



Mycosis Fungoides in a 60-year-old Male with Rapid Progression to Tumor Stage: A Case Report

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ABSTRACT

Mycosis fungoides is the most common primary cutaneous T-cell lymphoma with an indolent course. Typical manifestations include erythematous patches and plaque-type lesions, with about 25% of cases progressing into nodules and tumors. This case was presented at the Department of Dermatology, Mugda Medical College Hospital, in May 2023. A 60-year-old Bangladeshi male presented with diffuse infiltrative erythematous plaques and ulcerated pustules on the skin of his upper body for six months. The initial clinical diagnosis was exfoliative dermatitis, but a skin biopsy confirmed mycosis fungoides through immunohistochemistry. The patient later developed generalized lymphadenopathy. The immunohistochemistry findings of a lymph node biopsy revealed nodal involvement of the disease. Systemic chemotherapy was initiated, but he unfortunately passed away due to neutropenic sepsis. This case highlights the importance of early diagnosis in mycosis fungoides, a disease that can often be mistaken for inflammatory dermatoses. Early skin biopsy with immunohistochemistry can lead to an early diagnosis of MF, potentially changing the course and outcome of the disease.

Keywords: Mycosis Fungoides, T-cell Lymphoma, Immunohistochemistry.

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INTRODUCTION

Mycosis fungoides (MF), also known as granuloma fungoides or Alibert-Bazin syndrome, is the most common variant of primary cutaneous T-cell lymphoma [1, 2]. However, it is rare, with a reported incidence of 1 case per 1,000,000 [2]. It typically affects individuals aged 45 to 60, but can also be observed in children and adolescents [1-5]. The male-to-female ratio is 2:1 [3]. The disease usually follows an indolent clinical course, with early manifestations including dermal patches and plaque-type lesions that are often scaly and erythematous, resembling benign dermatologic conditions. In about 25% of cases, there is gradual

progression to nodules and tumors. Diagnosing MF can be challenging in the early stages due to nonspecific clinicopathological findings and delayed biopsy practices. It is often misdiagnosed for an extended period as another skin lesion, such as psoriasis, chronic contact dermatitis, or other similar conditions. A definitive diagnosis can be made through histopathology and immunohistochemistry of skin biopsy [2]. This case report describes a 60-year-old male with rapid progression to tumor stage IV. Reporting such cases is crucial in promoting the practice of early skin biopsy for timely diagnosis and improved prognosis.

Clinical History

A 60-year-old male from Bandarban, located in the Chittagong Hill Tracts, was referred to the Department of Dermatology at Mugda Medical College Hospital, Dhaka, Bangladesh in May 2023 due to a severe dermatological condition. The patient was a non-smoker and non-alcoholic, with no pertinent past medical or medication history.

MATERIALS AND METHODS

On clinical examination, the patient exhibited painful, pruritic, diffuse infiltrative erythematous plaques and ulcerated pustules that had progressed rapidly, affecting the scalp, face, chest, back, upper extremities, and abdomen, accompanied by significant facial edema (Figure 1A&B). Over the past two months, the lesions have continued to advance and enlarge, raising considerable concern regarding the underlying etiology and course of the condition. Initial management included topical corticosteroids and oral antihistamines, based on a preliminary diagnosis of eczema; however, this treatment yielded no improvement. The patient was hospitalized and underwent skin biopsy from an alopecic plaque of the occipital region of the scalp for a clinical diagnosis of exfoliative dermatitis with the differentials of pemphigus vegetans, mycosis fungoides and pustular psoriasis. The Department of Pathology at Mugda Medical College, conducted a comprehensive examination of his skin biopsy specimen. The microscopy revealed an epidermotropic infiltrate of atypical lymphocytes having pleomorphic, irregular nuclei in the superficial to mid-reticular dermis (Figure 1C). Pautrier's microabscess in epidermis and folliculotropism in dermis were also present. Differential Diagnoses were considered Exfoliative dermatitis, Pemphigus vegetans, Pustular psoriasis, and Mycoses fungoides. Immunohistochemistry conducted at the Department of Pathology, Bangladesh Medical University, Dhaka

revealed that the atypical lymphocytes were positive for CD3, CD2, CD5, CD4 and CD8 and negative for CD7 (Figure.1D-F), confirming the diagnosis of mycosis fungoides. Three days after hospital admission, the patient was found to have bilateral axillary and cervical lymphadenopathy. At the same time, his complete blood count showed neutrophilic leukocytosis, and the peripheral blood film revealed microcytic hypochromic anemia with eosinophilia. The largest cervical lymph node was biopsied. Grossly, the cut surface of the lymph node was waxy white, and microscopy revealed effacement of nodal architecture by a diffuse infiltrate of medium to large atypical lymphocytes with hyper-convoluted nuclei, irregular nuclear membrane and prominent nucleoli with brisk mitotic activity (Figure 2 A&B). These atypical lymphocytes were positive for CD3, CD2, CD5, CD4 and CD8 (CD4 predominant) and negative for CD7, supporting nodal involvement by the T-cell lymphoma (Figure. 2C-F).

