Our Dermatology Online

Case Report

Large, rapidly growing, ulcerated tumor in the abdomen

Jorge González-Torres[1], Amparo Hernández-Salazar[1], Betzabe Quiles-Martínez[1], Judith Domínguez-Cherit[1], Daniel Montante-Montes[2]

[1]Dermatology Department, National Institute of Medical Sciences and Nutrition "Salvador Zubirán", Mexico City, Mexico, [2]Pathology Department, National Institute of Medical Sciences and Nutrition "Salvador Zubirán" Mexico City, Mexico

Corresponding author: Jorge González-Torres, MD, E-mail: jorgeagtorres2019@gmail.com

ABSTRACT

Mycosis fungoides is the most common primary T-cell tumor, usually CD4+. Clinically, three stages are described: patch, plaque, and tumor. There are rare variants such as granulomatous lymphoma, accounting for less than 1.8% of the cases. Herein, we present the case of a 27-year-old male, previously healthy, having a large tumor on the abdominal wall, for which the diagnosis was delayed for one year, with an especially poor response to chemotherapy treatments, and with transformation to large cell mycosis fungoides.

Key words: Mycosis fungoides, Cutaneous lymphoma, Hospital Dermatology

INTRODUCTION

Mycosis fungoides is the most common primary T-cell tumor, usually CD4+. Clinically, three stages are described: patch, plaque, and tumor. The World Health Organization and the European Organization for Research in Cancer Therapy recognize three variants of classic mycosis fungoides: pagetoid reticulosis, folliculotropic, and granulomatous lax skin [1]. However, there are other varieties such as granulomatous lymphoma, which is a rare variant, accounting for less than 1.8% of primary cutaneous lymphomas [2], which may present at initial diagnosis or years after the classical form [1]. Herein, we present a case of this atypical variety.

CASE REPORT

This was a 27-year-old patient, who was previously healthy, native of a rural area in Mexico. One year prior to the admission, he reported an erythematous dermatosis that was treated as cellulitis without a response, which grew rapidly over six months. An ultrasound was performed in the patient's birthplace, which revealed a soft tissue tumor. A skin biopsy was performed and Rosai–Dorfman disease was diagnosed. An additional biopsy of the tumor revealed non-Hodgkin's T-cell lymphoma. In view of the presence of lymphadenopathy, a biopsy of the inguinal lymph node was performed, which revealed chronic non-crossing granulomatous lymphadenitis and sinusoidal histiocytosis. During this period, the patient experienced occasional nocturnal diaphoresis and unintentional weight loss of 8 kg. When he was examined at our institution, he had a skin dermatosis located on the trunk, unilateral, affecting the abdomen in the lower right quadrant, characterized by a large exophytic cupuliform neoformation measuring 20 x 20 cm, with two ulcers on its surface, one measuring 10 cm and the other 3 cm, both with irregular, hyperpigmented, dark brown, undermined edges, with purulent secretion on the periphery and a dry-looking discharge in the center. The rest of the tumor had a soft consistency, with irregular, firm, ill-defined edges, brown erythematous hyperpigmentation on the surface, and areas of atrophy. The tumor led to pain and reduced

How to cite this article: González-Torres J, Hernández-Salazar A, Quiles-Martínez B, Domínguez-Cherit J, Montante-Montes D. Large, rapidly growing, ulcerated tumor in the abdomen. Our Dermatol Online. 2024;15(2):167-170.

Submission: 08.10.2023; **Acceptance:** 21.01.2024
DOI: 10.7241/ourd.20242.15

ability to walk (Fig. 1). A skin biopsy was performed where, on H&E staining, atypical, small to medium-sized lymphocytes, histiocytes, and multinucleated giant cells were observed (Figs. 2a and 2b), with immunohistochemistry, with CD3+ (Fig. 3a) and CD20- (Fig. 3b), in the characterization of these lymphocytes with CD2+ (Fig. 3c), CD4+ (Fig. 3d), CD5+ (Fig. 3e) markers, and focally in histiocytes and CD68+ multinucleated giant cells (Fig. 3f), and CD8- (Fig. 3g). The pathology service reported granulomatous mycosis fungoides in the tumor phase.

