# Case 2

Persistent full facial swelling

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**Patient:** A 19-year-old Thai woman from Nakhon Sawan

**Chief complaint:** Facial swelling for 2 months

**Present illness:**

Two months previously, the patient developed intermittent full facial swelling, low grade fever, weight loss from 48 to 42 kilograms and anorexia.

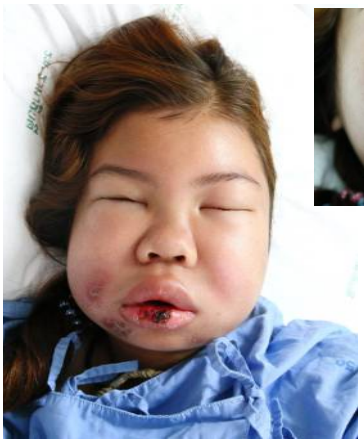
Three weeks later she developed painful oral ulcers and asymptomatic rashes on her face and right forearm. The facial swelling became increasingly prominent and persistent to the level that limited eye opening. She denied having past episodes of facial edema, history of photosensitivity, joint pain, current medical used or family history of facial edema.

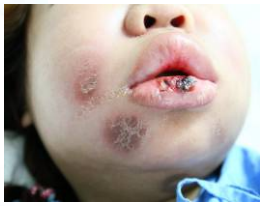
**Past history:** No underlying disease

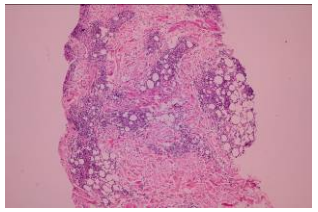
**Physical examination:**

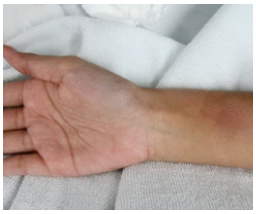
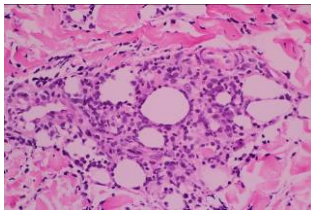
V/S: BT 37.5°C, PR 80 bpm, RR 20 times/min, BP 120/70 mmHg
GA: A Thai woman, good consciousness, mild pallor, no jaundice
HEENT: Diffuse facial edema with marked swelling of eyelids and lips,
mild pale conjunctiva, anicteric sclera, crusted erosions on lower lip
CVS: Normal $S_1S_2$, no murmur
RS: Normal breath sound, no adventitious sound Abdomen:
Soft, non-tender, liver 2 cm below right costal margin,
liver span 14 cm at right mid-clavicular line, spleen not palpable
Lymphatic system: No lymphadenopathy
Neurological system: Grossly intact

**Skin examination:** Few oval-shaped, erythematous to brownish indurated plaques with overlying scales on face and right forearm







**Histopathology: (**S14-1001A, face)

- Hyperkeratosis with follicular plugging of the epidermis and vacuolar alteration of basal cell layer
- Dense inflammatory-cell infiltrate of lymphocytes admixed with histiocytes and plasma cells in fat lobules of subcutaneous tissue
- Lymphocytes show mild to moderate nuclear atypia with hyperchromatid and pleomorphic nuclei
- Lymphophagocytosis also observe in the infiltrate
- Immunostains:
  - Positive for predominately CD3, CD8, Granzyme B, Ki67, Beta F1
  - Scatter positive for CD4
  - Negative for CD20 and Eber

**Investigation:**

Laboratory tests

CBC: Hct 30.8%, Hb 10 g/dL, WBC 2830/$mm^3$ (PMN 65% L 29% M 6%)**,** Platelet 250,000/$mm^3$

BUN: 9 mg/dL, Cr: 0.38 mg/dL

LFT: ALP 161 U/L, GGT 153 U/L, AST 153 U/L, ALT 61 U/L, TB 0.5 mg/dL, DB 0.2 U/L

UA: Normal

Thyroid function test and hepatitis profile: Normal

ANA: Negative**,** Anti-DNA: Negative**,** ANA 12 profile: Negative

Anti Cardiolipin IgG, Beta2 Glycoprotein1 IgG: Negative

p-ANCA and c-ANCA: Negative

CRP: 7.09 mg/L [<5], ESR: 31 mm/hr

CH50, C3c, C4: Normal levels

Imaging study

CT scan of the neck, chest and abdomen:

- Diffuse infiltrative enhancing lesions involving skin, subcutaneous and strap muscle of the neck extending to facial, orbits and thorax
- Hepatosplenomegaly
- No mediastinal and intra-abdominal lymphadenopathy

