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Case Report

Subcutaneous panniculitis-like T-cell lymphoma with unusual eschar-like crusting

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Abstract

Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) is a rare subtype of non-Hodgkin's lymphoma of the skin. Clinically, SPTCL presents as subcutaneous tumors located on the extremities or trunk, often associated with systemic symptoms like fever or fatigue. The therapeutic regimen for SPTCL is at present not standardized. We describe herein a case of a young woman who presented with intermittent fever and skin rash and was diagnosed later with SPTCL. The case is reported here for its rarity and rapidly changing unusual clinical manifestations. This case also highlights that monotherapy with systemic steroid can be a valuable treatment option for the management of SPTCL, especially in those without hemophagocytic syndrome.

Key words: subcutaneous panniculitis-like T-cell lymphoma, ulcerated, crusting

Introduction

Subcutaneous panniculitis-like T-cell lymphoma (SPTCL), a cytotoxic T-cell lymphoma, is characterized by infiltration of the subcutaneous tissue by pleomorphic T cells and benign macrophages that mimic lobular panniculitis [1, 2]. It was previously defined as a specific entity by the WHO but was regarded as a provisional entity by the European Organization for Research and Treatment of Cancer (EORTC). However, this entity has been redefined in the new WHO–EORTC classification (2005) [3] and this definition was subsequently adopted by the WHO classification in 2008 [4]. Cases with an α / β T-cell receptor (TCR) phenotype are usually CD8+, are confined to the subcutaneous tissue, and often have an indolent course.

On the other hand, cases with a γ / δ TCR phenotype are typically CD4– and CD8–, often express CD56, may involve the epidermis, and have a poor prognosis. Based on these facts, the group of SPTCL in the WHO–EORTC classification is now restricted only to cases with an α / β TCR phenotype. Cases with a γ / δ TCR phenotype are placed in the new category of cutaneous γ / δ T-cell lymphoma (CGD-TCL) [2, 3, 4].

SPTCL represents less than 1% of the cutaneous lymphomas [5]. We report here a case of a young woman with SPTCL who presented with intermittent fever and skin rash. The case is reported here for its rarity and rapidly changing unusual clinical manifestations.

Case synopsis

A 35-year-old woman presented with a history of a painful skin eruption associated with high grade intermittent fever that was unresponsive to antibiotics, for the preceding one and half months. In addition, there was a history of chills and generalized weakness. There was no significant family, past, or personal history. She had no history of weight loss.

On admission to the hospital, she had intermittent fever (temperature ranging from 102^o-103^o F). Her general physical examination was otherwise normal. Cutaneous examination showed multiple, tender, subcutaneous nodules and plaques of varying size (diameter ranging from 2–10 cm), distributed mainly on the right upper, and bilateral lower limbs (Figure 1, Figure 2). At presentation, there was ulceration and slight crusting on some of the indurated plaques. Gradually, within a span of a few days, extensive black-colored eschar-like crust developed over the lesions. No discharge was noted. There was no regional lymphadenopathy. The patient was offered paracetamol tablets for fever and pain.



Figure 1. (a) Ulcerated plaque on the right forearm (at presentation) (b) Escher-like crusting (on 10th day of hospitalization) (c) Residual hyperpigmentation (on 21st day of hospitalization)



Figure 2. (a) Erythematous ulcerated plaques on lower limb (at presentation) (b) Escher-like crusting (on 10th day of hospitalization) (c) residual hyperpigmentation (on 21st day of hospitalization)

Over the next few days, the overlying skin became almost normal leaving residual hyperpigmentation. However, the induration persisted.

Routine laboratory examination and biochemical panel initially showed leukopenia (total count: 2200/ml, neutrophil: 1408/ml), elevated ESR, and elevated lactate dehydrogenase level. Culture from the wound did not grow any organism. Antinuclear antibody (ANA) test was done by the hep-2 cell method, which was weekly positive (speckled pattern; 1:80 titer). Other markers of systemic lupus erythematosus including ds-DNA were negative. Histopathological examination (Figure 3) of incisional skin biopsy specimens from two different sites showed similar features. There was a small-to-medium-sized lymphoid infiltrate with typical panniculitis involving the lobular subcutaneous adipose tissue. Focal rimming of the adipocytes by lymphoid cells, focal areas of adipocytes necrosis, and multiple atypical lymphocytes were found. The tumor cells were admixed with a few histiocytes and demonstrated prominent karyorrhexis. The tumor cells expressed CD3, CD2, CD5, CD7, and CD8; it was immune-negative for CD20, CD4, CD25, CD30, ALK, and CD56 (Figure 4).

