CASE REPORT

Subcutaneous panniculitis-like T-cell lymphoma: A rare presentation

Dr. Meher Lakshmi Konatam[1,*], Dr. Triveni B[1], Dr. Sai Mallikarjun S[1] and Dr. Md Shoiab Zeesham[1]

[1] Assistant Professor, MNJ Institute of Oncology and & Regional Cancer Centre, Red Hills, Lakadikapul, Hyderabad-500004, Telangana, India.

Abstract

Subcutaneous panniculitis-like T-Cell Lymphoma (SPTL) is a very rare form of skin lymphoma that is localized primarily to the subcutaneous adipose tissue without palpable involvement of the lymph nodes. Diagnosis of SPTCL is a challenge as clinical features are non specific and mimic benign conditions. Unless there is expertise in oncopathology and strong suspicion, biopsy is also difficult to interpret. Due to misdiagnosis, steroids are started early which mask the disease leading to further delay in seeking appropriate care. We present here an illustrative case of a young lady with symptoms mimicking collagen vascular disease and diagnosis of SPTCL reached later.

Keywords: subcutaneous panniculitis like T cell lymphoma; SPTL; lymphoma

***Corresponding author:** Dr. Meher Lakshmi Konatam, Assistant Professor, MNJ Institute of Oncology and & Regional Cancer Centre, Red Hills, Lakadikapul, Hyderabad-500004, Telangana, India. Tel.: 9440592569; Email: mehercancercare@gmail.com

Received 21 July 2015; *Revised* 18 August 2015; *Accepted* 29 August 2015;.Published.5.September.2015

Citation: Meher Lakshmi K, Triveni B, Sai Mallikarjun S, Shoiab Zeesham Md. Subcutaneous panniculitis-like T-cell lymphoma: A rare presentation. J Med Sci Res. 2015; 3(4):183-186.
DOI: http://dx.doi.org/10.17727/JMSR.2015/3-035

Introduction

Subcutaneous panniculitis-like T-cell lymphoma (SPTL) was first described in 1991 by Gonzalez et al. [1] but was not recognized as a distinct entity by the World Health Organization until 2001 [2]. It is estimated that SPTCL accounts for less than 1% of all non-Hodgkins lymphomas [2]. Most often it presents as multiple, painless, subcutaneous nodules on the extremities and trunk. In its early phase, the nodules may resolve without treatment and subsequently new nodules may develop on the same or different skin locations. SPTL mimics benign conditions like panniculitis, eczema, dermatitis, psoriasis, and other skin and soft tissue infections leading to delay in diagnosis. Clinical and systemic symptoms are nonspecific and can include fever, chills, and weight loss; approximately half of the patients develop mild cytopenias. More serious conditions associated with SPTL include hepatosplenomegaly, mucosal ulcers, serosal effusions, hemophagocytosis syndrome (HPS), and pancytopenia, though these are less common [3, 4].

A case of young lady who became symptomatic during last trimester of pregnancy is reported here. After multiple misdiagnoses and false interpretation of skin biopsy, diagnosis of SPTCL was made and appropriate treatment was offered.

Case presentation

A 26-year-old lady with 8 months of amenorrhea presented with history of intermittent high grade fever with chills and erythematous patches over both upper limbs and lower limbs with few lesions ulcerated and few others healed with hyperpigmentation since January 2015. There was no history of oral ulcers, alopecia, Reynaud's, photosensitivity, malar rash, sicca symptoms, and skin tightening. No history of loss of appetite, weight loss or bleeding manifestations. She had history of pulmonary arterial hypertension (PAH) in first pregnancy. No history of abortions. She was admitted in another hospital in March 2015 with these symptoms. Possibilities of systemic lupus erythematosus (SLE)/ antiphospholipid syndrome (APS) were considered. But her antinuclear antibodies (ANA) and dsDNA antibodies were negative. Blood and urine cultures were sterile, procalcitonin levels and bone marrow aspiration was normal. She developed transaminitis and hypertension with cytopenias in due course. She was kept under obstetrician care with the suspicion of HELLP syndrome. She was managed conservatively and discharged on steroids, 1mg/ kg. She delivered a baby uneventfully in April 2015.

After delivery, she developed nodular swellings over upper limbs, chest and abdomen. There was no history of pain in the swellings and skin over swellings was normal and freely mobile. She continued to have fever spikes. Biopsy of skin lesion done at local hospital was s/o lobular panniculitis without vasculitis. Repeat ANA, dsDNA, C3C4 were normal. Bone marrow aspiration and biopsy were normal.