Diagnostic Timeline

December 2022: Start symptoms and had treatment for contact dermatitis

May 2023: Generalized symptoms developed and admitted Mugda Medical College Hospital

Biopsy and Histopathology were done in May, 2023

Immunohistochemistry was done at June 2023

Patient's condition was deteriorated before starting specific treatment and ultimately died

RESULTS

The treatment plan was immunotherapy. During follow-up, systemic chemotherapy was started, but the patient died due to neutropenic sepsis within 6 days. The patient was diagnosed as a case of Mycosis Fungoides, stage IVA2, based on criteria of the European Organization of Research and Treatment of Cancer and the International Society for Cutaneous Lymphoma.

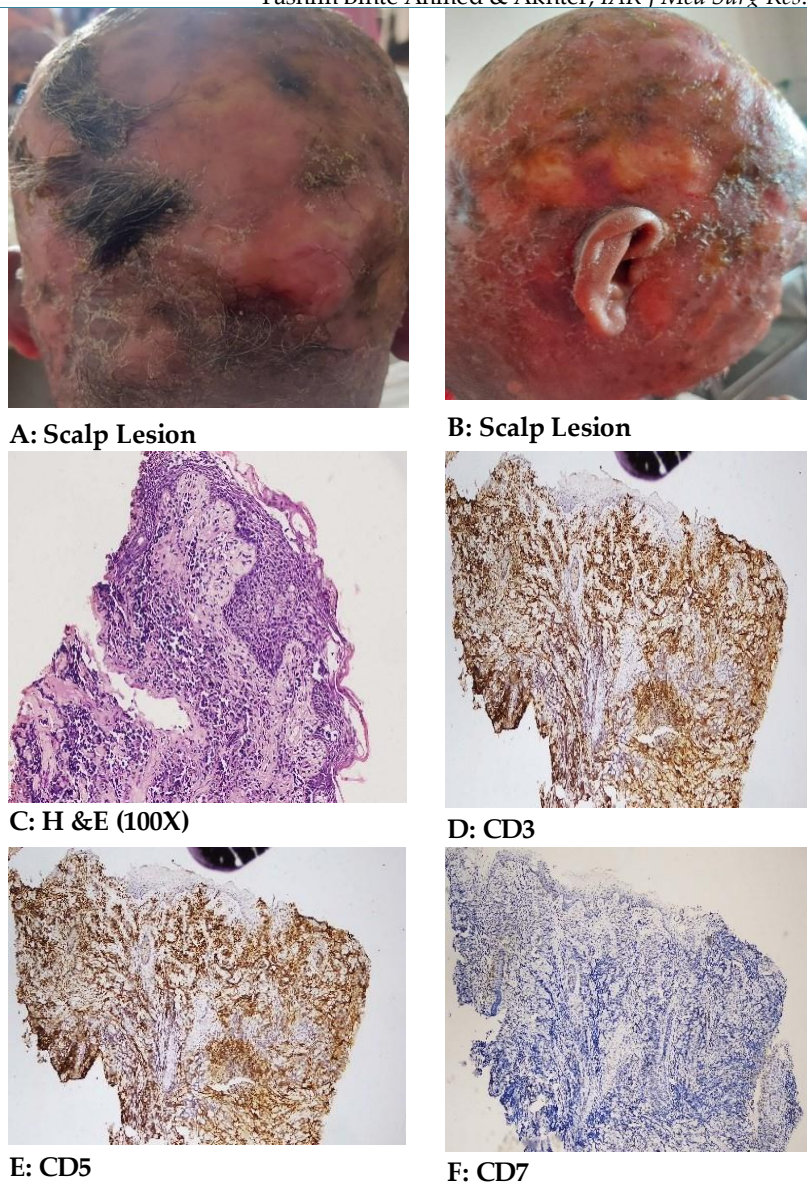


Figure 1: Clinical, Microscopic and Immunohistochemistry of Lesional Skin Biopsy from Scalp

Figure 1 shows A&B. Erythematous, ulcerated patches, plaques and nodules on back and lateral side of scalp. C. Microscopy of skin sections showing

epidermotropic atypical lymphocyte and also diffuse infiltrate in dermis; D-F. IHC stains showing lymphocytes are positive for CD3 and CD5 but negative for CD7.

