

Local Facial Edema: a Novel Presentation of Subcutaneous Panniculitis-like T-Cell Lymphoma in a 30-Year-Old Iranian Woman

Farid Kosari and Sanam Akbarzadeh Hosseini

Department of Pathology, Shariati Hospital, Tehran University of Medical Sciences, Tehran, Iran

Received: 2 Jun. 2013; Accepted: 17 May 2014

Abstract- Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) is an infrequent cytotoxic T-cell lymphoma of the skin with a unique immunophenotypic profile. Here we describe this lymphoma in a 30-year-old Iranian woman. She presented with periorbital edema, mandibular swelling with edematous areas on her face and two small lymph nodes (6-7mm) in the cervical area. Histopathological findings revealed lobular infiltration of atypical lymphoid cells in subcutaneous tissue, especially around individual fat cells. Immunohistochemical evaluation of the atypical lymphocytes showed CD3 and CD8 immunoreactivity and CD4 and CD56 negativity. She was diagnosed as SPTCL based on clinical, histopathological and immunohistochemical findings. This report suggests that SPTCL should be considered in the differential diagnosis of any atypical skin lesion involving subcutaneous fat. Suitable deep skin biopsy and immunohistochemical staining are essential to differentiate between SPTCL and other more common diagnoses.

© 2014 Tehran University of Medical Sciences. All rights reserved.

ActaMedicaIranica, 2014;52(12):950-953.

Keywords: Cutaneous Lymphoma; T- Cell; Panniculitis

Introduction

Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) is an uncommon type of peripheral T-cell lymphoma with a unique immunophenotypic profile and indicates less than 1% of all the non-Hodgkin lymphomas (1-2). It overallly occurs in men and is only seldom seen in females by a ratio of approximately 2:1(3). In the literature review, less than 50 cases are reported with SPTCL (4-5). It is a diagnostic challenge for the histologic and clinical features that can mimic benign causes of panniculitis. The association of hemophagocytic syndrome (HPS) indicates a poor prognosis (6). So far, the treatment of SPTCL is not standardized, and the response to treatment is different, with a high relapse rate (1,4,6,7). Here in, we describe a 30 aged Iranian woman who presented with local facial swelling and/or subcutaneous nodules and fever, and were subsequently given diagnoses of SPTCL. Here we suggest that SPTCL may present in the atypical clinical presentation and should be considered in the various diagnoses of any atypical skin lesion involving subcutaneous fat. The patient responded to treatment with prednisolone and CHOP chemotherapy.

Case Report

A 30-year-old Iranian woman presented with a history of low-grade fever, chills, general fatigue, anorexia, swelling and induration of face from one month ago. A systemic review was unremarkable except for marked mandibular swelling, induration and periorbital edema. Her medical and family histories were unremarkable. On examination, the patient was pale and febrile (temperature of 38.5°C). She had two small lymph nodes (6-7mm) in the cervical area. Local skin examination revealed bilateral erythematous ill-defined skin and subcutaneous induration and swelling. No other skin lesion is identified. No Mucosal lesions were seen. Laboratory findings revealed anemia (Hb: 7.9 gr/dl), elevated ESR (51mm/h, Normal range: female up to 22) and C-reactive protein test (+4). Renal function tests, lipid profile, hepatic function tests, serologic and other hematologic tests were unremarkable.

Abdominal and pelvic ultrasonography showed mild hepatosplenomegaly and dilatation of intra-hepatic veins. One para aortic echogenic lesion detected suggestive of para aortic adenopathy. Bone marrow aspirates and trephine biopsy showed normocellular marrow with some lymphoid aggregates, free of the

Corresponding Author: F. Kosari

Department of Pathology, Shariati Hospital, Tehran University of Medical Sciences, Tehran, Iran
Tel: +98 21 84901000, Fax: +98 21 88633078, E-mail address: faridkosari@yahoo.com

tumoral involvement. A deep punch biopsy of skin from the right side lower jaw revealed essentially intact epidermis and dermis. Subcutaneous fat showed lobular lymphoid infiltrate and septal panniculitis (10 × magnifications) (Figure 1:A). On high-power view (Figures 1:B and C), rimming of adipocytes by neoplastic lymphoid cells with enlarged, irregular hyperchromatic nuclei and scanty cytoplasm noted. Scattered histiocytes, some with phagocytic cell debris

were also seen. No erythrophagocytosis or granuloma was observed. Immunohistochemical staining of mentioned lymphoid cells showed positivity for CD3, CD8, granzyme B and negativity for CD4 and CD56 (Figures 1: D, E, and F). Prednisolone 40 mg once daily prescribed for the patient then tapered gradually and followed by CHOP chemotherapy. The fever and other systemic symptoms and skin lesions resolved within two weeks after the initiation of the treatment.

