CASE REPORT

Remission of subcutaneous panniculitis-like T-cell lymphoma in a pregnant woman after treatment with oral corticosteroids as monotherapy

Emily S. West, MD, Kanade Shinkai, MD, PhD, Weiyun Z. Ai, MD, PhD, and Laura B. Pincus, MD

San Francisco, California

Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) is a rare form of cutaneous T-cell lymphoma characterized by neoplastic α/β T cells infiltrating subcutaneous tissues in a lobular pattern. Few data support the optimal treatment regimen for patients, given the rarity of this condition, and even fewer data describe treatment when diagnosed during pregnancy. We describe a case of SPTCL in a pregnant patient who achieved clinical remission after treatment with corticosteroid monotherapy. Our case suggests that corticosteroids should be considered as first-line treatment in pregnant patients with SPTCL. (J Am Acad Dermatol 2017;3:87-9.)

Key words: corticosteroid; cutaneous lymphoma; immunosuppression; malignancy; pregnancy; T-cell lymphoma.

INTRODUCTION

Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) is a rare form of cutaneous T-cell lymphoma characterized by neoplastic α/β T cells infiltrating subcutaneous tissues in a lobular pattern. Patients typically have indurated and sometimes erythematous plaques and nodules, often on the lower extremities, although the upper extremities and the face can be involved. Prognosis is generally favorable with cumulative survival at 5 years approaching 82%. There is no standardized treatment protocol for SPTCL, and even fewer data guide treatment for patients diagnosed during pregnancy.

CASE REPORT

A 20-year-old woman who was 18 weeks pregnant had an erythematous subcutaneous nodule on her right thigh. Six weeks after onset, fever and tachycardia developed, and the patient was hospitalized. Physical examination at that time found several indurated subcutaneous erythematous plaques and nodules on her trunk and thigh ranging in size between 2 and 15 cm in diameter (Fig 1).

Laboratory investigations found leukopenia (3.9 × 10^9/L; normal range, 4.5–13.2 × 10^9/L) and anemia (7.7 g/dL; normal range, 12–15.5 g/dL). ANA, anti-double-strand deoxyribonucleic acid, anti–SS-A, and anti–SS-B antibodies were not detected. Blood and tissue cultures from a skin biopsy found no growth. Results of a purified protein derivative test for tuberculosis were negative. Bone marrow biopsy and flow cytometry from peripheral blood and marrow results were normal.

Skin biopsy from a subcutaneous nodule on her right abdomen found a dense infiltrate composed mostly of lymphocytes within the subcutaneous lobules with minimal epidermal and dermal involvement. In some foci, the lymphocytes appeared quite

From the Department of Dermatology, *Department of Medicine, Division of Hematology/Oncology, *and Department of Pathology, †University of California at San Francisco.

Funding sources: None.

Conflicts of interest: None declared.

Correspondence to: Emily S. West, MD, Department of Dermatology, University of California at San Francisco, 1701 Divisadero St. Room 4-20, San Francisco, CA 94115. E-mail: Emily.west2@ucsf.edu.

Abbreviation used:

SPTCL: Subcutaneous panniculitis-like T-cell lymphoma

2352-5126 © 2017 by the American Academy of Dermatology, Inc. Published by Elsevier, Inc. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

http://dx.doi.org/10.1016/j.jder.2016.12.007
crowded together. Some of the lymphocytes were atypical with pleomorphic nuclei, and scattered mitotic figures were evident among the lesional lymphocytes (Fig 2, A and B). Immunohistochemical staining showed a predominance of CD3⁺, CD4⁺, CD8⁺, TIA-1⁺ and BF1⁺ T lymphocytes, and these cells rimmed adipocytes in numerous foci. A GM3 stain did not show significant labeling among the lymphocytes rimming adipocytes. In situ hybridization for Epstein-Barr virus was negative. The proliferation rate among the lymphocytes rimming adipocytes was very high (approximately 70%) as assessed by a Ki-67 immunoperoxidase stain. Genotypic analysis of a skin biopsy found a clonal rearrangement of T-cell receptor γ chain genes.