She presented to our hospital in May 2015 with persistent subcutaneous nodules. On examination, patient was conscious, coherent with performance status 1. Subcutaneous nodules were present over left arm, breast and upper abdomen, healed necrotic patches over both upper and lower limbs, few ulcerated lesions on upper and lower limbs (Figure 1). Pulse rate 102 per minute, other vital data normal. Systemic examination was unremarkable.

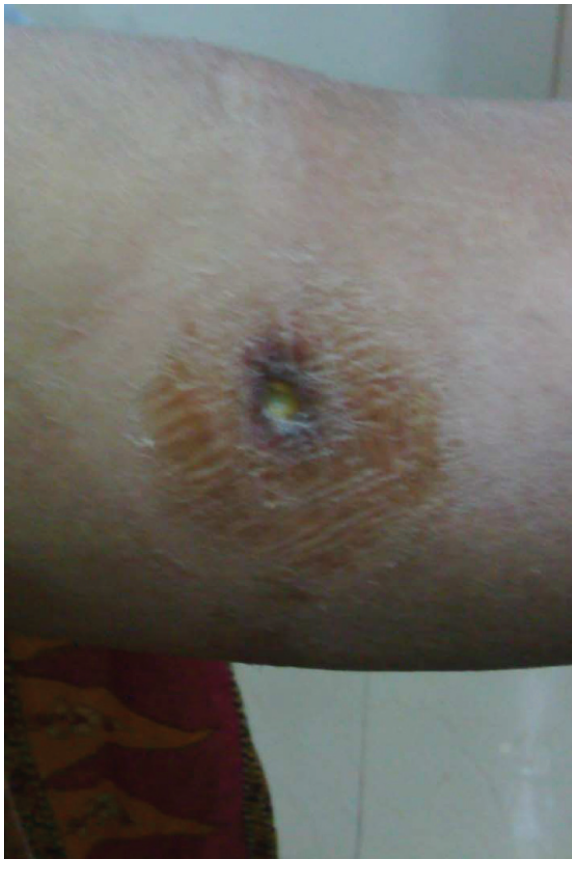

Figure 1: Ulcerated lesion on medial aspect of left arm.

Review of skin biopsy slides was done at our hospital. Microscopically epidermis is unremarkable. Subepithelial and underlying dermis and subcutaneous tissue shows multiple foci of atypical lymphoid cells in aggregates and scattered discretely. Cytoplasm is scant with large dark nucleus. These cells are also seen around adnexa and perineurally. Intervening stroma shows thick collagen bundles. Immunohistochemistry (IHC) was done and CD3 positive, CD20 negative, CD68 positive in few histiocytes, cytokeratin negative, Ki 67 around 40%. Conclusion of SPTL was arrived at (Figures 2 and 3) contrast enhanced computed tomography (CECT) chest and abdomen showed extensive subcutaneous fat stranding in chest and abdomen parietal wall (Figure 4). Blood counts, liver and kidney function tests were normal.

She was started on CHOP chemotherapy. After therapy her skin nodules regressed completely with complete resolution of her symptoms. She is now due for 2nd cycle chemotherapy and is doing well.

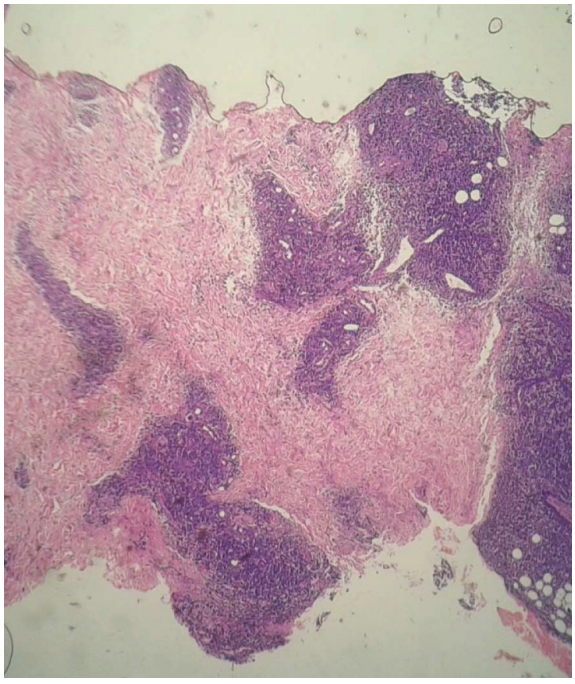

Figure 2: Low power view showing unremarkable epidermis, multiple foci of atypical lymphoid cells in aggregates and scattered discretely in subepithelial and underlying dermis and subcutaneous tissue. These cells are also seen around adnexa and perineurally. Intervening stroma shows thick collagen bundles.