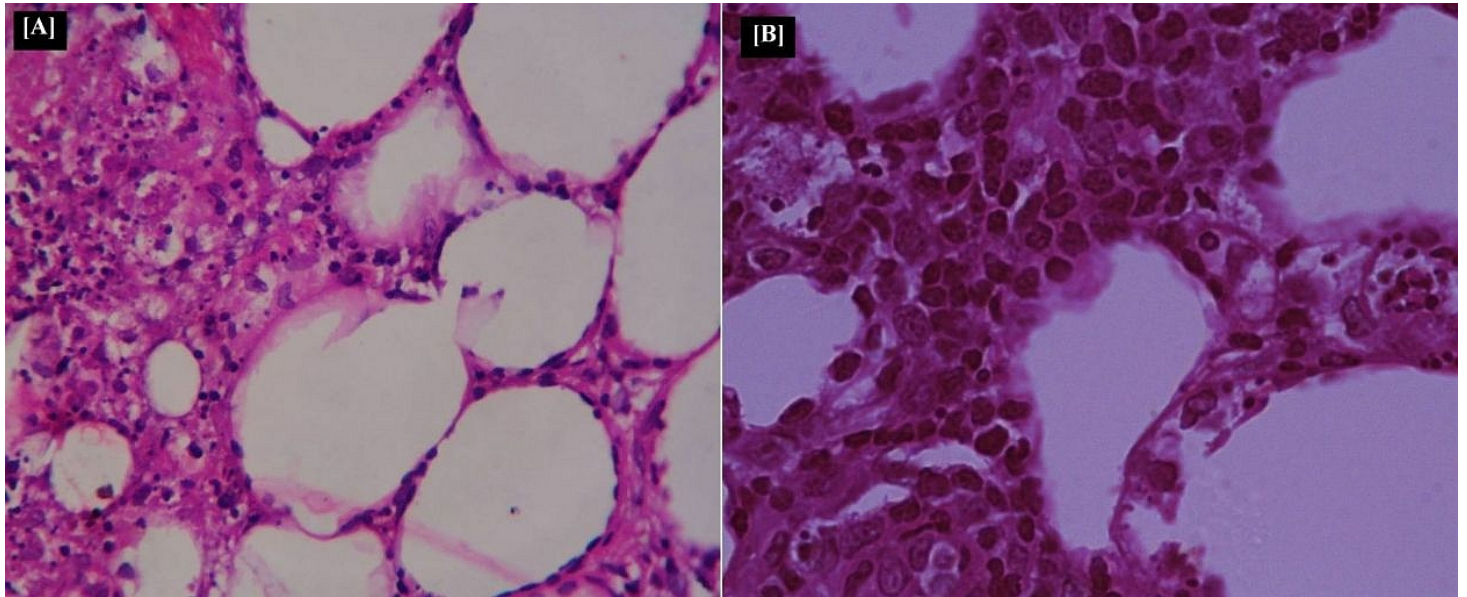


Figure 3. (A) Lymphocytic lobular panniculitis, rimming of the individual adipocytes by lymphocytes, and prominent karyorrhexis (H and E x 400) (B) Atypical lymphocytic infiltration around adipocytes (H and E x 1000)