In order to establish the stage, a PET-CT scan was performed. It showed an infiltrative lesion in the soft tissues of the right abdominal wall with associated hypermetabolism and hypermetabolic right axillary lymph nodes (Fig. 4), for which an axillary node biopsy was performed. The pathology report indicated infiltration by granulomatous mycosis fungoides with partial involvement ISCL/EORTC (International Society for Cutaneous Lymphomas and the Cutaneous Lymphoma Task Force of the European Organization of Research and Treatment of Cancer) NE/category 3 DUTCH system and no involvement was reported in the bone marrow aspirate revealed no abnormalities.

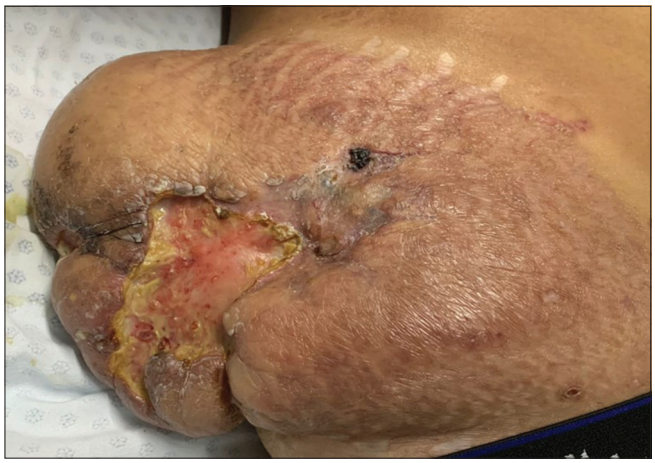

Figure 1: Dermatosis.

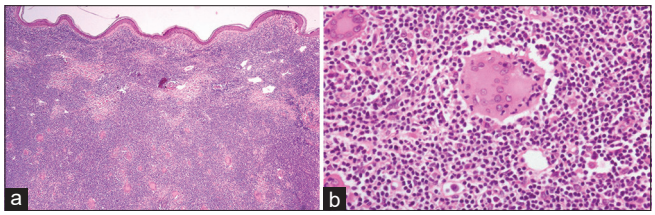

Figure 2: (a) Diffuse infiltrate of basophilic cells and multinucleated giant cells (H&E; 40x). (b) Magnification cells of intermediate to small size with blastic-looking chromatin with periadnexal involvement and even adipose tissue (H&E; 400x).

Based on these findings, the subject was classified as being in stage IVB.

The hematology and lymphoma department began treatment with gemcitabine for eight cycles, with little response and adverse reactions to chemotherapy (diarrhea, fever, tubulointerstitial necrosis) and malignant hypercalcemia. Due to the partial response, treatment with brentuximab, adriamycin, and cyclophosphamide was initiated with the aim of reducing the tumor load. One year after beginning the treatment, the patient manifested a new disseminated dermatosis on the trunk and four extremities, characterized by oval infiltrated plaques measuring 0.5 to 1.5 cm, erythematous, brown, with fine scaling on the surface, pruritic, with an evolution of one month (Fig. 5), in addition to the persistence of the initial lesion. A new biopsy was performed, which showed transformation to large cell mycosis fungoides with an immunophenotype CD3+, CD20-, CD2+, CD4+, CD5+, CD8-, CD56-, CD30 (60%). Treatment with brentuximab is currently awaiting initiation.

DISCUSSION

Clinical features of granulomatous mycosis fungoides are generally atypical. Patches or plaques may be absent [3]. Some features that may suggest this form of presentation are thick plaques or nodules, no scaling and no laxity. It may mimic granuloma annulare, sarcoidosis, or granulomatous rosacea. Other reported forms include psoriasiform or dermatofibroma-like plaques [3] and morphea-like plaques [2]. The form of presentation in this case was atypical, as it presented as a large, subcutaneous, exophytic tumor with central ulceration, with no previous patch or plaque stage. The initial presumptive diagnosis was sarcoma. The biopsy is characterized by CD4+ T-cell infiltrate, in different patterns such as lichenoid, perivascular, perivascular, and interstitial. Up to 33% lymphocytic infiltrate or nuclear atypia may be absent. If epidermotropism is observed, it is moderate and focal. Multinucleated giant cells, histiocytes, and granuloma formation are found. Immunohistochemistry is dominated by CD4+ cell population, CD4: CD8 > 4:1 and 50% loss of CD7 expression [2]. Histological criteria include a prominent granuloma formation of any type, with histiocytes and multinucleated giant cells. Loss of elastic fibers is common, yet elastophagocytosis is rare [1]. Histology of this tumor showed abundant histiocytes, multinucleated giant cells, and atypical lymphocytes.