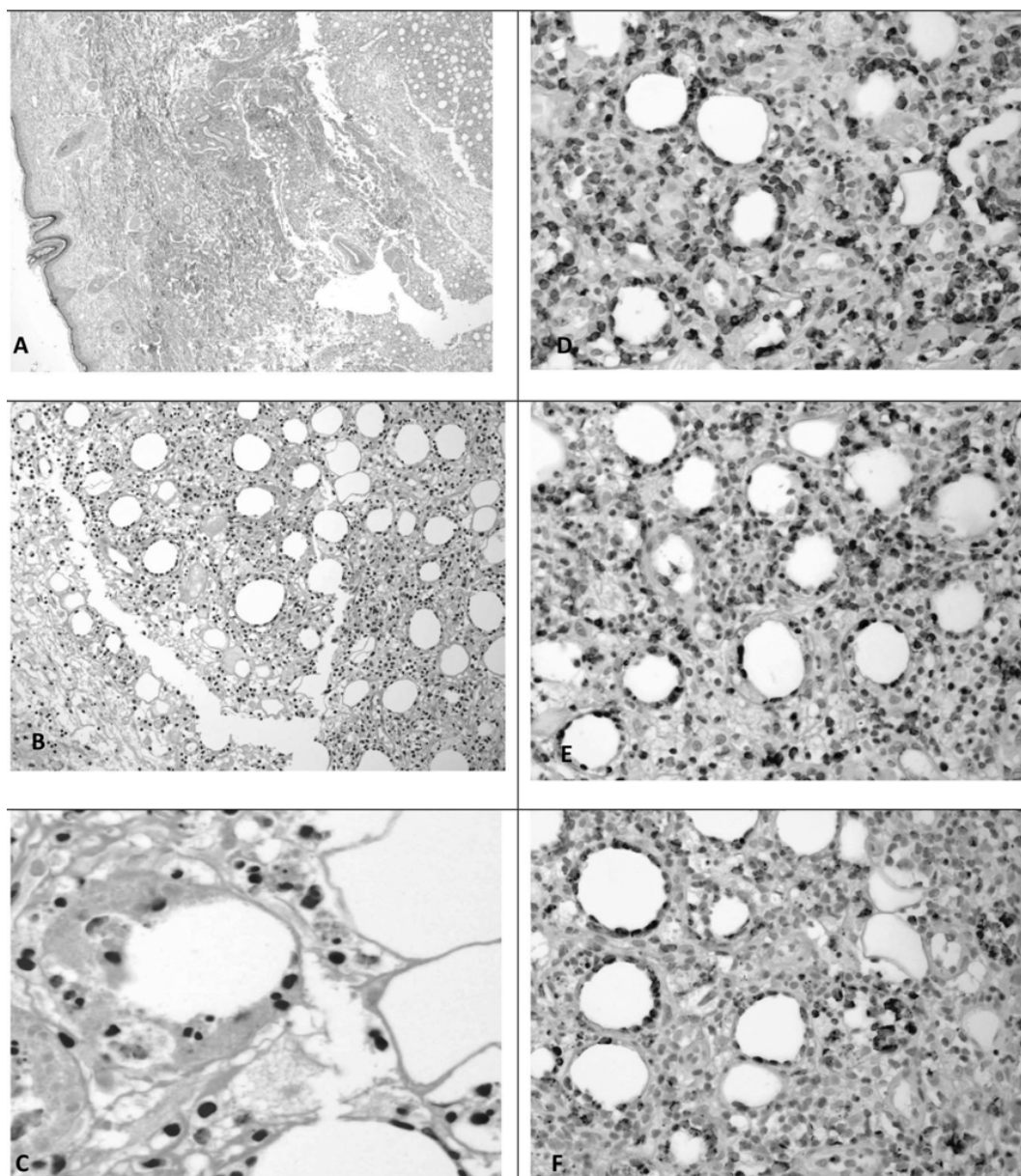


Figure 1. Subcutaneous panniculitis-like T-cell lymphoma

(A) Skin biopsy revealed intact epidermis and dermis with lobular lymphoid infiltrate in subcutaneous fat and septal panniculitis. (B) Rimming of adipocytes by neoplastic lymphoid cells are noted. (C) Scattered histiocytes, some with phagocytic cell debris are seen. (D) The neoplastic cells are CD3+. (E) The rimming of fat spaces by tumor cells is highlighted by staining for CD8. (F) The neoplastic cells express Granzyme B

Discussion

Gonzalez *et al.*, originally described SPTCL (1), as an unusual form of cutaneous lymphoma that was localized within the subcutis and mimicked lobular panniculitis. It can occur with or without visceral disease. The most frequent clinical differential diagnosis includes panniculitis, particularly erythema nodosum, infectious panniculitis, and cellulitis. Patients typically present with subcutaneous nodules on the trunk or lower extremities and less generally on the upper extremities or face. In our patient, the tremendous swelling of the face led to the misdiagnosis of contact dermatitis. The disease can have a slow course or can be rapidly progressive and fatal. The presence of HPS, which is characterized by fever, hepatosplenomegaly, pancytopenia, and coagulopathy, is a poor prognostic sign (6). HPS is thought to product from lymphokines secreted from neoplastic T cells.

