

## BRIEF ARTICLE

**A Man with Worsening Scaling Plaques and a New Onset Large Draining Ulcerated Tumor**Hannah Pile, DO<sup>1</sup>, Alexandra Taylor, BS<sup>2</sup>, David Altman, MD<sup>1</sup><sup>1</sup> St. Joseph Mercy Hospital, Ann Arbor, MI<sup>2</sup> Campbell University of Osteopathic Medicine, Lillington, NC

## ABSTRACT

Mycosis Fungoides (MF) is the most common form of cutaneous T-cell lymphoma, a rare condition which typically presents with erythematous patches and plaques, skin nodules and/or tumors. Advanced disease may progress to a more aggressive form known as a large-cell transformation (LCT). MF may be easily confused for psoriasis, a deep fungal infection, and pyoderma gangrenosum. The name “mycosis fungoides” is misleading as the disease is not due to a fungal infection. Psoriasis and MF share common features associated with the abnormal functioning of T cells, and in the absence of an infectious process, pyoderma gangrenosum (PG) may be considered. Definitive diagnosis relies upon histopathology revealing characteristic features of MF including cerebriform nuclei, intraepidermal Pautrier microabscesses, epidermotropism and haloed lymphocytes, ruling out the other etiologies. Here is a case of a 62-year-old male with an atypical case of MF with LCT who presented with worsening skin eruption and a large draining ulcerated abdominal plaque. MF is typically a slow and indolent disease with multiple treatment options. Brentuximab vedotin (BV) is a monoclonal antibody targeting the CD30 antigen on cancer cells and is increasingly used in the treatment of MF patients with LCT. The patient was referred to hematology/oncology where further workup revealed CD30 positive large T-cells. He was started on BV and has already shown signs of significant disease regression.

## INTRODUCTION

Mycosis fungoides (MF) represents the most common form of cutaneous T-cell lymphoma, which is typically a progressive and chronic disease.<sup>1</sup> There are multiple stages that classify the severity of the disease, which can range from generalized itching, to skin tumors, and finally a fully disseminated disease.<sup>2</sup> MF may progress through a large-cell transformation, a more aggressive form conferring a worse prognosis.<sup>1</sup> The diagnosis

of MF is usually made with a variety of techniques including histology, immunophenotyping and T-cell receptor gene analysis.<sup>3</sup> Treatment is multidisciplinary, requiring hematology and oncology referral and possibly investigational therapies.<sup>2</sup>

## CASE REPORT

A 62-year-old male presented as a new patient for evaluation of a worsening eruption

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on his arms, face, legs, and trunk. The rash was described as red, blistering, painful and had been present for the past eight months. The patient also reported a periumbilical plaque that started as a red-violaceous nodule which progressed to a large draining ulcerated plaque over the past six months. Past medical history is significant for untreated psoriasis diagnosed 10-15 years prior. The patient has been washing his abdomen daily with soap and water and reported no cough, diarrhea, fevers, or joint aches. Physical examination revealed scattered erythematous oval patches and plaques on his bilateral upper and lower extremities and trunk, with variable overlying scale. On the lower abdomen there was a 20 cm periumbilical ulcerated indurated tumor with adherent yellow-green fibrinous drainage (**Figure 1**). The patient had significant cervical, axillary, and inguinal lymphadenopathy. Punch biopsies from the periumbilical area and right ventral forearm were sent for histopathologic examination (**Figure 2 and 3**).



**Figure 1.** Ulcerated abdominal tumor.

## Microscopic Findings and Clinical Course

Histopathologic examination demonstrated irregular epidermal hyperplasia with patchy spongiosis. There are dense diffuse infiltrates of large atypical epithelioid to reniform cells with interspersed small lymphocytes and frequent eosinophils in the superficial and deep dermis (**Figure 3**). The epidermis contained similar atypical cells, as well as eosinophils, as single units and small clusters. Focal ulceration and necrosis were also found within the lesions. A CD3 stain marked most of the large dermal cells (**Figure 2**). The patient was referred to hematology where further workup revealed CD30 positive large T-cells. The patient has been started on BV and has already shown signs of disease regression.