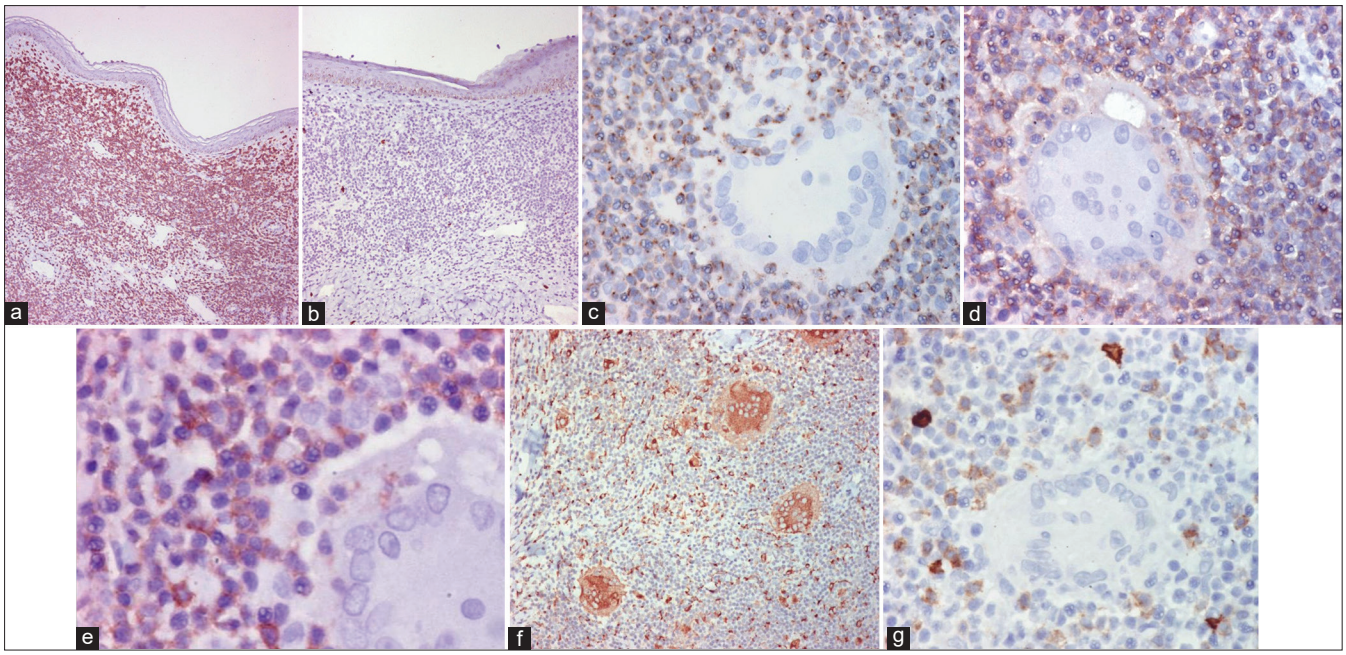

Figure 3: (a) Immunohistochemistry, CD3 intensely positive. (b) Immunohistochemistry, CD20 negative. (c) Immunohistochemistry, CD2 positive. (d) Immunohistochemistry, CD4 positive. (e) Immunohistochemistry, CD5 positive. (f) Immunohistochemistry, CD68 positive for multinucleated giant cells and interstitial histiocytes. (g) Immunohistochemistry, CD8 negative.