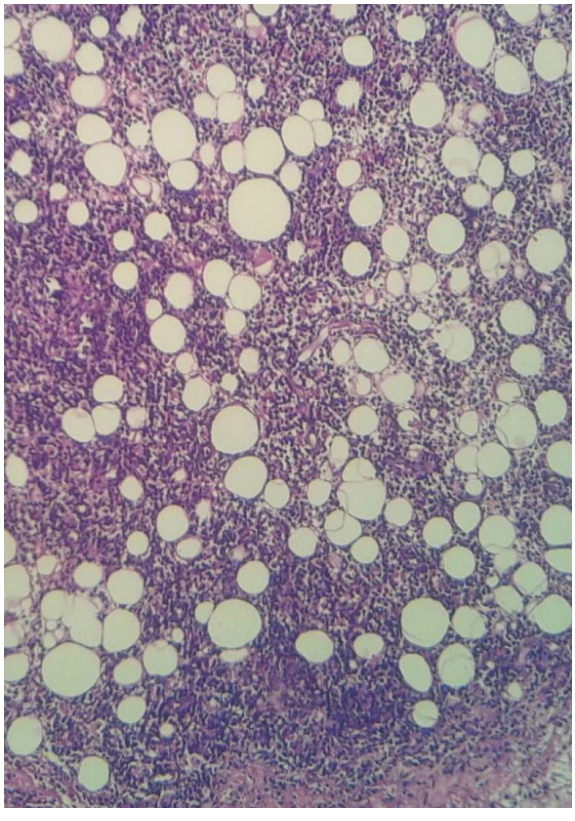

Figure 3: Lymphoid cells seen surrounding fat cells.

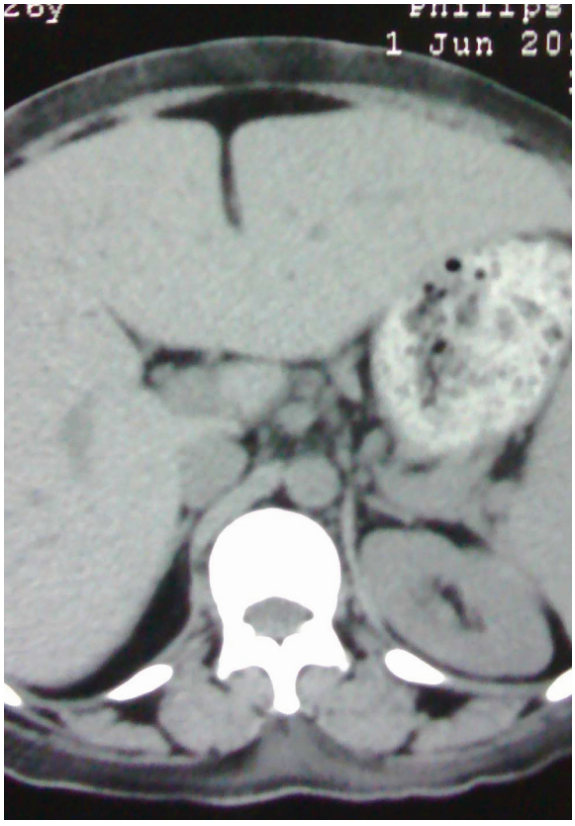

Figure 4: CT abdomen showed extensive subcutaneous fat stranding in parietal wall.

Discussion

In the WHO classification, subcutaneous panniculitis-like T-cell lymphoma (SPTL) is defined as a distinct type of T-cell lymphoma with an aggressive clinical behavior. Recent studies suggest that distinction should be made between SPTL with an alpha/ beta T-cell phenotype (SPTL-AB) and SPTL with gamma/ delta T-cell phenotype (SPTL-GD). SPTL-ABs were generally confined to the subcutis, had a CD4-, CD8+, CD56-, betaF1+ phenotype, were uncommonly associated with a hemophagocytic syndrome (HPS; 17%), and had a favorable prognosis (5-year overall survival [OS]; 82%). SPTL-AB patients without HPS had a significantly better survival than patients with HPS (5-year OS; 91% vs 46%; P < .001). SPTL-GDs often showed (epi) dermal involvement and/ or ulceration, a CD4-, CD8-, CD56+/-, betaF1- T-cell phenotype, and poor prognosis (5-year OS; 11%), irrespective of the presence of HPS or type of treatment. These results indicate that SPTL-AB and SPTL-GD are distinct entities, and justify that the term SPTL should further be used only for SPTL-A [4].

