

Received: 2020.02.07

Accepted: 2020.05.05

Available online: 2020.08.13

Published: 2020.09.18

Mycosis Fungoides Palmaris et Plantaris on the Plantar Aspect of the Foot: A Case Report

Authors' Contribution:
 Study Design A
 Data Collection B
 Statistical Analysis C
 Data Interpretation D
 Manuscript Preparation E
 Literature Search F
 Funds Collection G

BCDEF 1 **Ian Beiser**
 BCDE 2 **Joon Yim**
 BCDE 1 **Erin Robles-Sherman**
 AE 1 **Gene S. Mirkin**
 ABCDEF 2 **Xingpei Hao**

1 Department of Podiatry, Foot and Ankle Specialists of The Mid-Atlantic, LLC, Rockville, MD, U.S.A.

2 Department of Pathology, Foot and Ankle Specialists of The Mid-Atlantic, LLC, Rockville, MD, U.S.A.

Corresponding Author: Xingpei Hao, e-mail: xhao@footandankle-usa.com

Conflict of interest: None declared

Patient: Male, 54-year-old
Final Diagnosis: Mycosis fungoides
Symptoms: Pain • patch
Medication: —
Clinical Procedure: Topical psoralen plus ultraviolet A (PUVA) photochemotherapy
Specialty: Dermatology • Oncology • Pathology • Podiatry

Objective: Rare disease

Background: Mycosis fungoides palmaris et plantaris (MFPP) is a rare variant of the cutaneous T cell lymphoma mycosis fungoides (MF). Here we report the case of a middle-aged man with MF on the sole of his left foot.

Case Report: A 54-year-old man had a diffuse, hard lesion in the middle of the arch on the sole of his left foot for 3 years. Physical examination revealed a 3-cm scaly, keratotic patch with slight erythema on the left plantar central arch. Histopathological evaluation of a punch biopsy specimen revealed infiltration of atypical lymphocytes in the upper dermis. Immunostaining of the atypical lymphocytes showed strong expression of CD3, CD4, and CD5; reduced expression of CD7 and CD8; and no expression of CD20. Periodic acid-Schiff staining was negative for fungi. The patient's lesion was diagnosed as MFPP and he was treated with topical psoralen plus ultraviolet A (PUVA) photochemotherapy. At 5-year follow-up, his condition was in complete remission.

Conclusions: MFPP is a rare clinical variant of MF restricted to the palmoplantar area, and is histologically characterized by upper dermal infiltration of atypical lymphocytes with preserved CD3, CD4, and CD5 expression but decreased CD7 and CD8 expression. PUVA photochemotherapy is a treatment option associated with excellent prognosis.

MeSH Keywords: Foot Diseases • Lymphoma, T-Cell, Cutaneous • Mycosis Fungoides • PUVA Therapy

Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/923361>



950



—



1



15



Background

Mycosis fungoides (MF), first described as a mushroom-like lesion by Alibert in 1806 [1], accounts for more than half the cases of cutaneous T cell lymphoma (CTCL). Clinical presentations of MF include patches, plaques, tumors, erythroderma and other variants [2]. MF palmaris et plantaris (MFPP) (keratoderma-like), which manifests primarily on the palms and soles, is rare [1]. Fewer than 30 cases have been reported in the English-language literature [3–5]. Here, we describe the clinical presentation and histopathology in a case of MFPP with the goal of raising awareness of this rare variant of MF.

Case Report

A 54-year-old man presented with a diffuse, hard lesion in the middle of the arch of his left foot, which had been there for 3 years. He described the onset of the lesion as insidious and its nature as cracked, dry, scaly, and not painful. The patient reported a history of severe eczema with dry patches in the same area, which he had treated unsuccessfully with topical cortisone cream and over-the-counter acid. Over the past several months, the lesion had gradually thickened, hardened, and becoming noticeably tender, progressing to the point where it was painful to walk. The patient's surgical and medical histories included thyroidectomy (for thyroid cancer) at age 28 years, headaches/migraines, hypertension, and bipolar disorder. His family history was unremarkable. A review of systems did not reveal any major abnormalities.

Dermatological examination revealed a 3.0-cm, scaly, keratotic patch with slight erythema in the plantar central region of the left arch, which was tender to palpation. Neither clear signs of skin atrophy nor lymphadenopathy were found. Results of laboratory testing were within normal limits.

Histological evaluation of a specimen from skin punch biopsy revealed infiltration of atypical lymphocytes in the upper dermis (Figure 1A). Most of the atypical lymphocytes were round or ovoid with a cerebriform nuclear contour but with no clear nuclear membrane or nucleoli (Figure 1B). The atypical lymphocytes infiltrated up into the epithelial layers (epidermotropism) in single units or small clusters (Figure 1C), and down into the eccrine sweat glands (syngiotropism) (Figure 1D) and the walls of the blood vessels in the dermis (Figure 1E). Immunostaining of the atypical lymphocytes showed almost uniformly strong positive staining for CD3, CD4 (Figure 1F), and CD5, and about 30% positivity for CD7 (Figure 1G) and CD8 (Figure 1H), but staining for CD20 was negative. The ratio of CD4- to CD8-positive cells was about 3: 1. Periodic acid-Schiff staining was negative for fungal elements (both spores and hyphae). The histomorphology and immunostaining profiles

were consistent with MFPP. Treatment options were discussed with the patient in detail, and he was referred to a dermatologist for topical psoralen plus ultraviolet A (PUVA) photochemotherapy. Complete remission was achieved and the patient had no recurrence at 5-year follow-up.