Based on the high proliferation rate of lymphocytes rimming adipocytes, an α/β immunophenotype, and the clinical presentation, a subcutaneous
A panniculitis-like T-cell lymphoma was diagnosed. After discussion of treatment options with the obstetrics team, including combination treatment with oral corticosteroids and cyclosporine or oral corticosteroids alone, she started prednisone, 80 mg daily (1.17 mg/kg/d), monotherapy with resolution of fevers and cutaneous findings. Every 2 weeks the dose was tapered, and she continued 30 mg daily until delivery. She was in remission at the time of delivery and slowly tapered off prednisone over the next 4 months. She remained in remission at her most recent clinic visit 18 months after diagnosis.

DISCUSSION

There is no standardized treatment protocol for SPTCL, and there are only 4 reports of pregnant patients with SPTCL. Treatments used for pregnant patients reported in the literature include corticosteroids, methotrexate, chemotherapy, radiation therapy, and stem cell transplant (Table I). Two reported cases were fatal, one after treatment with corticosteroids then radiation therapy (case 1) and the other after treatment with corticosteroids, chemotherapy, and stem cell transplant (case 2). Neither of those patients was confirmed to have SPTCL with an α/β cytotoxic T-cell phenotype, and both patients had rapid disease progression and hemophagocytic syndrome, thus raising concern that they may have had γ/δ T-cell lymphoma. Two patients were confirmed to have SPTCL and both survived, although one (case 3) received chemotherapy and stem cell transplantation, and the other (case 4) was treated with prednisone and methotrexate immunosuppression. Our patient with SPTCL also had a favorable clinical outcome, achieving clinical remission after several months of oral corticosteroid monotherapy. Treatment with cyclosporine monotherapy or combination therapy was initially also considered but deferred given known excretion of cyclosporine in maternal breast milk, risks of hypertension and nephrotoxicity, and a laboratory monitoring requirement associated with cyclosporine use.

In light of the often indolent course and favorable prognosis of SPTCL, this case suggests that first-line treatment with systemic corticosteroids alone should be considered before combination therapy or alternative therapies such as cyclosporine, radiation therapy, or stem cell transplant in pregnant patients with SPTCL.

REFERENCES

1. 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.
2. Perniciaro C, Zalla MJ, White JW, Menke DM. Subcutaneous T-cell lymphoma: report of two additional cases and further observations. Arch Dermatol. 1993;129:1171-1176.
3. Romero LS, Goltz RW, Nagi C, et al. Subcutaneous T-cell lymphoma with associated hemophagocytic syndrome and terminal leukemic transformation. J Am Acad Dermatol. 1996;34:904-910.
4. Reimer P, Rudiger T, Muller J, et al. Subcutaneous panniculitis-like T-cell lymphoma during pregnancy with successful autologous stem cell transplantation. Ann Hematol. 2003;82(5):305-309.
5. Noble EL, Garry-McCoy A, Marks VJ, Tyler W. Subcutaneous panniculitis-like T-cell lymphoma in pregnancy. J Am Acad Dermatol. 2012;66(4):AB140. S1.
6. Moretti ME, Sgro M, Johnson DW, et al. Cyclosporine excretion into breast milk. Transplantation. 2003;75(12):2144-2146.

Table I. Literature review of cases of subcutaneous panniculitis-like T-cell lymphoma during pregnancy

| Case no. | Study            | Age, y | Pregnancy outcome | Therapy                              | Disease outcome                                  |
|----------|------------------|--------|-------------------|--------------------------------------|-------------------------------------------------|
| 1        | Perniciaro et al² | 21     | Delivery          | Corticosteroids, XRT                 | Death 2 months after XRT with disease progression |
| 2        | Romero et al³    | 22     | Spontaneous abortion | Prednisone, CHOP, mini-BEAM, ICE and autologous SCT | Death 6 weeks after ICE and SCT                 |
| 3        | Reimer et al⁴    | 35     | Delivery          | CHOP, ESHAP, myeloablative radiochemotherapy and autologous SCT | Remission sustained at follow-up 5 months after myeloablative radiochemotherapy |
| 4        | Noble et al⁵     | 31     | Delivery          | Prednisone, MTX                      | Recurrence 2 months after delivery               |
| 5        | Current study     | 20     | Delivery          | Prednisone                          | Remission after 5 months of treatment           |

CHOP, Cyclophosphamide, doxorubicin, vincristine, and prednisone; ESHAP, etoposide, prednisolone, cytarabine, cisplatin; ICE, ifosfamide, carboplatin, etoposide; mini-BEAM, Carmustine, etoposide, cytarabine, melphalan; MTX, methotrexate; SCT, stem cell transplant; XRT, external beam radiation therapy.