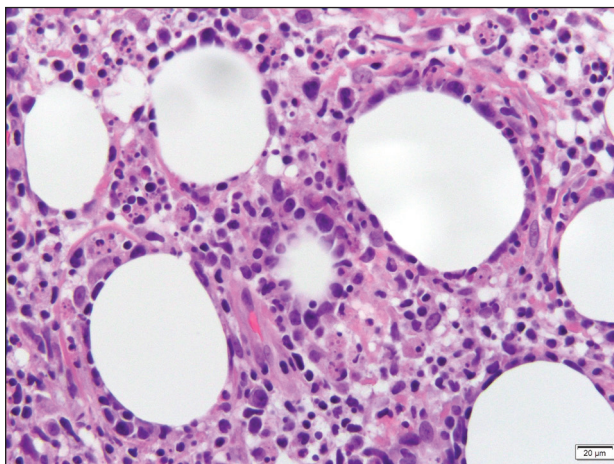


Tender Soft Tissue Mass on the Base of the Neck

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H&E, original magnification $\times 400$. Reference bar indicates 20 μm .

A 47-year-old man presented with a tender soft tissue mass on the upper back with increasing discomfort over the last 4 weeks. He noted that he felt feverish a few times. Physical examination revealed a 3 \times 4-cm area of induration involving the upper mid back with faint erythema of the overlying skin; no drainage was noted. A prominent left posterior cervical lymph node also was appreciated, and a punch biopsy of the mass was performed.

THE BEST DIAGNOSIS IS:

- erythema nodosum
- fat necrosis
- lipodermatosclerosis
- lupus erythematosus profundus
- subcutaneous panniculitislike T-cell lymphoma

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The authors report no conflict of interest.

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doi:10.12788/cutis.0155

THE DIAGNOSIS:

Subcutaneous Panniculitislike T-cell Lymphoma

Subcutaneous panniculitislike T-cell lymphoma (SPTCL) is a rare form of cutaneous lymphoma of mature cytotoxic T cells simulating panniculitis and preferentially infiltrating the subcutaneous tissue.¹ Subcutaneous panniculitislike T-cell lymphoma can affect all ages but predominantly affects younger individuals, with 20% being younger than 20 years.² It is a rare lymphoma that accounts for less than 1% of all non-Hodgkin lymphomas.³ It presents clinically as multiple subcutaneous masses, nodules, or plaques generally on the trunk or extremities.^{1,2} The skin surrounding the nodules may be erythematous, and the nodules may become necrotic; however, ulceration typically is not seen. Systemic symptoms such as fever, night sweats, and chills are present in half of cases.¹ According to the World Health Organization, cytopenia and elevated liver function tests are common, and a hemophagocytic syndrome may be present in 15% to 20% of cases.³ The presence of a hemophagocytic syndrome yields a poor prognosis.^{1,3} Current guidelines denote that SPTCL T-cell receptor (TCR) $\alpha\beta$ is a distinct entity from the TCR $\gamma\delta$ phenotype, known as cutaneous $\gamma\delta$ -positive T-cell lymphoma.^{3,4} Cutaneous $\gamma\delta$ -positive T-cell lymphoma is associated with rapid decline and a worse prognosis.⁴

Histology of SPTCL is characteristic for a lobular panniculitislike infiltrate.¹ The heavy subcutaneous lymphoid infiltrate is composed of atypical small- to medium-sized lymphocytes with mature chromatin and inconspicuous nucleoli lining adipocytes. The dense inflammatory infiltrate composed predominantly of neoplastic T cells and macrophages may diffusely invade into the subcutaneous tissue.¹ Admixed histocytes and karyorrhectic debris as well as rimming of the lymphocytes around the fat cells is typical and was seen in our patient (quiz image). The T cells of SPTCL have the following immunophenotype: TCR-beta F1⁺, CD3⁺, CD4⁻, CD8⁺, CD56⁻. They can express numerous cytotoxic proteins, such as T1a-1, granzyme B, and perforin.^{2,3} Although the CD8⁺ T cells may be sparse, they generally surround the adipocytes in a rimming manner and may distort the adipocyte membrane.¹

Lupus erythematosus profundus (LEP) is a form of chronic cutaneous lupus that affects the deep dermis and fat.⁵ It also can present clinically as tender plaques or nodules. It most frequently involves the upper arms, shoulders, face, or buttocks—areas that are less commonly involved in other panniculitides.⁶ Histologically, LEP is similar to chronic discoid lupus with features such as epidermal atrophy, interface changes, and a thickened basement membrane (Figure 1). Lupus erythematosus profundus can present as a lobular panniculitis with mucin as well as a superficial and deep lymphocytic infiltrate that can involve the septa.⁵ Some cases of LEP have a predominantly lobular lymphocytic panniculitis in the absence of the typical epidermal or

dermal changes of lupus erythematosus. Lymphoid follicles with germinal center formation are present in half of cases and reportedly are characteristic of LEP.^{6,7} The lymphoid follicles often have plasma cells, can extend into the septa as well as in between collagen bundles, and may have nuclear fragmentation.⁵ Another characteristic feature of LEP is hyaline sclerosis of lobules with focal extension into the interlobular septa. Immunofluorescence studies usually show linear deposition of IgM and C3 at the dermoepidermal junction. Antinuclear antibodies can be present in patients who have LEP but are not entirely specific.⁶

