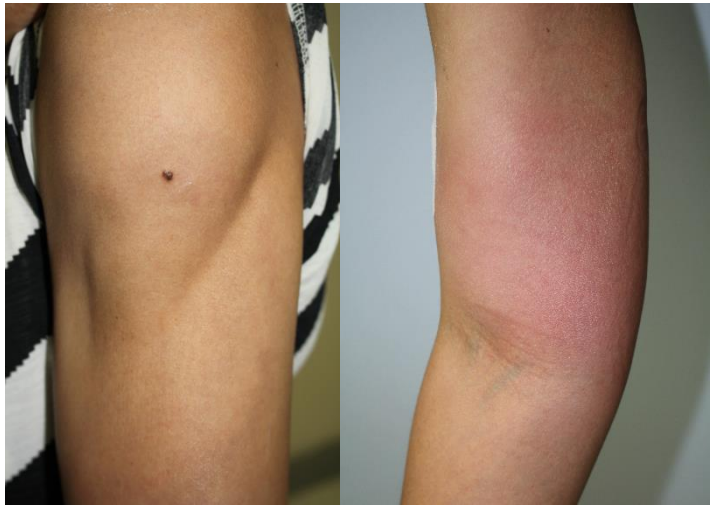
Case 22
A 34-year-old woman from Nakhon Sawan
Chief complaint: Multiple tender mass on trunk and extremities for 1 month

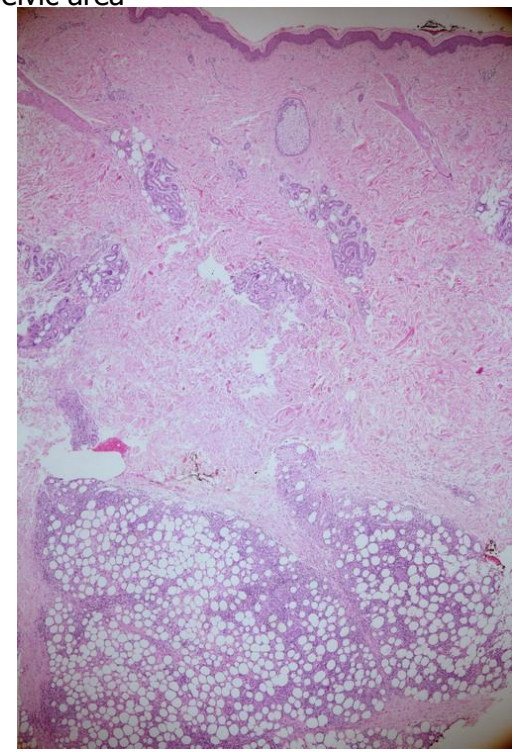


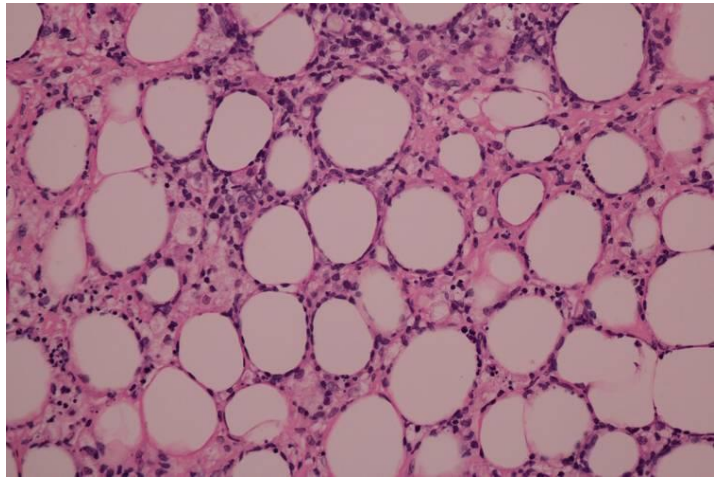
Present illness: Four months previously, the patient experienced some dimpled skin around left triceps and right pelvic areas. She was diagnosed as localized lipodystrophy, unfortunately she lost to follow up while not yet fully investigated. Two months previously, the patient developed intermittent low grade fever. She denied any associated symptoms such as weight loss or anorexia. Four weeks later, she developed persistent high grade fever with multiple painful mass on chest, abdomen, both arms and genitalia. She denied a history of photosensitivity, joint pain and current medical used.