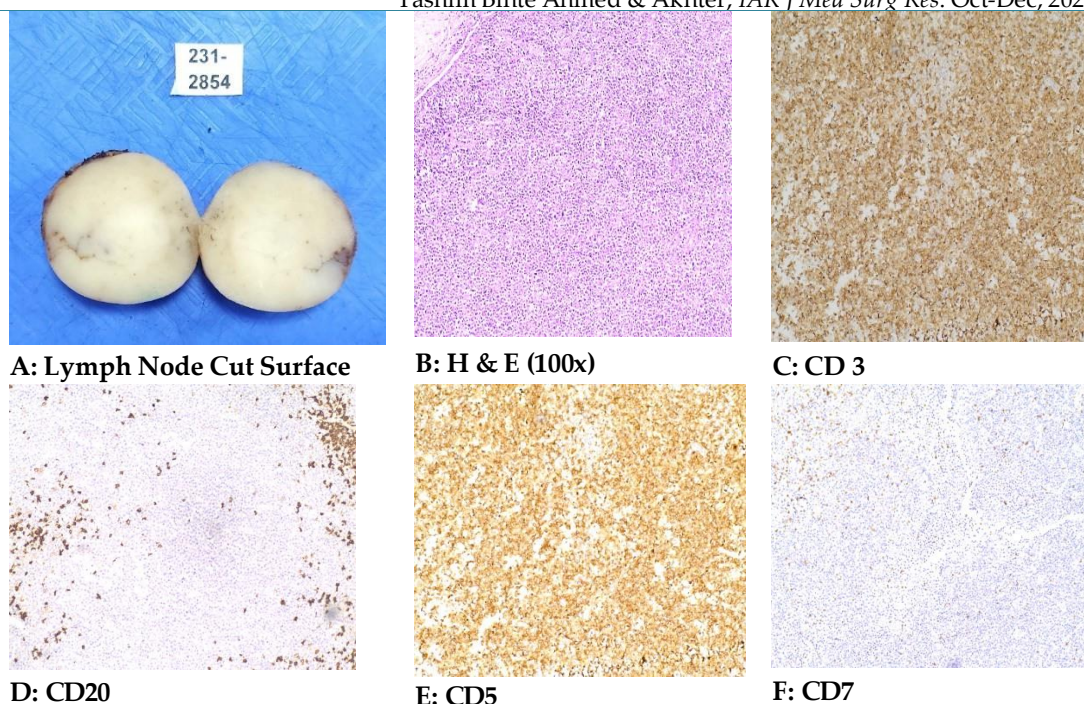


Figure 2: Gross, Microscopic and Immunohistochemistry of Cervical Lymphnode

Figure 2 shows A. Grossly waxy-white cut surface of lymph node; B. Infiltrate of atypical lymphocytes in microscopic sections; C-F. IHC stains showing lymphocytes are CD3 and CD5 positive but CD7 and CD20 negative.

DISCUSSION

Mycosis Fungoides (MF) typically presents as generalized erythematous patch-like lesions and is traditionally classified into three clinical presentations: patches, plaques, and nodules. Patch and plaque lesions are mainly located in areas protected from sunlight, such as the buttocks, medial thighs, back, and breasts, although any part of the skin can be affected. This condition is often misdiagnosed as chronic contact dermatitis or psoriasis, especially in the early stages. The cause of MF remains unknown, with various factors, including genetic predispositions, environmental exposures, and infectious agents, suggested as potential triggers for lymphocyte activation and transformation [1, 6-12]. Continuous occupational exposure to specific substances in industries like glass manufacturing, ceramics, paper processing, painting, and agriculture may lead to persistent antigenic stimulation of CD4⁺ memory T cells, creating an environment of chronic inflammation that could eventually result in the development of a malignant T-cell

clone [1, 3, 5, 12]. While some studies have identified HTLV-1 or HTLV-2 gene sequences in peripheral blood or biopsies from MF lesions, others have found no significant link between MF and HTLV-1 [8, 9]. In this specific case, our patient was a farmer with no known chemical or occupational exposures, and most notably, no testing for HTLV-1 or HTLV-2 was performed. The diagnostic histopathological parameters for mycosis fungoides (MF) include dense infiltration of atypical lymphocytes with hyper-convoluted or cerebriform nuclei in the dermis, epidermotropism, and Pautrier's microabscesses in the epidermis.

The typical immunophenotype of atypical lymphocytes in MF is CD2⁺, CD3⁺, CD4⁺, CD5⁺, CD8⁻, CD45RO⁺ and TCR GAMMA. CD20 may be positive in a few exceptional cases. Loss of CD7, a frequent characteristic in all stages of the disease, is an important diagnostic marker as well as, CD30⁺ in all stages, including the advanced stage. Our patient's immunophenotype also showed similar results except for a negative CD30. No molecular test was conducted in this case. Similar clinicopathological findings have been observed in the USA, Nigeria, Africa, Syria, Indonesia and China [1, 2, 5, 10-12]. The clinical stage, which is a reflects of the type and extent of skin involvement and extra-

cutaneous invasion, is a crucial determinant of the disease's prognosis. Understanding this role is essential for effective treatment planning [13]. Several factors can indicate a poor prognosis, including advanced stage, failure to respond in early treatment, age over 60, and raised lactate dehydrogenase. In this case, the age of the patient, delayed diagnosis, and rapid progression of the disease, including extra cutaneous involvement, were the parameters associated with a rapidly worsening prognosis.

CONCLUSION

Diagnosing mycosis fungoides poses significant challenges and is often delayed due to its phenotypic similarities with inflammatory dermatoses like eczema, psoriasis, or lichen planus. These delays can negatively impact patient outcomes. In suspected cases of mycosis fungoides, an early skin biopsy followed by histopathology and immunohistochemistry is essential for an accurate diagnosis by distinguishing it from other skin conditions of almost similar presentations.

Ethical Statement

Data was collected after obtaining written informed consent from patient by maintaining Helsinki declaration.

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