Lupus erythematosus profundus and SPTCL are part of a spectrum and may have overlapping clinical and histopathologic characteristics; therefore, distinguishing them may be difficult.⁶⁻⁸ It is important to monitor these patients closely, as their disease may progress to lymphoma.⁶ Patients with SPTCL are more likely to present with advanced symptoms such as fever and hepatosplenomegaly and to succumb to hemophagocytic syndrome than patients with LEP.⁹

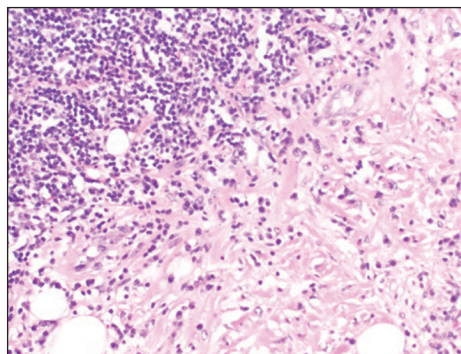


FIGURE 1. Lupus erythematosus profundus. A dense collection of lymphocytes in a lymphoid follicle with associated plasma cells (H&E, original magnification $\times 40$).

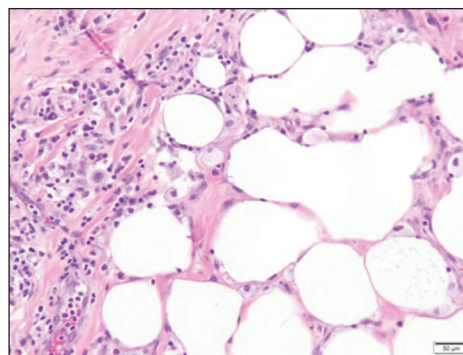


FIGURE 2. Fat necrosis. Lipid-laden macrophages along with chronic inflammatory cells (H&E, original magnification $\times 200$). Reference bar indicates 50 μm .

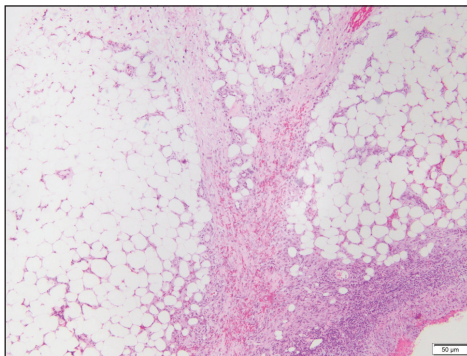


FIGURE 3. Erythema nodosum. Septal panniculitis with a mixed inflammatory background (H&E, original magnification $\times 40$). Reference bar indicates 50 μm .

Although SPTCL usually is clonal, several cases of LEP with clonality also have been described. Clonal LEP cases generally are identified in patients who present with fever and cytopenia.⁸ Lymphoid atypia and morphologic abnormalities may be seen in cases of LEP, further complicating the distinction between LEP and SPTCL. An elevated Ki67 level may be seen in cases of SPTCL with periadipocytic rimming.⁹ LeBlanc et al¹⁰ used Ki67 “hot spots” along with CD8 immunohistochemistry to identify atypical lymphocytes associated with SPTCL. Lymphocyte rimming was defined by the presence of CD8⁺ lymphocytes with an elevated Ki67 index. Clinical, histopathologic, and molecular findings all should be used when dealing with challenging cases.

Fat necrosis can occur in any part of the body where trauma has occurred and can be associated with many disease processes. Patients typically present with a palpable mass, but a clinical history of trauma is not always present. Histopathologic findings include necrotic fat alongside lipid-laden foamy macrophages and scattered inflammatory cells (Figure 2).¹¹ Fragments of normal as well as degenerating adipose tissue and multinucleated giant cells can be present.

Erythema nodosum (EN) is the most frequently encountered panniculitis and usually is seen in women in early adulthood.¹² Patients present with several tender subcutaneous nodules and plaques that most commonly are present on the anterior surface of the legs.^{12,13} Patients may have a constellation of symptoms including fever and leukocytosis, but the disorder generally is self-limited.¹² Erythema nodosum may be associated with a variety of diseases or infections including sarcoidosis, inflammatory bowel disease, and malignancy.¹⁴ The etiology of EN is diverse; therefore, a proper clinical workup may be necessary. Histopathology is that of a septal panniculitis with lymphocytes, histiocytes, and occasional eosinophils (Figure 3).¹³

Lipodermatosclerosis also occurs on the legs, most commonly in patients with venous insufficiency.^{12,15} Patients present clinically with pain, induration, redness, or swelling of the legs. Histopathology predominantly is

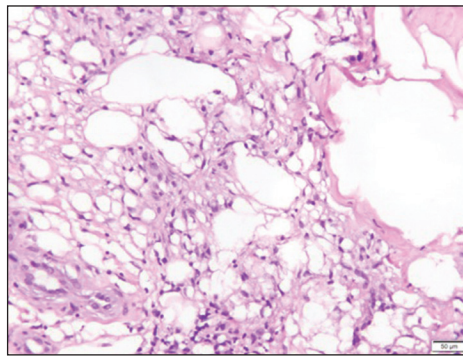


FIGURE 4. Lipodermatosclerosis. Membranous fat necrosis with cystic cavities lined by a hyaline membrane (H&E, original magnification $\times 200$). Reference bar indicates 50 μm .

characterized by membranous fat necrosis, fibrosis, and fatty microcysts that may be lined by a thickened hyaline membrane (Figure 4). Lipodermatosclerosis lesions generally do not resolve spontaneously and may need to be treated.¹⁶

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