Past history: No known underlying disease

Physical examination:
V/S: T 38.3°C, PR 90 bpm, RR 20 times/min, BP 96/64 mmHg
GA: A Thai woman, good consciousness, mild pallor, no jaundice
HEENT: No pale conjunctiva, anicteric sclera
CVS: Normal S_1S_2, no murmur
RS: Normal breath sound, no adventitious sound
Abdomen: Soft, non-tender, liver and spleen not palpable
Lymphatic system: Multiple axillary and groin nodes enlargement
Nervous system: Grossly intact

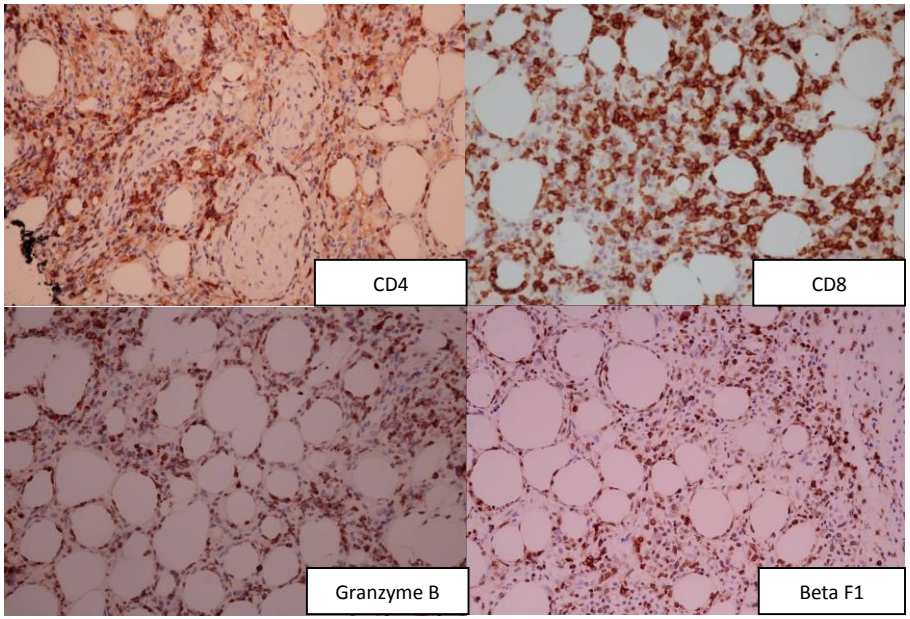
Skin examination:
- Multiple erythematous to brownish indurated plaques distributed on chest, abdomen, upper back and both arms
- Few skin-colored atrophic plaques distributed on left arm and right pelvic area





Histopathology: (S15-015737, left arm)

- Diffuse inflammatory cell infiltrate of lymphocytes, admixed with some plasma cells lymphoid cells in the fat lobules of subcutaneous tissue and lower dermis.
- Some lymphocytes show mild to moderate nuclear atypia



Immunohistochemistry (S15-017629):

- Most of lymphocytes positive for CD3, CD4, CD8, Granzyme B, Beta F1
- Negative for CD20, CD30, CD56

Laboratory investigation:

CBC: Hct 29.8%, Hb 10.1 g/dL, WBC 4120/mm^3 (PMN 75% L 22% M 3%), Plt 233000/mm^3
BUN 7 mg/dL, Cr 0.65 mg/dL
LFT: ALP 43 U/L, GGT 17 U/L, AST 51 U/L, ALT 18 U/L, TB 0.5 mg/dL, DB 0.2 U/L, alb 22.2 g/L
LDH 698 U/L
UA: Normal
Chest X-ray: No active pulmonary disease
CT chest and abdomen: Lymphadenopathy at bilateral axillary and inguinal regions. No intraabdominal or mediastinal lymphadenopathy. No hepatosplenomegaly.
Bone marrow biopsy: Hypercellular bone marrow
Lymph node biopsy (Left axillar): Reactive lymphoid hyperplasia