Discussion

MFPP manifests primarily on the palms and soles and is a rare variant of MF. Fewer than 30 cases have been reported since the condition was first described by Resnik in 1995 [1,3–5]. Clinically, the age of patients with MFPP (on the soles) has ranged from 11 to 83 years, with the mean and median being 51.8 and 52 years, respectively. Male patients are predominant, with the ratio of men to women being 2.3: 1. The duration of the lesions varies from 3 months to 25 years, with the mean and median being 4.2 and 3 years, respectively [3]. Clinical presentations include psoriasiform [6], erythematous hyperkeratotic patches, plaques, blisters, ulceration, and tumor [3–5,7]. The lesions need to be differentiated from other those of other dermatoses, including palmoplantar psoriasis vulgaris, dermatophytosis, secondary syphilis, eczema, hyperkeratotic lichen planus, contact dermatitis, and warts [8]. The accuracy of diagnosis can be improved with histological evaluation of biopsies that includes immunostaining profiling and genetic analysis of polymerase chain reaction (PCR)-based monoclonal rearrangements of T cell receptors (TCRs). No TCR clonal analysis was conducted in our case because the morphological evaluation with immunostaining profiling was diagnostic.

Treatment options for MFPP include monotherapy with PUVA [1,9] or PUVA combined with topical or systemic administration of corticosteroids [1], vitamin A [1], vitamin D [9], nitrogen mustard [10], or doxorubicin, gemcitabine and methotrexate [9–11]; carbon dioxide laser [12]; excimer laser [13]; or radiation therapy [3,14]. Patient response (complete or partial) to any type of treatment reportedly is excellent, with a response rate of 95% [3]. The progression of MFPP can be controlled with further treatment, even when the tumor has relapsed or recurred [1,9,10]. Results from a recent study showed that treatment with brentuximab vedotin, an antibody that selectively targets tumor cells expressing CD30 antigen, led to complete remission of a tumor-stage MFPP with large cell transformation and partial expression of CD30 [5].

The patient in the present study was treated with topical PUVA photochemotherapy, which resulted in complete remission at 5-year follow-up. The efficacy of PUVA as a monotherapy for MF has been demonstrated in multiple studies, as reviewed by Olsen [15]. Complete remission of MFPP treated with PUVA has been reported previously [1]. Selection of treatment should be based on the location, size, and stage of the MFPP;

