An atypical presentation of subcutaneous panniculitis-like T-Cell lymphoma

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ABSTRACT:

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Subcutaneous panniculitis-like T-Cell Lymphoma (SPTL) is a rare cytotoxic T-cell lymphoma primarily localized to the subcutaneous tissue. Clinical presentations are usually consisted of painless indurated subcutaneous nodules or plaques with variable number and sizes. The most common sites of presentation are extremity and trunk. We reported a case of SPTL patient who presented with progressive painless facial and periorbital swelling with B symptoms.

Key words: Facial swelling, Subcutaneous Panniculitis-like T-Cell Lymphoma (SPTL)

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Introduction

Subcutaneous panniculitis-like T-Cell lymphoma (SPTL) is a rare cytotoxic T-cell lymphoma primarily localized to the subcutaneous tissue[1]. Cutaneous manifestations are consisted of painless indurated subcutaneous nodules or plaques on extremity and trunk[2]. In this present study, we reported a case of SPTL with facial swelling which it was atypical presentation.

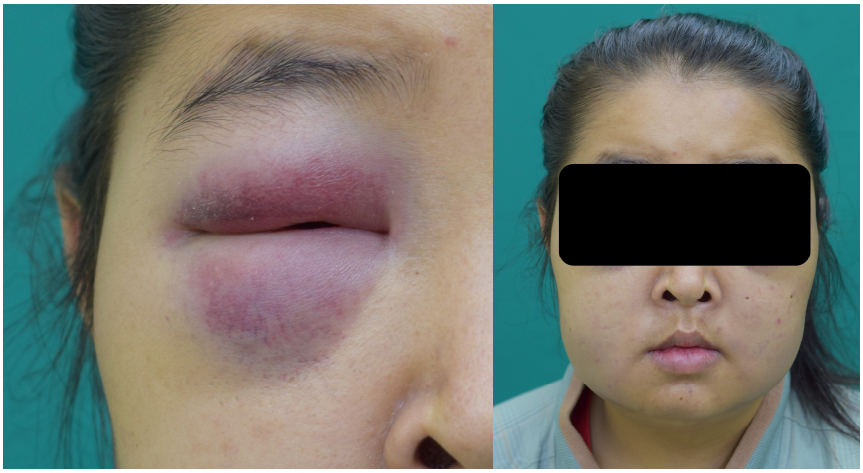

Figure 1,2 periorbital edema and erythema, multiple ill-defined border, discrete, not tender subcutaneous nodules on both cheeks

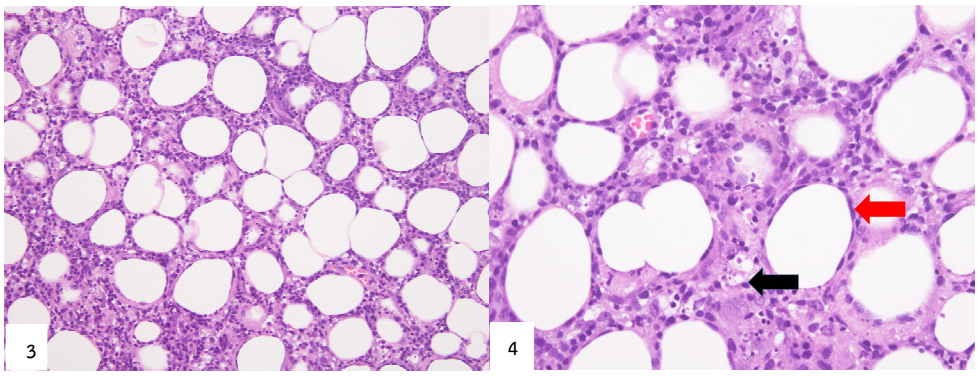

Figure 3,4 Skin biopsy showed lobular panniculitis with atypical mononuclear cell, large hyperchromatic nuclei with rimming individual adipocytes (red arrow) and phagocytic histiocytes (black arrow)**.** (Hematoxylin-eosin, original magnification: **3** X100, **4** X400)

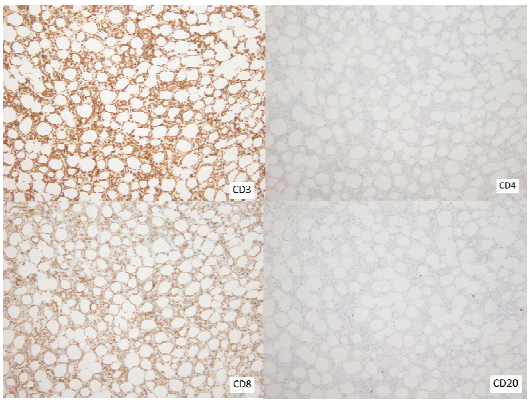

Figure 5 The immunohistochemistry showed positive for CD3, CD8 and negative for CD4, CD20 (original magnification: X100)