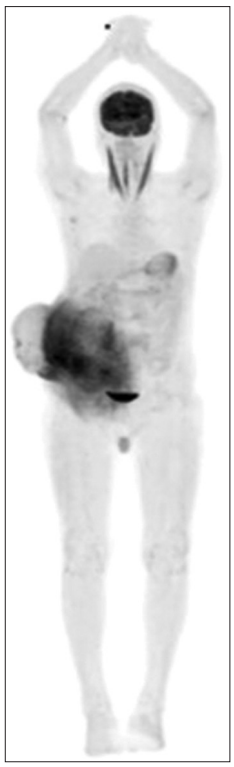

Figure 4: Hypermetabolism in the right abdominal wall and axillary adenopathies (PET 18F-FDG).

Immunohistochemistry was as expected. Diagnosis may be difficult because epidermotropism may be absent and because of the predominance of granulomatous infiltrate [1], as in this case, the initial biopsies were

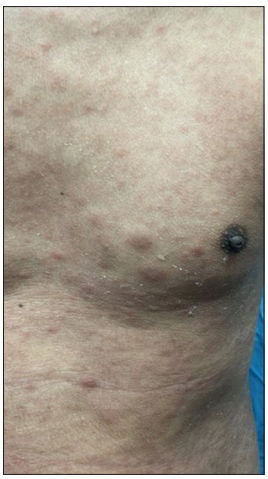

Figure 5: Progression: oval, erythematous, brown plaques with fine scaling on the surface.

diagnosed with granulomatous disease and Rosai–Dorfman disease. This showed how important it is that the diagnosis is made by pathologists with experience in this type of disease. Atypical presentations show a more rapid progression, worse prognosis, or transformation to cutaneous or extracutaneous large cell lymphoma [3], as in this case. The five-year survival rate is 66% in this form of presentation, with progression up to 46% [2]. This patient had a poor response to the first-choice treatment, gemcitabine. When the patient manifested a new disseminated dermatosis, the histology showed an infiltrate with more than 25% of cells of large

size and has now transformed to large cell mycosis fungoides, with a worse prognosis. This transformation is defined as more than 25% predominance of large cells with oval or irregular nuclei and prominent CD30 (+/-) nucleoli [4]. 50% occur in the tumor phase, and in 66%, the transformation occurs in the skin [5], as in this case. Overall survival in this phase is 12 to 20 months, and there is a mortality rate of 40% [4]. The patient is currently awaiting treatment with brentuximab.

CONCLUSION

Herein, we have presented an exceptional case due to the initial clinical presentation as a giant tumor in the abdomen, which progressed despite treatments. This form of presentation, which is rare, confers a worse prognosis.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

REFERENCES

1. Hodak E, Amitay-Laish I. Mycosis fungoides: A great imitator. Clin Dermatol. 2019;37:255-67.
2. Lenskaya V, de Moll EH, Hussein S, Phelps RG. Subtlety of granulomatous mycosis fungoides: A retrospective case series study and proposal of helpful multimodal diagnostic approach with literature review. Am J Dermatopathol. 2022;44:559-67.
3. Sidiropoulou P, Tsaoutou K, Constantinou A, Marinos L, Voudouri D, Iliakis T, Kanellis G, Pouliou E, Stratigos A, Nikolaou V. New insights into granulomatous mycosis fungoides (GMF): A single-center experience. Eur J Cancer. 2021;156 Suppl 1:S69-S70.
4. Vural S, Akay BN, Botsalı A, Atilla E, Parlak N, Okçu Heper A, Şanlı H. Transformation of mycosis fungoides/Sezary syndrome: Clinical characteristics and prognosis. Turk J Haematol. 2018;35:35-41.
5. Gómez Díez S, Fresno Forcelledo MF, Raya Aguado C, Vázquez López F, Rodríguez Vigil T, González López M, Rodríguez Prieto MA, Pérez Oliva N. Transformation of mycosis fungoides/Sézary syndrome to large cell lymphoma. Actas Dermo-Sifiliográficas Vol. 92. No. 9. Pages 391-396 (September 2001).

Source of Support: This article has no funding source.

Conflict of Interest: The authors have no conflict of interest to declare.