Histologically, SPTCL is characterized by an atypical lymphocytic infiltrate primarily in a lobular distribution in the subcutaneous tissue with common cell necrosis and no epidermotropism. The cells in the reported cases of SPTCL often show features of cytotoxic T cells (8).

In current patient, no typical skin lesion is seen. The patient only has mandibular swelling with ill - defined erythematous induration and periorbital edema.

Histopathology of SPTCL revealed essentially intact epidermis and dermis. Subcutaneous fat presents lobular atypical lymphoid infiltrate, rimming of individual fat cells, and septal panniculitis (9,10). There are small to medium, or sometimes large sized pleomorphic T cells with enlarged irregular hyperchromatic nuclei and scanty cytoplasm admixed with benign histiocytes, plasma cells and neutrophils (9,11). Necrosis, Scattered mitosis, karyorrhexis, cytophagocytosis, granulomatous reaction, and angioinvasion can be present (8,9). The neoplastic cells in SPTCL have cytotoxic T cell features which are CD3+ and CD4-. α/β + TCL is CD8+ and usually CD30- and CD56-. Γ/δ -TCL is the main differential diagnosis and usually CD8-, CD56+, and CD30+ (12,13). Granzyme B, a cytotoxic granular protein, is positive in the tumor cells (2,8). No specific genetic abnormalities are identified.

In present patient, the skin biopsy showed histopathological and immunophenotypical characteristics of α/β + TCL; including atypical lymphoid cells were positive for CD3 and CD8 and negative for CD4 and CD56 accompanied by scattered B

cells were CD20+. These findings exclude most differential diagnoses.

In conclusion from our recent experience, in whom the diagnosis of SPTCL was delayed because of confusion with contact dermatitis, cellulitis, or panniculitis. The scarcity of this entity combined with the resemblance of the clinical presentation of SPTCL to more benign inflammatory disorders, particularly panniculitis, can result to misdiagnosis. Therefore, SPTCL should be considered in the differential diagnosis of any atypical skin lesion involving subcutaneous fat tissue. Suitable deep skin biopsy and immunohistochemical staining are essential to differentiate between SPTCL and other more common diagnoses.

References

- Gonzalez CL, Medeiros LJ, Brazier RM, et al. T-cell lymphoma involving subcutaneous tissue: a clinicopathologic entity commonly associated with hemophagocytic syndrome. *Am J Surg Pathol* 1991;15(1):17-27.
- Kumar S, Krenacs L, Medeiros J, et al. Subcutaneous panniculitic T-cell lymphoma is a tumor of cytotoxic T lymphocytes. *Hum Pathol* 1998;29(4):397-403.
- Alsaleh QA, Nanda A, Al-Ajmi H, et al. Clinicoepidemiological features of mycosis fungoides in Kuwait, 1991-2006. *Int J Dermatol* 2010;49(12):1393-8.
- Craig AJ, Cualing H, Thomas G, et al. Cytophagic histiocytic panniculitis—a syndrome associated with benign and malignant panniculitis: case comparison and review of the literature. *J Am Acad Dermatol* 1998;39(5 Pt 1):721-36.
- Au WY, Choy C, Kwong WL. Aggressive subcutaneous panniculitic-like T-cell lymphoma: complete remission with fludarabine, mitoxantrone, and dexamethasone. *Br J Dermatol* 2000;143(2):408-10.
- Von den Driesch P, Staib G, Simon M, et al. Subcutaneous T-cell lymphoma. *J Am Acad Dermatol* 1997;36(2 Pt 2):285-9.
- Wang CYE, Su WPD, Kurtin PJ. Subcutaneous T-cell lymphoma. *Int J Dermatol* 1996;35(1):1-8.
- Salhany KE, Macon WR, Choi JK, et al. Subcutaneous panniculitic-like T-cell lymphoma: clinicopathologic, immunophenotypic, and genotypic analysis of alpha/beta and gamma/ delta subtypes. *Am J Surg Pathol* 1998;22(7):881-93.
- Bagheri F, Cervellione KL, Delgado B, et al. An Illustrative Case of Subcutaneous Panniculitis-Like T-Cell Lymphoma. *J Skin Cancer* 2011;2011:824528.

10. Lozzi GP, Massone C, Citarella L, et al. Rimming of adipocytes by neoplastic lymphocytes: a histopathologic feature not restricted to subcutaneous T-cell lymphoma. *Am J Dermatopathol* 2006;28(1):9-12.
11. Vijaya B, Sunila MD, Manjunath GV. Subcutaneous panniculitic-like T-cell lymphoma: a red alert! The role of a vigilant histopathologist. *Indian J Pathol Microbiol.* 2011;54(2):376-8
12. 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):838-45.
13. Willemze R, Jaffe ES, Burg G, et al. WHO-EORTC classification for cutaneous lymphomas. *Blood* 2005;105(10):3768-85.