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Case Report
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Folliculotropic Mycosis Fungoides: An Atypical Clinical Presentation

Alexandria B Glass, Christopher Buckley and Marcus B Goodman*

Department of Dermatology, Philadelphia College of Osteopathic Medicine, Georgia, USA

Abstract

Mycosis Fungoides (MF) is the most commonly encountered type of cutaneous T-cell lymphoma, with three rare distinct variants including pagetoid reticulosis, granulomatous slack skin disease, and Folliculotropic MF (FMF). This article discusses a case report of FMF presenting in a 42-year-old Caucasian male in an atypical location. The patient presented with a two-year history of erythematous plaques with overlying follicular papules on his right arm and left lateral buttocks. He also had a patch of alopecia containing flesh colored follicular papules on his right anterior lower leg. The lesion on the arm and leg were biopsied showing abnormal folliculotropic lymphocytes positive for CD3, an elevated CD4:CD8 ratio, and diminished CD7 positivity. Stains for CD20 and CD30 were negative confirming the diagnosis of FMF. This case shows an atypical presentation of FMF due to its presence on the extremities instead of the typical locations of the head and neck. This case exemplified why FMF should be in the differential diagnosis of erythematous papules and/or focal areas of alopecia, regardless of the location.

Keywords: Cutaneous T-cell lymphoma; Mycosis fungoides; FMF; CD4; CD8

Introduction

Mycosis Fungoides (MF) is the most commonly encountered type of Cutaneous T-cell Lymphoma (CTCL), comprising about 60% of all cases. The classic form of MF, also referred to as the "Alibert-Bazin" type, generally is more prevalent in males and is diagnosed during the fifth to sixth decades. It has an indolent clinical course and progresses slowly over years, from patch, to plaque, and finally tumor stage. Histology shows medium to large atypical T-cells with cerebriform nuclei infiltrating the epidermis. When diagnosed early, MF has a favorable prognosis with only a minority of disease related deaths. According to the World Health Organization-European Organization for Research and Treatment of Cancer (WHO-EORTCF), there are three distinct variants of MF: pagetoid reticulosis, granulomatous slack skin disease, and Folliculotropic MF (FMF) [1,2]. FMF accounts for about 10% of all MF cases, making it the most common variant [1]. There are multiple similarities between classic MF and FMF, such as being more prevalent in adult males and having an increased CD4 to CD8 ratio. However, the sites most often affected, the histology, and clinical course are very different [2,3].

Case Presentation

A 42-year old Caucasian male with a past medical history of testicular cancer presented to the office with a chief complaint of a waxing and waning itchy red areas on his right arm, right lower leg, and left lateral buttocks for two years. He stated the rash first appeared on his arms then left lateral buttocks as slightly pruritic erythematous papules. He then developed a patch of alopecia on his right leg. He denied pain, tenderness, and edema of these sites. Review of systems was otherwise negative including fever, chills, weight loss, night sweats, nausea/vomiting, diarrhea, and other cutaneous manifestations. Other than testicular cancer, the patient's medical history and family history were negative. He did not take any medications, reported no allergies, and denied drug use. He reported occasional alcohol use and being a daily smoker. On physical exam, the patient was well-appearing in no acute distress. He had ill-defined faint erythematous plaques with few overlying follicular papules, crusting, and alopecia present on his right posterior forearm and left lateral buttocks. On his right anterior medial lower leg, accentuated flesh colored follicular papules and alopecia were observed (Figure 1). A 3 mm punch biopsy was performed on the right posterior forearm and a shave biopsy on the right anterior medial lower leg.

On histopathological examination, both biopsies demonstrated a dense T-cell infiltrate

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Correspondence:
Alexandria B Glass, Department of Dermatology, Philadelphia College of Osteopathic Medicine (PCOM), 2500 Hospital Boulevard, Suite 280, Roswell, Georgia 30076, Georgia, USA, Tel: +1-770-754- 0787; E-mail: alexandriabglass@gmail.com