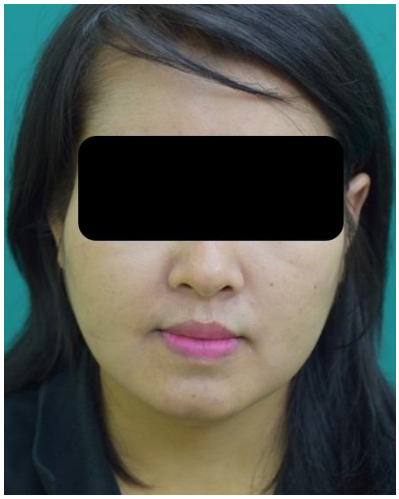

Figure 6 Clinical improvement after treatment

Our case

A 24-year-old Thai female with prolonged fever and weight lost of 8 kg during 3 months was studied. One month ago, the patient had developed progressive and persistent painless facial and periorbital swelling. She denied any history of trauma to the affected area. The patient had no history of underlying disease and no family history of similar skin diseases. The dermatologic examination revealed periorbital edema and erythema, multiple ill-defined border, discrete, not tender subcutaneous nodules on both cheeks (Figure 1, 2). In addition, the physical examination revealed hepatomegaly as well. The lymphadenopathy or neck vein engorgement was not found. The other physical examinations were unremarkable. Furthermore, laboratory investigations showed mild anemia (hematocrit 32.3%) and transaminitis (aspartate aminotransferase 403 U/L, alanine aminotransferase 186 U/L, alkaline phosphatase 459 U/L). Skin biopsy taken from the right cheek of patient revealed the lobular panniculitis with atypical mononuclear cells and large hyperchromatic nuclei with rimming individual adipocytes in a lace-like pattern (Figure 3, 4). The immunohistochemistry showed the positive results for CD3, CD8, granzyme B and T-cell intracellular antigen-1 (TIA-1) (Figure 5). Computed tomography of chest and abdomen indicated diffuse nodular and ill-defined soft tissue density lesions with borderline enhancement involving with subcutaneous fatty tissue of chest, abdominal wall, upper thighs and arms. Bone marrow biopsy revealed no evidence of the bone

marrow involvement. Therefore, the clinical and pathological findings from above examinations were compatible with SPTL. Our patient was received an initial treatment with prednisolone and cyclosporine which patient subsequently achieved response after 1 month later (Figure 6).

Discussion

SPTL is a rare primary cutaneous lymphoma of mature cytotoxic T cells. This variant is usually found in 1% of non-Hodgkin's lymphomas[1]. The updated classifications from 2016 revision of the World Health Organization classification of lymphoid neoplasms describe the term of SPTL as it is only used for cases with T-cell receptor (TCR) α/β phenotype, while cases expressing TCR γ/δ phenotype are reclassified as primary cutaneous gamma/delta T-cell lymphoma[2]. In addition, SPTL is generally appeared in young to middle-aged patients. Studies have established that women have a higher prevalence of SPTL than men[3]. The clinical manifestation are usually consisted of multiple painless indurated nodules and infiltrated plaques with a variable diameter[1,3]. Furthermore, ulcerative lesions and lipoatrophy are also observed in some cases[3,4]. The sites of skin involvement are typically presented on lower extremities and trunk. Face is the less common areas of skin involvement[3]. The patients may present with constitutional symptoms such as fever, and night sweats[3,5]. Lymphadenopathy and hepatosplenomegaly are considered as uncommon[1,2,3]. The associated autoimmune diseases are also found in approximately 25% of cases such as lupus erythematosus, hypothyroidism and multiple sclerosis[3].

The laboratory abnormalities including anemia, leukopenia, thrombocytopenia, abnormal liver function, elevated erythrocyte sedimentation rate, elevated lactate dehydrogenase and elevated beta-2-microglobulin level are found as well[1,3]. The histopathologic findings are consisted of lobular panniculitis with dense infiltration of atypical lymphocytes. Moreover, a typical finding is the rimming of individual adipocytes by atypical lymphocytes[1,3,4]. Macrophages filled with nuclear debris in the cytoplasm (bean-bag appearance) is a common finding. Additionally, other immunohistochemical studies demonstrate that these atypical cells are positive for CD3, CD8, Beta F1, cytotoxic proteins such as TIA-1, perforin and granzyme B whereas negative for CD4, CD30, CD56, and EBER[3,6].

SPTL generally has a favorable prognosis. The five-year survival rate is about 85 % in patients without hemaphogocytic syndrome[1,6].

The treatment of SPTL include immunosuppressive drugs, biologic agents, multidrug chemotherapy, radiotherapy, and/ or

bone marrow transplantation[7]. The spontaneous remission is rarely found[1,3]. Solitary or localized skin lesions could be treated with radiotherapy[1,6]. Typically, SPTL is treated with prednisolone with or without other immunosuppressive agents such as cyclosporine, methotrexate, cyclophosphamide and chlorambucil[7,8]. The aggressive disease or failure of initial therapy should be considered about chemotherapy such as CHOP regimen or cisplatin-based, melphalan-based, ifosfamide-based, and purine nucleoside analogue–based regimens[7]. Additionally, stem cell transplantation performed with high dose chemotherapy is a treatment option for recurrent or refractory diseases[1,7]. Moreover, combination of high-dose chemotherapy and autologous peripheral blood stem cell transplantation are successful treatment for SPTL with hemophagocytosis[9].

In the review of 7 patients in the age-group 2-30 years who presented with facial swelling, clinical courses of disease were similar to the other site involvement. The patients showed good clinical response after received the treatment with prednisolone and other immunosuppressive agents[10,11,12,13,14,15,16].

Finally, the result of the present study indicated that after 1 month of initial treatment with prednisolone and cyclosporine, the clinical improvement was significantly observed in patient. Thus, we had consulted hematologist for the further management in our analysis.

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