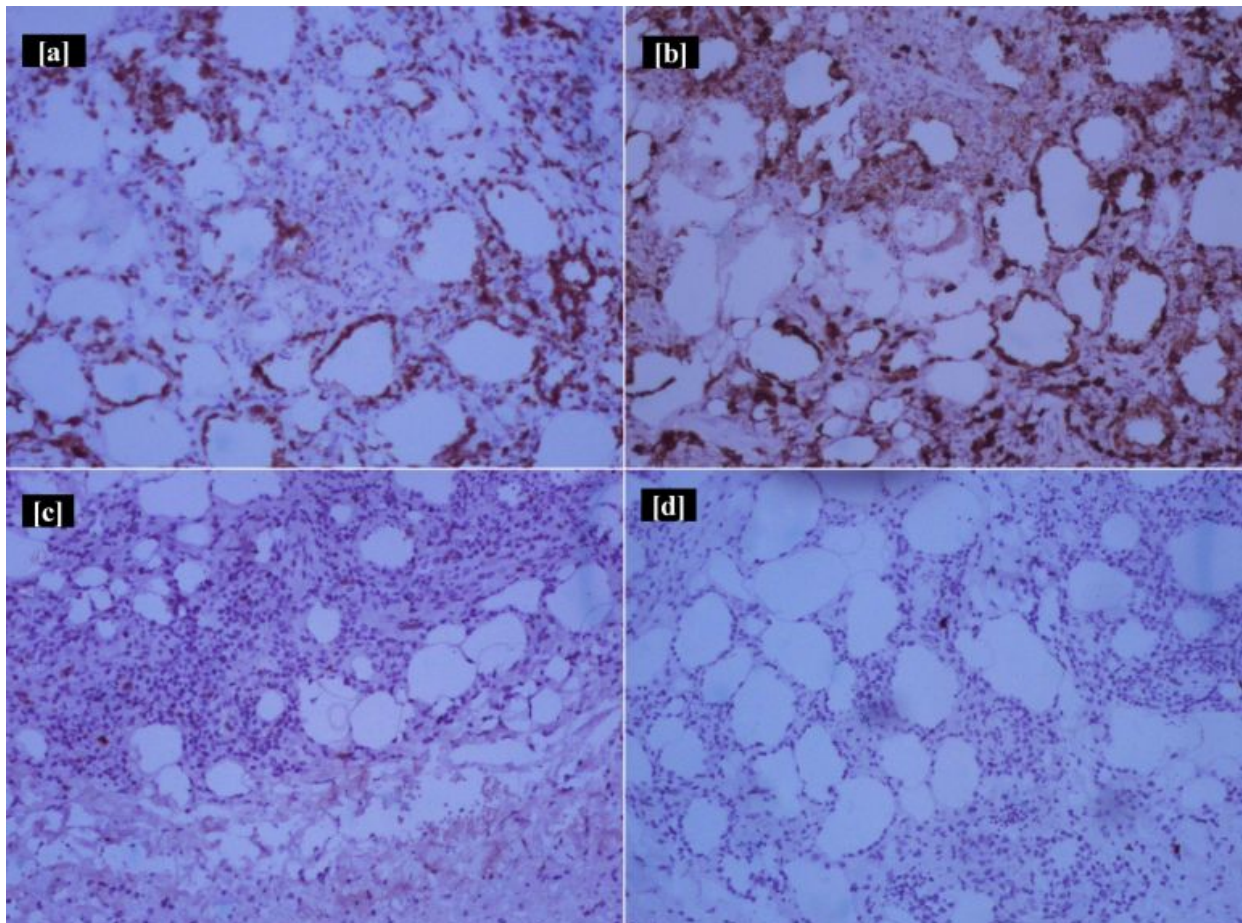


Figure 4. Immunohistochemistry (x100) showing (a) CD3 positivity (b) CD8 positivity (c) CD20 negativity and (d) CD56 negativity

A diagnosis of subcutaneous panniculitis-like T-cell lymphoma was made, based on the clinical, histopathological, and immunohistochemical findings. Bone marrow biopsy did not reveal involvement. Ultrasonography of the abdomen and chest X-ray was normal and our patient did not fulfill the criteria laid down for the diagnosis of hemophagocytic syndrome (HPS) [6]. We started a tapering course of oral prednisolone therapy (initial dose: 1mg / kg body weight). Within a couple of weeks the patient showed significant clinical improvement and the lesions were gradually improved leaving a mild degree of post-inflammatory hyperpigmentation. After 5 months of treatment, the patient is now on low dose prednisolone therapy (10 mg on alternate days) and shows no evidence of disease activity (Figure 5).



Figure 5. Residual hyperpigmentation of the cutaneous lesion with slight atrophy

Discussion

Although SPTCL usually runs an indolent course, the disease-related morbidity is often severe. In a previous series, the median age of SPTCL at diagnosis was 39 years (range, 0.5– 84.0 years) [7] and there were approximately equal numbers of male and female patients [1]. Clinically, SPTCLs usually present with well-circumscribed solitary or multiple nodules and plaques, which often involve the extremities. Ulceration is rare. In a large series, ulceration was found in 6% of the patients with SPTCL [7]. In the present case, a rapidly changing clinical scenario and unusual eschar-like crusting on the ulcerated plaques was a notable and unique feature. Systemic symptoms (B symptoms) like fever, fatigue, and weight loss were found in 59% of patients in a large series [7]. A hemophagocytic syndrome, which is generally associated with a rapidly progressive course, may occur in SPTCL. However, it is relatively more common in cutaneous CGD-TCL lymphomas with panniculitis-like lesions.