**Diagnosis: Subcutaneous panniculitis-like T-cell lymphoma**

**Treatment:**

- Systemic chemotherapy
  (one cycle of CHOP regimen then 6 cycles of CHOEP regimen)

**Discussion:**

This patient presented with diffuse and persistent facial swelling, indurated plaques, hepatomegaly, fever and night sweat. Initial presenting

symptoms with facial swelling gave high suspicion for conditions that mimic both acquired or non- acquired angioedema. Given that our patient also had cutaneous involvement of multiple plaques on the face and right forearm, this led us to suspect lymphoproliferative diseases and lupus erythematosus. Moreover, with the history of intermittent fever and night sweat along with hepatomegaly, lymphoproliferative disease was the provisional diagnosis. The skin biopsy performed on indurated plaques of face and forearm showed lobular infiltration of lymphocytes, plasma cells, bean bag cells and nuclear dust in the subcutaneous fat. The immunophenotypic features show positive CD3, CD8, beta F1 and negative CD20, CD56 and EBER. Therefore, the finally diagnosis in our patient is subcutaneous panniculitis-like T-cell lymphoma**.**

Subcutaneous panniculitis-like T-cell lymphoma (SPTL) is a rare T-cell lymphoma that preferentially affects the subcutaneous tissue and may clinically mimic panniculitis. It accounts for less than 1% of non-Hodgkin's lymphomas.[1] It typically presents in young adults with has no clear gender predisposition. Clinical presentation usually consists of painless multiple indurated, poorly circumscribed subcutaneous nodules or plaques ranging from 0.5 to more than 10 cm in diameter on the trunk and extremities. Isolated reports describe plaques on the face and chest.[1,2] The facial swelling observed our patient is an unusual presentation that has been described in one case report in the literature.[3] Common skin findings of SPTL include various stages of healing and areas of lipoatrophy which result from the spontaneous regression of subcutaneous nodules without treatment.[1,4] Up to 60% of patients presented with prodromal symptoms such as fevers, chills and night sweat.[5] Lymphadenopathy and hepatosplenomegaly are uncommon, it accounts for 8%.[1,4] Up to 50% of patients had laboratory abnormalities, most commonly include anemia, leukopenia, thrombocytopenia or combined cytopenias, and elevated liver function tests.[1] SPTL has been associated with autoimmune disorders, including systemic lupus erythematosus, juvenile rheumatoid arthritis, type 1 diabetes mellitus, Sjogren's syndrome, and idiopathic thrombocytopenic purpura. The relationship between SPTL and autoimmune disease is unclear.[1,2,6]

Histopathologic findings are dense lymphoid infiltrates of small- to medium- to large-sized lymphocytes present preferentially in the subcutaneous tissue, predominantly in a lobular pattern. The neoplastic infiltrate is composed of pleomorphic T cells of variable sizes with irregular and often hyperchromatic nuclei. Rimming of individual adipocytes by neoplastic T cells is a common feature. The findings of atypical lymphocytic lobular panniculitis have been found to overlap features of LEP and SPTL.[1,7] Histopathologic criteria favoring the diagnosis of LEP include epidermal involvement, mucin depositions, the presence of reactive germinal centers, clusters of B cells or considerable number of admixed plasma cells, and polyclonal TCRγ. These findings suggest that all patients with suspected SPTL should be screened for LE.[2] The immunophenotype is performed as a diagnostic method. World Health Organization-European Organization for Research and Treatment of Cancer (WHO-EORTC) classification of primary cutaneous lymphomas in 2008 redefined SPTL as a purely TCR α/β phenotype, which is typically positive for βF1, CD3, CD8, and cytotoxic proteins and negative CD4, CD5, CD56, and EBER that have somewhat better outcome.[4,7]

SPTL has generally been associated with a favorable prognosis and a protracted disease course with a 5-year survival rate of more than 80% and low risks for developing hemophagocytic syndrome (approximately 17%), which is

associated with poor outcomes.[4,8] Metastatic disease and visceral involvement are less common. Due to the indolent course of disease, SPTL is usually treated with prednisolone or other immunosuppressive agents such as methotrexate and cyclosporine, which provide benefits equivalent to CHOP or CHOP-like regimens.[1 9-13] A recent study showed benefits of bexarotene use in treatment of SPTL.[14,15] Patients with aggressive SPTL, those who failed initial immunosuppressive therapy, or those with recurrent disease can be treated with CHOP regimen. Additionally, high-dose chemotherapy followed by stem cell transplantation should also be considered in these refractory cases.[1,10]

Our patient received CHOP regimen due to visceral organs involvement with resolution to lipoatrophy and brownish patches in 3 months.

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