Apart from SPTL-GD and other aggressive T-cell lymphomas, SPTL-AB should be differentiated from lupus erythematosus panniculitis (LEP). The relationship between SPTL and LEP, which may be clinically indistinguishable, is controversial [5, 6]. In a detailed report on 11 cases of LEP, Massone, et al. proposed histopathological criteria useful in differentiating between LEP and SPTL-AB, suggesting that these represent distinct entities [6]. Useful histopathologic criteria favoring a diagnosis of LEP included epidermal involvement, mucin depositions, the presence of reactive germinal centers, clusters of B cells or considerable numbers of admixed plasma cells, and polyclonal TCR (T-cell receptor) gene rearrangement. In contrast, Magro et al. emphasized overlapping features between LEP and SPTL and suggested that both conditions form a spectrum of disease [5].

Initial treatment in SPTL-AB varied widely from only radiotherapy or prednisone to doxorubicin-based chemotherapy. Since the WHO classification from 2001 did not yet distinguish between SPTL-AB and SPTL-GD and describes SPTL as an aggressive type of lymphoma, in most centers doxorubicin based chemotherapy is the preferred type of treatment, sometimes in combination with alemtuzumab or followed by an auto-SCT [7-9]. High-dose chemotherapy followed by auto-SCT or allo-SCT has been suggested as an important option in patients with refractory or recurrent SPTL [9].

Our patient was initially started on steroids in outside hospital with initial partial response. By the time she presented to our hospital, she developed new lesions. She responded well to CHOP chemotherapy with complete resolution of symptoms and lesions.

Conclusions

High index of suspicion is required to diagnose subcutaneous panniculitis-like T-Cell Lymphoma (SPTL) to avoid delay in early diagnosis and for timely and accurate treatment.

Conflict of interest

The authors declare no conflict of interest.

References

1. Gonzalez CL, Medeiros LJ, Braziel RM, Jaffe ES. T-cell lymphoma involving subcutaneous tissue. A clinicopathologic entity commonly associated with hemophagocytic syndrome. Am J Surg Pathol. 1991; 15(1):17–27.
2. Jaffe ES, Harris NL, Stein H, Vardiman JW. Pathology and Genetics of Tumours of Haematopoietic and Lymphoid Tissues. World Health Organization Classification of Tumours, IARC Press, Lyon, France, 2001.
3. Willemze R, Jaffe ES, Burg G, Cerroni L, Berti E, et al. WHO-EORTC classification for cutaneous lymphomas. Blood. 2005; 105(10):3768–3785.
4. Willemze R, Jansen PM, Cerroni L, Berti E, Santucci M, 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.
5. Magro CM, Crowson AN, Kovatich AJ, Burns F. Lupus profundus, indeterminate lymphocytic lobular panniculitis and subcutaneous T-cell lymphoma: a spectrum of subcuticular T-cell lymphoid dyscrasia. J Cutan Pathol. 2001; 28(5):235–247.
6. Massone C, Kodama K, Salmhofer W, Abe R, Shimizu H, et al. Lupus erythematosus panniculitis (lupus profundus): clinical, histopathological, and molecular analysis of nine cases. J Cutan Pathol. 2005; 32(6):396–404.
7. Go RS, Wester SM. Immunophenotypic and molecular features, clinical outcomes, treatments, and prognostic factors associated with subcutaneous panniculitis-like T-cell lymphoma: a systematic analysis of 156 patients reported in the literature. Cancer. 2004; 101(6):1404–1413.
8. Ghobrial IM, Weenig RH, Pittelkow MR, Qu G, Kurtin PJ, et al. Clinical outcome of patients with subcutaneous panniculitis-like T-cell lymphoma. Leuk Lymphoma. 2005; 46(5):703–708.
9. Alaibac M, Berti E, Pigozzi B, Chiarion V, Aversa S, et al. High-dose chemotherapy with autologous blood stem cell transplantation for aggressive subcutaneous panniculitis-like T-cell lymphoma. J Am Acad Dermatol. 2005; 52(5 Suppl 1):S121–S123.