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

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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%.[1] There is no standardized treatment protocol for SPTCL, and even fewer data guide treatment for patients diagnosed during pregnancy.

Abbreviation used:

SPTCL: Subcutaneous panniculitis-like T-cell lymphoma

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 × 10E9/L; normal range, 4.5−13.2 × 10E9/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

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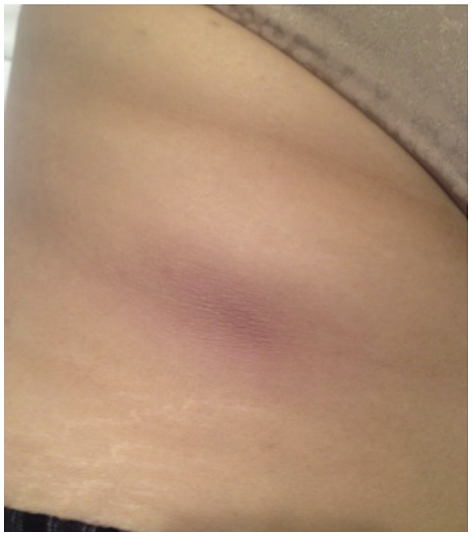

Fig 1. Subcutaneous panniculitis-like T-cell lymphoma. Right flank with tender indurated subcutaneous plaques and nodules with overlying erythema.

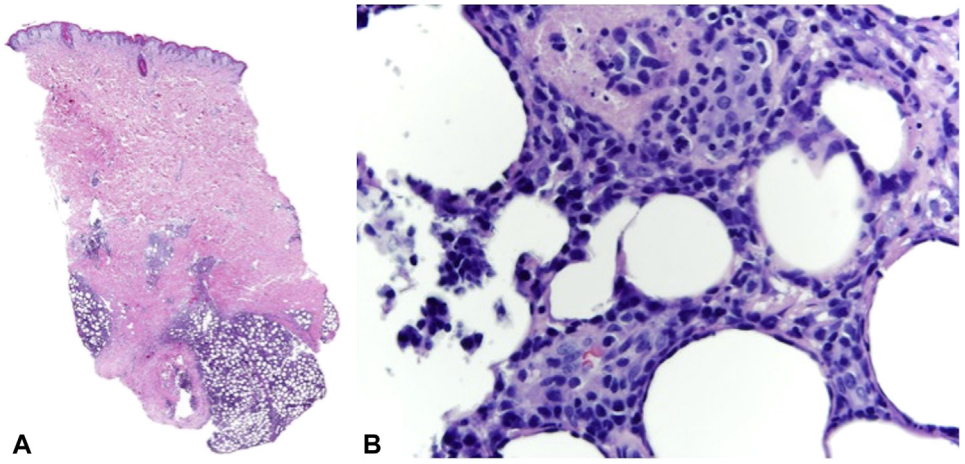

Fig 2. Subcutaneous panniculitis-like T-cell lymphoma. **A**, Low-power image shows a lymphocytic infiltrate centered on the subcutis with minimal alteration of the epidermis or dermis. **B**, Higher-power image shows that some of the lesional lymphocytes are large and have pleomorphic nuclei, (**A** and **B**, hematoxylin-eosin stain; original magnifications: **A**, ×20; **B**, ×200.)

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 $\beta F1^+$ 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

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 al[2]	21	Delivery	Corticosteroids, XRT	Death 2 months after XRT with disease progression
2	Romero et al[3]	22	Spontaneous abortion	Prednisone, CHOP, mini-BEAM, ICE and autologous SCT	Death 6 weeks after ICE and SCT
3	Reimer et al[4]	35	Delivery	CHOP, ESHAP, myeloablative radiochemotherapy and autologous SCT	Remission sustained at follow-up 5 months after myeloablative radiochemotherapy
4	Noble et al[5]	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.

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).[2-5] 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).[2,3] 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 chemoradiation and stem cell transplantation, and the other (case 4) was treated with prednisone and methotrexate immunosuppression.[4,5] 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,[6] 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.

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