Mycosis Fungoides Masquerading as Acrodermatitis Continua of Hallopeau

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ABSTRACT

Herein, we present a case of long-standing mycosis fungoides of the digit that was previously misdiagnosed. We highlight the unusual presentation of the lesion, as well as its extreme response to focal radiation therapy. This case demonstrates the importance of early biopsies in lesions with mycosis fungoides on the differential. In addition, close monitoring during initial treatment of cutaneous T cell lymphoma (CTCL) can help patients in tolerance of therapy.

INTRODUCTION

Mycosis Fungoides (MF) is characterized by patches, plaques, and tumors of cutaneous T lymphocytes.¹ Though MF is the most common form of CTCL, it does not impact much of the population, and thus is not seen routinely by dermatologists.¹ MF has unique histological findings, however, it can mimic other more-common plaque-like pathologies such as psoriasis. This can lead to long-term misdiagnosis, such as with our patient.

CASE

A 70-year-old male, with history of Basal Cell Carcinoma of the arm and suspected plaque psoriasis, presented with a swollen, red and scaly left fifth digit. He noted that the finger had been particularly inflamed and painful for two months with a worsening course over several years. He endorsed frequent skin breakdown and weeping. On exam, yellow granulation tissue on an edematous base and serous crusting were noted.

About one year prior, the patient presented to an outside dermatologist. Punch biopsy was performed which demonstrated a lichenoid superficial infiltrate and deep lymphocytic reaction. An atypical mycobacterial infection was suspected, and oral minocycline was prescribed. The patient failed to improve on antibiotic therapy.

Approximately one year later, the patient presented to a wound care center for a second opinion. An additional punch biopsy was performed which led to increased swelling and tenderness in the digit. He was treated with amoxicillin for suspected cellulitis. At this time, he was referred to our clinic for further evaluation by both the dermatology and hematology/oncology teams. Relevant medical history included long-standing suspected plaque psoriasis.
On examination, erythematous, scaly plaques were observed on the bilateral upper and lower extremities. Two punch biopsies were performed- one of the left fifth digit and the other of a left forearm plaque. Both specimens demonstrated atypical lymphoid infiltrates with epidermotropism, favoring a diagnosis of mycosis fungoides. Infiltrates were predominantly CD3+ with rare CD20+ B cells. The differential diagnosis included mycosis fungoides, peripheral T-cell lymphoma, and primary cutaneous CD4+ small/medium T-cell proliferative disorder. Whole body positron emission tomography/computed tomography (PET/CT) was ordered as well as T-cell receptor (TCR) gene rearrangement studies for both tissue and blood. PET/CT showed a hypermetabolic left fifth finger mass and a nonspecific hypermetabolic cutaneous/subcutaneous metabolic activity along the right anterior distal calf/ankle. Peripheral blood was negative for involvement by cutaneous T-cell lymphoma. Tissue TCR gene rearrangement of the left fifth digit biopsy site showed mild staining of the lymphocytes for beta and rare staining for delta. Tissue TCR gene rearrangement of the left forearm plaque was negative for beta and delta. Radiation oncology was consulted.

The patient was referred to radiation oncology for focal radiation to both the left 4th and 5th digit as well as the anterior right distal ankle. He underwent the first series of focal radiation treatments shortly thereafter.

Approximately one month later the patient began his second half of radiation treatments. A few weeks after receiving his tenth and final treatment, the patient was seen in cutaneous lymphoma clinic. He was immediately advised to go to the emergency room due to worsening digital pain and concern for osteomyelitis. Crusting and necrosis of the finger were noted. Complete blood count (CBC), lactic acid testing and a hand x-ray were performed. CBC and lactic acid were both within normal limits and the left-hand x-ray was negative for osteomyelitis. Viral cultures were negative for HSV, CMV, and VZV. Wound cultures were positive for enterobacteria, and the patient was treated with ciprofloxacin.

Plastic surgery was consulted, and wound debridement was not recommended. An MRI hand was ordered and was negative for acute bone marrow edema or enhancement but did show infiltrative circumferential soft tissue thickening of the left fifth digit as well as soft tissue edema throughout the rest of the left hand. He was prescribed a repeat course of ciprofloxacin 500 mg BID for seven days along with recommendations to apply hydrogen peroxide and polysporin to the digit.

Two weeks after the consultation with plastics he had marked improvement of his left fifth digit (Figure 1). Peginterferon alfa-2a 180 mcg weekly was recommended for maintenance therapy to control his mycosis fungoides.
This case highlights the importance of early and repeat biopsies when there is clinical concern for CTCL, especially when lack of clinical improvement is noted with traditional therapies (i.e. corticosteroids for psoriasis). This patient had been misdiagnosed with psoriasis for years. Performing multiple biopsies earlier in the disease course, as well as referral to an academic center, could have allowed for earlier diagnosis and treatment. In addition, it is possible that with earlier diagnosis, our patient may have had further treatment options available to him, discounting the need for radiation and its side effects. Key histological differences between psoriasis and mycosis fungoides allow distinction between the two diseases. Pautrier microabscesses, or intraepidermal inclusions of malignant lymphocytes are characteristic of mycosis fungoides. In contrast, Munro microabscesses, or intraepidermal inclusions of neutrophils, are characteristic of psoriasis. Most mycosis fungoides lesions exhibit CD4 positivity, with CD8 positivity only seen in a minority of cases (i.e. hypopigmented variant). Thus, T-Cell monoclonality testing can be used to differentiate between psoriasis and CTCL. On exam, mycosis fungoides typically presents as pruritic patches/plaques/tumors primarily in non-sun exposed areas. Rarely, will it present as symmetric, well-demarcated lesions, this being more consistent with psoriasis. Involvement with lymph nodes, though rarely associated with psoriasis, is typically an indication of CTCL.

Patient Perspective

The patient had an active role in their care, taking photographs throughout their radiation treatments and sharing them with our team. Though the lesion’s reaction to the radiation was concerning to the patient, the patient appreciated the outcome after allowing time to heal.

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