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BRIEF ARTICLES

Subcutaneous Panniculitis-Like T-cell Lymphoma: A Case Report

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ABSTRACT

We present a case of a woman diagnosed with subcutaneous panniculitis-like T-cell lymphoma treated with prednisone and methotrexate with sustained symptom remission, suggesting that uncomplicated subcutaneous panniculitis-like T-cell lymphoma can be managed effectively without multiagent chemotherapy or radiation.

INTRODUCTION

Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) is an unusual cutaneous T-cell lymphoma (TCL) accounting for <1% of non-Hodgkin lymphomas which typically presents with subcutaneous nodules or deep plaques. We present the case of a woman diagnosed with SPTCL successfully treated with oral corticosteroids and methotrexate.

REPORT OF A CASE

A woman in her 60s presented with a 4-month history of numerous painful 4-8cm subcutaneous nodules on the cheeks, arms, breasts, and buttocks (Figure 1a). Her past medical history was unremarkable. Review of symptoms was notable for fever, fatigue, and a 60-pound unintentional weight loss.

Biopsy of a lesion demonstrated a dense atypical lymphoid infiltrate in the subcutis (Figure 2) that was CD3+, CD8+, CD56-, reactive for TIA, perforin, granzyme B, and BF1, with rare CD20+ cells. Tissue cultures and stains were negative. Laboratory studies were notable for elevated dehydrogenase (556 U/L), mildly elevated transaminases, and mild pancytopenia. Flow cytometry analysis of peripheral blood was unrevealing. Positron Emission Tomography demonstrated numerous subcutaneous nodules in her cheeks, arms, neck, breasts, mesentery, mediastinum, back. and abdominal cavity without lymphadenopathy or hepatosplenomegaly.

The patient was diagnosed with SPTCL and started on oral methotrexate and a prednisone taper with atrophy of her subcutaneous nodules and resolution of her elevated transaminases, cytopenias, and symptomatic improvement. She was continued on 15mg weekly methotrexate with

January 2020 Volume 4 Issue 1

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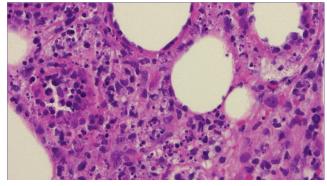
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sustained symptom remission 4 months after diagnosis (Figure 1b).

Figure 1. (a) Subcutaneous nodules on the bilateral breasts at presentation (b) and after four months of treatment with methotrexate and a prednisone taper



Figure 2. A dense, lobular lymphocytic panniculitis with rimming of the adipocytes by atypical T-cells. The lymphocytes demonstrate nuclear atypia, there is adipocyte membrane crowding and disruption, and cellular fat necrosis. 40x magnification. Hematoxylin and eosin staining.



DISCUSSION

SPTCL is an uncommon TCL characterized by subcutaneous nodules that may be scattered across the body. Cytologically, it is defined by a T-cell infiltrate expressing an $\alpha\beta$ T-cell receptor phenotype and is typically characterized by a CD4-, CD8+, CD56-, BF1-lymphoid infiltrate. These lymphoid infiltrates are typically confined to subcutaneous tissue. $^{1-3}$

SPTCL has been associated with underlying autoimmune and rheumatologic disorders such as lupus erythematosus or rheumatoid arthritis in 20-40% of patients. Associated

symptoms may include fevers, chills, weight loss, fatigue, and myalgia. Laboratory findings may include cytopenias and elevated liver function tests, and radiographic studies may demonstrate lymphadenopathy and hepatosplenomegaly. It may be accompanied by hemophagocytic syndrome (HPS) in approximately 20% of patients, which portends a notably worse prognosis.³

Though traditional therapy for SPTCL has centered multiagent combination on chemotherapy, such as with CHOP (i.e. Cyclophosphamide, Adriamycin, Vincristine, and Prednisone) or CHOP-like regimens.4 these treatments are associated with significant morbidity, often surpassing that of the disease itself. Thus, there has been a renewed push to induce disease remission with less toxic and immunosuppressive regimens such as systemic corticosteroids, methotrexate, and other more common immunosuppressive rather than chemotherapy regimens.² Response rates similar to that of traditional chemotherapy, with the majority of patients achieving complete disease remission with immunosuppressive regimens such systemic corticosteroids, sparing patients the side effects cytotoxic of traditional chemotherapy. 1,5 Multiagent chemotherapy may be reserved for patients who do not respond to systemic corticosteroids or patients with HPS, with or without stem cell transplantation. Radiotherapy may also be considered in cases of solely local disease.⁴

Conflict of Interest Disclosures: None

Funding: None

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January 2020 Volume 4 Issue 1



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References:

- Willemze R, Jansen PM, Cerroni L, et al. Subcutaneous panniculitis-like T-cell lymphoma: definition, classification, and prognostic factors: an EORTC Cutaneous Lymphoma Group Study of 83 cases. *Blood*. 2008;111(2):838-845. doi:10.1182/blood-2007-04-087288
- Parveen Z, Thompson K. Subcutaneous Panniculitis-like T-Cell Lymphoma: Redefinition of Diagnostic Criteria in the Recent World Health Organization–European Organization for Research and Treatment of Cancer Classification for Cutaneous Lymphomas. *Arch Pathol Lab Med*. 2009;133(2):303-308. doi:10.1043/1543-2165-133.2.303
- 3. Swerdlow S, Campo E, Harris N, Jaffe E, Pileri S, Stein H. WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues, Fourth Edition. Lyon, France: IARC; 2008.
- Go RS, Wester SM. Immunophenotypic and molecular features, clinical outcomes, treatments, and prognostic factors associated with subcutaneous panniculitis-like T-cell lymphoma. *Cancer*. 2004;101(6):1404-1413. doi:10.1002/cncr.20502
- Guenova E, Schanz S, Hoetzenecker W, et al. Systemic corticosteroids for subcutaneous panniculitis-like T-cell lymphoma. *Br J Dermatol*. 2014;171(4):891-894. doi:10.1111/bjd.13053