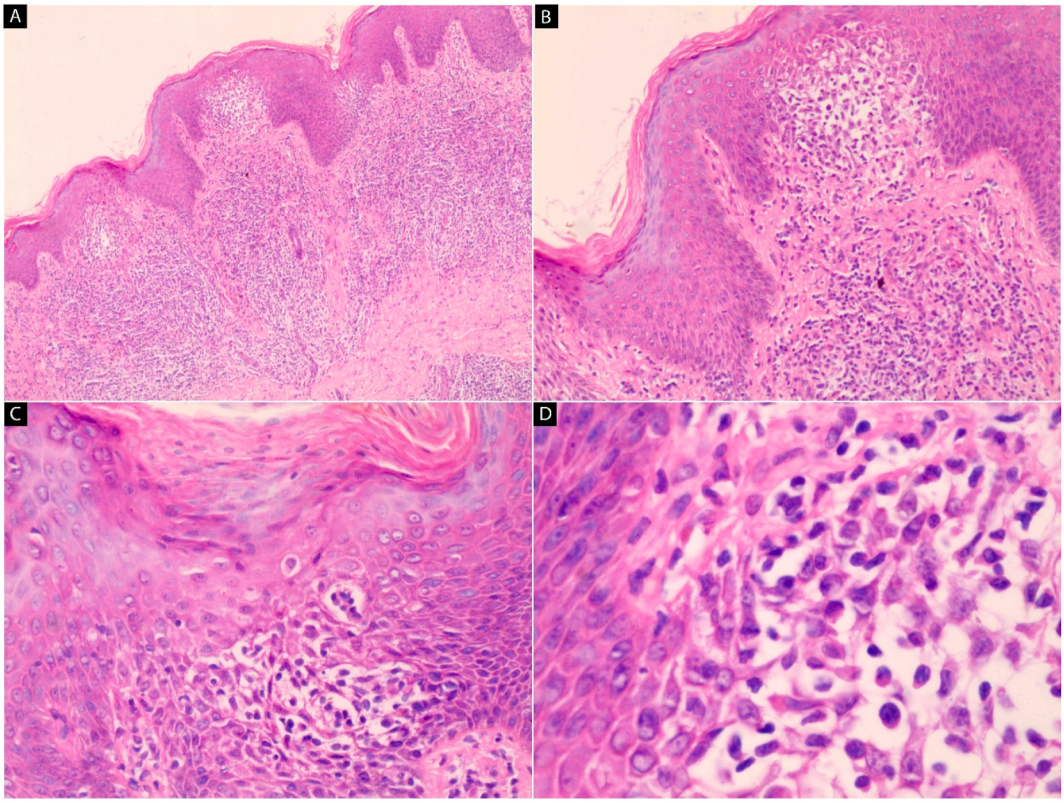

Fig. 2. Hematoxylin-eosin staining (A–D) Microscopic images of Mycosis Fungoides. (A) Marked infiltration of malignant T-cells in the epidermis and dermis (x40). (B) Papillary dermal fibrosis is present (x100). (C) Atypical lymphoid cells surrounded by a clear halo in the epidermis are seen (x200). (D) Malignant cells are pleomorphic with irregular cerebriform nuclei (x400).

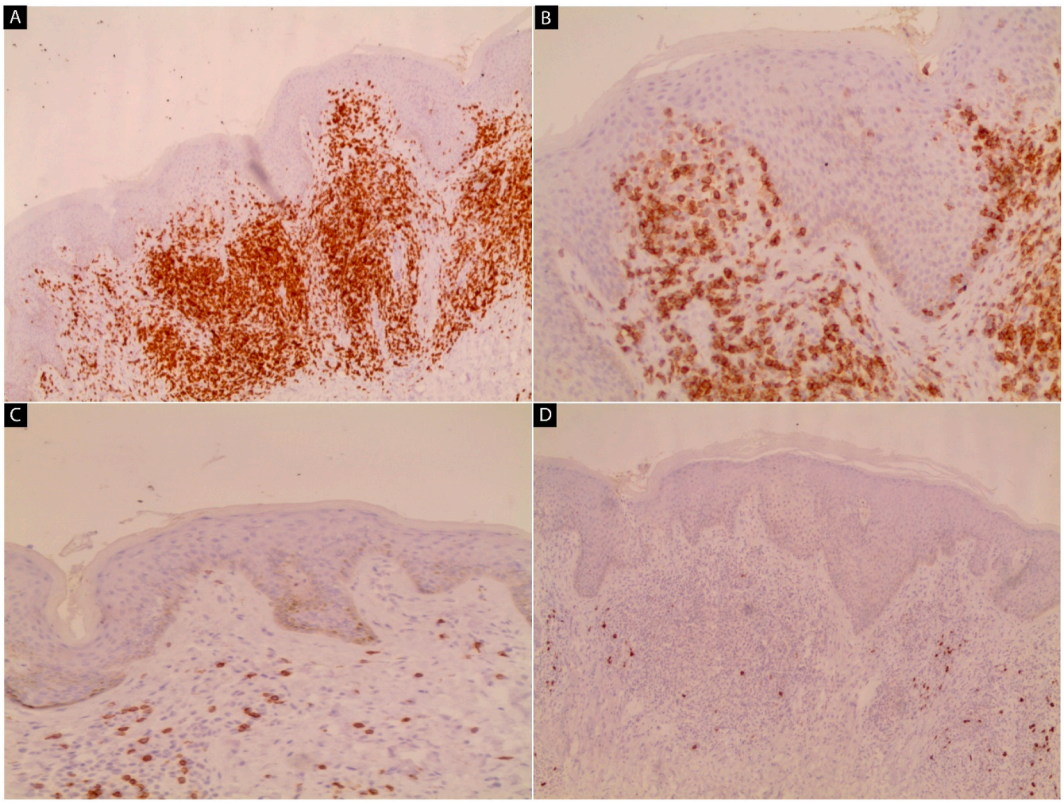

Fig. 3. Immunohistochemical staining (A–D). (A and B) Tumor cells are positive for CD4 and CD3 respectively. (C and D) Tumor cells are negative for CD8 and CD20 respectively.

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Declaration of competing interest

The authors have no conflicts of interest to declare.

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