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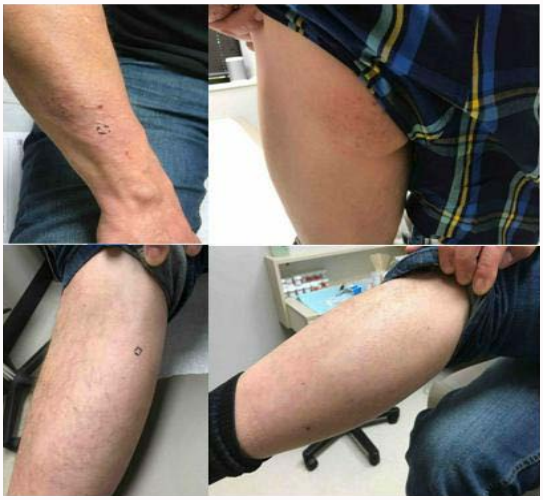

Figure 1: Clinical images of patient on presentation. (Purple marking indicates biopsy site locations). (Top left) right posterior forearm (top right) left lateral buttocks (bottom left, bottom right) right anterior medial lower leg.

surrounding the hair follicle. Further workup confirmed the diagnosis of FMF with positivity for CD3 and an elevated CD4:CD8 ratio. CD7 showed diminished positivity with negative CD20 and CD30 (Figure 2).

The patient was informed of their diagnosis and referred to oncology for further workup and treatment.

Discussion

Folliculotropic Mycosis Fungoides has characteristic clinical and histological findings that make it a distinct entity from classic mycosis fungoides. Unlike classic MF which typically presents on the buttocks and other sun-protected sites, FMF has a predilection for the head and neck region. It usually presents as grouped follicular papules, acneiform papulopustular lesions, and/or areas of alopecia. In addition, lesions can be quite pruritic unlike classic MF. On histology, atypical lymphocytes surrounding the hair follicle, usually with mucinous degeneration of the follicle (follicular mucinosis), can be observed [1,3,4]. This folliculotropic infiltrate, usually sparing the epidermis, is in contrast to the epidermotropic infiltrate seen in classic MF. Immunohistochemical analysis, like MF, show a CD3+, CD4+, CD8- phenotype with sometimes admixed CD30+ cells. The deeper follicular infiltrate in FMF can make this CTCL more difficult to treat with topical therapies. In addition to being more refractory to treatment, FMF has a more aggressive course than classic MF [1,4]. Prognosis for patients with FMF is worse than classic patch-stage MF and outcomes are similar to patients with tumor-stage MF. Many independent risk factors are associated with decreased overall survival such as age greater than 60 years at diagnosis, more progressed clinical stage, and secondary bacterial infection [3]. Other factors, such as sex, the duration of disease prior to diagnosis, the extent of cutaneous lesions, pruritus, and presence of follicular mucinosis appear to have no effect on survival [5]. Treatment for FMF varies depending on the clinical stage at diagnosis. Early-stage FMF is typically treated with topical therapies including topical steroids, Ultraviolet B (UVB), and

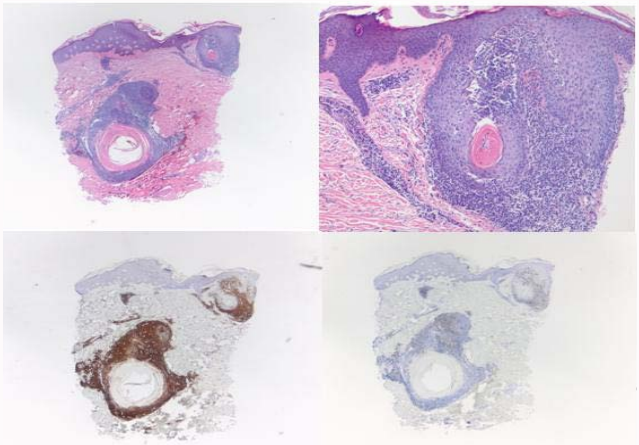

Figure 2: Histological images from the patient's biopsy. Dense atypical lymphocytic inflammation can be observed surrounding the hair follicle. (Top left) 2x magnification (top right) 10x magnification (bottom left) 10x magnification with CD4 stain (bottom right) 10x magnification with CD7 stain.

Psoralen Plus Ultraviolet A (PUVA). In advanced-stage FMF, more aggressive forms of treatment are used such as local radiotherapy, total skin electron beam irradiation, and PUVA combined with local radiotherapy [6].

This case demonstrates an atypical presentation of FMF. The patient discussed in this case was not only younger than the average age at diagnosis (42 years of age versus 55-60 years of age), but also had an atypical site location with the initial rash observed on his arm, buttocks, and leg in comparison to the typical head and neck region. FMF remains to be a diagnostic challenge due to its various clinical presentations and should be considered in areas of alopecia and/or erythematous papules.

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