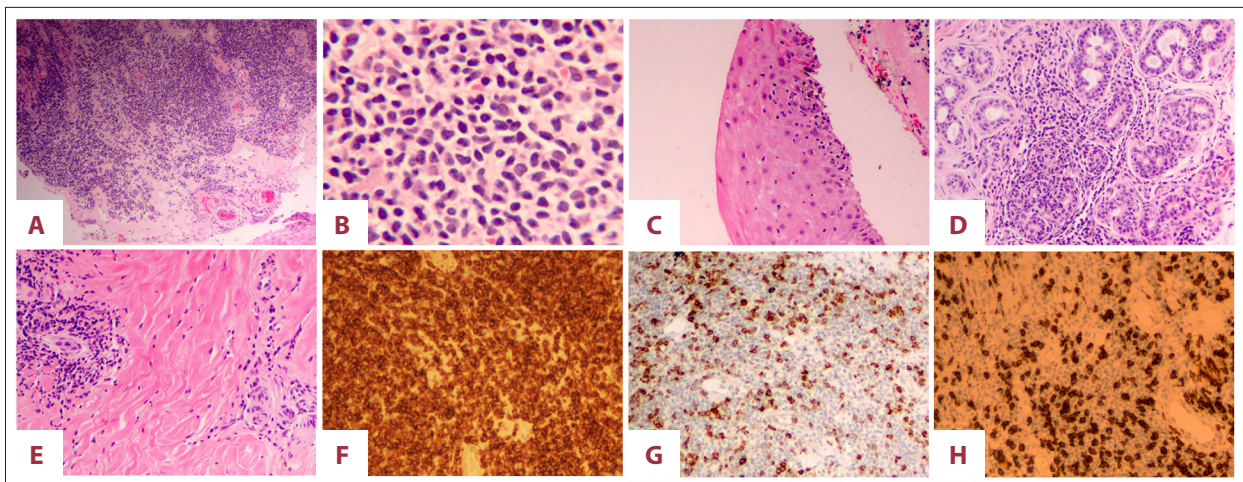


Figure 1. Histopathology and immunostaining profiles of mycosis fungoides palmaris et plantaris. (A) An infiltrate of the atypical lymphocytes in the upper dermis (hematoxylin and eosin [H&E], 40×1). (B) Round or ovoid atypical lymphocytes with cerebriform nuclear contour and no clear nuclear membrane or nucleoli (HE, 400×1). (C) Atypical lymphocytes infiltrating into the epidermis (epidermotropism) (HE, 100×1). (D) Atypical lymphocytes infiltrating into eccrine sweat glands (syringotropism) (HE, 100×1). (E) Perivascular infiltration of the atypical lymphocytes in the dermis (HE, 100×1). (F) Strong CD4 expression in atypical lymphocytes (3, 3 -diaminobenzidine [DAB], 100×1). (G) Reduced CD7 expression in atypical lymphocytes (DAB, 100×1). (H) Reduced CD8 expression in atypical lymphocytes (DAB, 100×1).

the patient's general health; and available medical resources. Therapy should be adjusted according to patient response and any adverse effects of the chosen treatment.

Conclusions

MFPP is a rare variant of MF with an indolent course. It is histologically characterized by epidermotropism of atypical convoluted lymphocytes and infiltration of atypical lymphocytes

in the upper dermis. Treatment options include topical or systemic chemotherapy, PUVA photochemotherapy, radiation therapy, and immunotherapy, which result in complete or partial remission in most patients. Periodic follow-up visits are needed because of the potential for recurrence or spread of MFPP to other areas of the body.

Conflicts of Interest

None.

References:

1. Resnik KS, Kantor GR, Lessin SR et al: Mycosis fungoides palmaris et plantaris. *Arch Dermatol*, 1995; 131(9): 1052–56
2. Larocca C, Kupper T: Mycosis fungoides and Sezary syndrome: An update. *Hematol Oncol Clin North Am*, 2019; 33(1): 103–20
3. Nakai N, Hagura A, Yamazato S, Katoh N: Mycosis fungoides palmaris et plantaris successfully treated with radiotherapy: Case report and mini-review of the published work. *J Dermatol*, 2014; 41(1): 63–67
4. Tanita K, Fujimura T, Kakizaki A et al: Periostin in the cancer stroma of mycosis fungoides palmaris et plantaris: A case report and immunohistochemical study. *Case Rep Dermatol*, 2016; 8(1): 31–35
5. Pham AK, Carter JB, Ratcliffe NR et al: Tumor-stage mycosis fungoides in palmoplantar localization with large-cell transformation and partial CD30 expression shows complete response to brentuximab vedotin. *J Cutan Pathol*, 2018; 45(6): 458–62
6. Doukaki S, Arico M, Bongiorno MR: A rare presentation of mycosis fungoides mimicking psoriasis vulgaris. *Case Rep Dermatol*, 2009; 1(1): 60–65
7. Toritsugi M, Satoh T, Higuchi T et al: A vesiculopustular variant of mycosis fungoides palmaris et plantaris masquerading as palmoplantar pustulosis with nail involvement. *J Am Acad Dermatol*, 2004; 51(1): 139–41
8. Wong HK, Mishra A, Hake T, Porcu P: Evolving insights in the pathogenesis and therapy of cutaneous T-cell lymphoma (mycosis fungoides and Sezary syndrome). *Br J Haematol*, 2011; 155(2): 150–66
9. Kim ST, Jeon YS, Sim HJ et al: Clinicopathologic features and T-cell receptor gene rearrangement findings of mycosis fungoides palmaris et plantaris. *J Am Acad Dermatol*, 2006; 54(3): 466–71
10. Sandwich JT, Davis LS: Mycosis fungoides palmaris et plantaris. *Arch Dermatol*, 1996; 132(8): 971
11. Lambert TJ, Prieto VG, Duvic M: Multiple plaques on the hands and feet. Mycosis fungoides (MF) palmaris et plantaris (MFPP). *Arch Dermatol*, 2007; 143(1): 109–14
12. Goldberg DJ, Stampien TM, Schwartz RA: Mycosis fungoides palmaris et plantaris: Successful treatment with the carbon dioxide laser. *Br J Dermatol*, 1997; 136(4): 617–19
13. Jin SP, Jeon YK, Cho KH, Chung JH: Excimer laser therapy (308 nm) for mycosis fungoides palmaris et plantaris: A skin-directed and anatomically feasible treatment. *Br J Dermatol*, 2010; 163(3): 651–53
14. Wang CM, Duvic M, Dabaja BS: Acral erosive mycosis fungoides: Successful treatment with localised radiotherapy. *BMJ Case Rep*, 2013; 2013: bcr2012007120
15. Olsen EA, Hodak E, Anderson T et al: Guidelines for phototherapy of mycosis fungoides and Sezary syndrome: A consensus statement of the United States Cutaneous Lymphoma Consortium. *J Am Acad Dermatol*, 2016; 74(1): 27–58