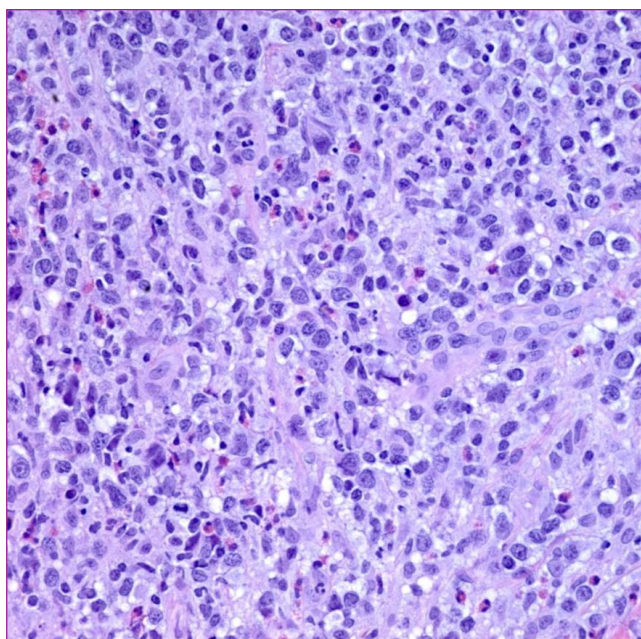


**Figure 2.** CD3 stain.

## DISCUSSION

Mycosis fungoides (MF) is the most common subtype of cutaneous T-cell lymphoma and is characterized by the evolution of patches, plaques, tumors, and in some individual's progression to a more aggressive form known as large-cell transformation (LCT).<sup>1,2</sup>

While LCT is very rare in early stage I of MF, it has been described in up to 50% of patients with stage IV disease.<sup>2</sup> MF with LCT typically confers a worse prognosis with 50% survival by 1 year compared to tumor stage MF without LCT, which has an average survival of 5.5 years.<sup>2</sup>



**Figure 3.** High power dermal infiltrate.

Classic MF presents with erythematous patches and/or plaques in the early stages, which may progress to thicker plaques, skin nodules and tumors.<sup>1</sup> These mushroom-like tumors can ulcerate and are typically 1 to 15 cm in diameter presenting with a bluish or red-brown hue.<sup>3</sup> While LCT is most likely to occur in the tumor stage of disease, it has also been associated in smaller papules with rapid progression of disease which may help aid in early detection of transformation.<sup>3</sup> The diagnosis of MF relies upon histology, immunophenotyping and T-cell receptor gene analysis.<sup>4</sup> Characteristic histologic features in MF include atypical mononuclear cells with cerebriform nuclei, intraepidermal Pautrier microabscesses, epidermotropism and haloed lymphocytes.<sup>3</sup> MF with LCT is defined histopathologically as >25% of the

population of cells being large (about 4 times the size of normal lymphocytes).<sup>3</sup> The cells may display other atypical features such as pleomorphism, lobulated nuclei, and CD30 positivity.<sup>2</sup>

Psoriasis and MF share common clinical and pathogenetic features associated with the abnormal functioning of T cells.<sup>4</sup> They are two distinct skin disorders that may coexist, present after one another or resemble each other, making differentiation increasingly difficult.<sup>5</sup> Additionally, repeated biopsies may not confirm MF for months to years. Disease confirmation often requires a thorough exam, histopathology and T cell monoclonality.<sup>4</sup> Psoriasis is histologically characterized by epidermal hyperplasia, parakeratosis, neutrophils infiltrating the stratum corneum and spinous layer, hypogranulosis, and suprapapillary plate thinning with papillary blood vessel dilation.<sup>6</sup> Lymphadenopathy and internal organ involvement are much more likely to be seen in MF than psoriasis.<sup>6</sup>

Tumor stage MF should be differentiated from other causes of indurated skin lesions. The name “mycosis fungoides” is misleading due to the fungal sounding nature of the name. However, MF is typically an indolent lymphoma with skin lesions that may resemble mushrooms.<sup>1</sup> Deep fungal infections may be diagnosed via PCR or culture.<sup>7</sup> Histologic evaluation of a skin biopsy would reveal a granulomatous process with fungal hyphae or yeast forms highlighted by periodic acid-Schiff or Grocott methenamine silver stains.<sup>7</sup> In ulcerated skin lesions a Gram-stain can be helpful at ruling out bacterial infections.<sup>7</sup> In the absence of an infectious process, pyoderma gangrenosum (PG) may be considered. While the infiltrate of PG may be robust, it’s typically neutrophilic and lack the monoclonality and atypical T cells appreciated in MF.<sup>8</sup>

MF is typically a slow and indolent disease with treatments including topical steroids, phototherapy, topical nitrogen mustard, methotrexate, interferons, oral retinoids, photopheresis and radiation.<sup>1</sup> LCT of MF typically displays a more aggressive disease course and confers a poor prognosis requiring a multidisciplinary treatment approach with referral to hematology/oncology.<sup>2</sup> BV, an anti-CD30 monoclonal antibody, is an ideal treatment in CD30 positive MF patients with LCT.<sup>9</sup> Overall BV is well tolerated when compared to other systemic and radiation therapies with the most common side effect being peripheral sensory neuropathy.<sup>9</sup>

## CONCLUSION

While cutaneous manifestations are the hallmark of MF, a large draining ulcerated abdominal plaque is not a common presentation. This case runs through the appropriate diagnosis modalities and histopathological findings for MF with LCT, in addition to ruling out other common etiologies. While prognosis is typically poor at later stages of disease, treatment with BV may slow disease progression in patients with these specific tumor markers.

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