SPTCL may rarely be associated with autoimmune disorders, such as lupus erythematosus, juvenile rheumatoid arthritis, a combination of Sjogren disease and rheumatoid arthritis, type-1 diabetes mellitus, idiopathic thrombocytopenia, multiple sclerosis, Raynaud disease, and Kikuchi disease [7]. However, apart from a weakly positive ANA test (1:80 titer) we could not find any such association in the present patient.

SPTCL may be preceded for years or decades by an apparently benign panniculitis [3].

Histopathologically [2, 8, 9], it is characterized by a lymphocytic panniculitis (predominantly lobular) and rimming of the individual adipocytes by lymphocytes. The infiltrate comprises of small, medium, or large atypical lymphocytes with irregular hyperchromatic nuclei. Histiocytes are also present. Although, the rimming of adipocytes is a helpful diagnostic clue, it is not an exclusive feature of SPTCL. Similar rimming may also be found in other lymphomas and leukemias involving subcutaneous tissue [10]. Karyorrhectic nuclear fragments are consistently present in SPTCL.

“Bean-bag histiocytes” containing cell debris or red blood cells are occasionally found. Fat necrosis with foamy macrophages may also be seen. Sparse plasma cells are present in some cases. However, neutrophils and eosinophils are generally absent. Angiocentricity, angioinvasion, and large areas of necrosis are not characteristic of SPTCL [8, 9].

Immunohistochemical staining is the cornerstone of the diagnosis of SPTCL. The neoplastic cells, by definition, have a T-cell phenotype and are CD3+, CD4-, and CD8+. They also express cytotoxic markers TIA1 and granzyme B. CD30 and CD56 are not expressed. The tumors are EBV-negative [8, 9].

The differential diagnosis of SPTCL includes lupus erythematosus panniculitis (LEP), CGD-TCL, natural killer (NK)/T-cell lymphoma, and idiopathic lobular panniculitis, which often involves subcutaneous tissue. Histopathologically, the useful features that distinguish LEP from SPTCL are the presence (in lupus panniculitis) of epidermal involvement, lymphoid follicle formation with reactive germinal centers, a mixed cell infiltrate with prominent plasma cells, interstitial mucin deposition, and predominant CD4+ T cells [8,10]. CGD-TCL shows involvement of subcutis, epidermotropic infiltrates, and angiocentric growth. Immunophenotypically, CGD-TCL is CD4- CD8-, CD56+ and bF1+, unlike SPTCL [4, 11]. In NK/T-cell lymphoma, there is a diffuse or angiocentric and periappendageal cellular infiltrate that involves the dermis and subcutaneous tissue. The tumor lymphocytes are CD56+, CD2+, and surface CD3- and the lesions usually show positivity for Epstein-Barr virus [8, 11]. Idiopathic lobular panniculitis is usually a diagnosis of exclusion.

Subcutaneous panniculitis-like T-cell lymphoma has a favorable prognosis. The overall 5-year survival rate exceeds 80% [2, 11]. In a recently published Australian study, the five year survival rate of SPTCL was 100% [12]. On the other hand, CGD-TCL is associated with a poor prognosis (a 5-year survival rate of 11%) regardless of its association with HPS [10].

The therapeutic regimen for SPTCL is currently not standardized [2, 10, 13]. Prior to the WHO-EORTC re-classification, doxorubicin-based chemotherapy, radiotherapy, and bone marrow transplantation were usually used treatment modalities [10]. Because SPTCL has an indolent course, the use of aggressive therapeutic regimens (such as combination chemotherapy or bone marrow transplantation) have recently come into question [10]. Recent studies suggest that many patients can be controlled for long periods of time with systemic corticosteroids [10, 14]. The favorable prognosis and 5-year survival rate of SPTCL further justify an initial treatment approach with systemic steroids alone [2, 13], particularly if HPS is not present [10].

Subcutaneous panniculitis-like T-cell lymphoma often poses a real diagnostic challenge. In this report, we seek to emphasize that whenever a patient presents with a long history of panniculitis-like skin lesions that are associated with fever, weight loss, and other constitutional symptoms, SPTCL should be considered. Neutropenia, which does not respond to antibiotics, is another clue to the diagnosis [11]. Furthermore, only a high degree of suspicion can hasten the diagnosis and hence reduce the morbidity and mortality of this condition. This case also highlights that monotherapy with systemic steroids might be a valuable treatment option for the management of SPTL, especially in those without HPS.

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