Diagnosis: Subcutaneous panniculitis-like T-cell lymphoma

Treatment: CHOP regimen

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Consultant: Ploysyne Rattanakaemakorn, MD

Discussion:

Initial presenting symptoms with multiple sites of panniculitis with areas of lipoatrophy gave high suspicion for lymphoproliferative diseases or lupus paniculitis. Given that our patient also had a history of intermittent fever along with multiple lymphadenopathy, lymphoproliferative disease was the provisional diagnosis. The incisional skin biopsy performed on indurated plaques of left arm showed diffuse inflammatory cell infiltrate of lymphocytes, plasma cells and some atypical lymphoid cells within subcutaneous fat. The immunophenotypic features show positive CD3, CD4, CD8, beta F1 and negative CD20, CD30, CD56 and EBER. Therefore, the finally diagnosis in our patient is subcutaneous panniculitis-like T-cell lymphoma (SPTL).

SPTL is a rare cytotoxic T-cell lymphoma primarily localized to the subcutaneous tissue that can be misdiagnosed as a benign panniculitis. It accounts for less than 1% of non-Hodgkin's lymphomas.[1] SPTL was redefined by World Health Organization-European Organization for Research and Treatment of Cancer (WHO-EORTC) classification of primary cutaneous lymphomas in 2008 as a distinct entity of TCR α/β phenotype which had an indolent clinical course in contrast to TCR γ/δ phenotype.[2] It typically presents in young adults with has no clear gender predisposition. Clinical presentation usually consists of painless indurated subcutaneous nodules or plaques with variable number and size.[1,3] Ulceration has also been reported in some cases.[4] The sites of skin involvement are commonly on extremity, trunk, face and neck, respectively. Common skin findings include various stages of healing and areas of lipoatrophy which result from the spontaneous regression of subcutaneous nodules without treatment.[1,2] Up to 60% of patients presented with prodromal symptoms such as fevers, chills and night sweat.[5] Lymphadenopathy and hepatosplenomegaly are also uncommon, it accounts for 8%.[1,2] Laboratory abnormalities, most commonly include anemia, leukopenia, thrombocytopenia or combined cytopenias, and elevated liver function tests.[1] The diagnosis of SPTL is made merely by histology and immunohistochemistry. The histopathologic findings consist of lobular panniculitis with dense infiltration of atypical lymphocytes. The typical finding is the rimming of individual adipocytes by atypical lymphocytes.[1,3,4] Macrophages filled with nuclear debris in the cytoplasm giving a feature of bean-bag appearance is a common finding. The immunophenotype is typically positive for βF1, CD3, CD8, and cytotoxic proteins and negative CD4, CD5, CD56, and EBER.[2,4]

The prognosis of SPTL is favorable with a 5-year survival rate of more than 80%. The risks for developing hemophagocytic syndrome are approximately 17% which is associated with poor outcomes.[2,6] Metastatic disease and visceral involvement are less common. Due to the indolent course, SPTL is usually treated with prednisolone or other immunosuppressive agents such as cyclosporine, chlorambucil, methotrexate, cyclophosphamide, interferon-alpha or gemcitabine which provide benefits equivalent to CHOP or CHOP-like regimens.[1,7-9] A recent study showed benefits of bexarotene in treatment of SPTL.[10,11] Radiotherapy or surgery has been used for solitary or localized skin lesions.[1] The spontaneous remission is rare (approximately 3%)[1]. Patients with aggressive disease or failed initial immunosuppressive therapy can be treated with CHOP regimen. Additionally, high-dose chemotherapy followed by stem cell transplantation should also be considered in refractory or recurrence diseases.[1,12,13]

Our patient is a case of SPTL involving skin and lymph nodes, presenting with multiple indurated plaques on the common sites. Up to now, the patient received two cycles of CHOP regimen with resolution of indurated plaques and lymphadenopathy within 3 weeks.

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