Subcutaneous Panniculitis-like T-Cell Lymphoma with Co-Existing Lupus Panniculitis: A Case Report

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Abstract

Subcutaneous panniculitis-like T-cell lymphoma (SPTL) is an extraordinary form of peripheral T-cell lymphoma, accounting for much less than 1% of all non-Hodgkin lymphoma. It usually happens amongst women with a median age of onset of 36 years. It can arise with autoimmune disease ~20%. SPTL may also coexist with different autoimmune diseases (e.g., lupus erythematosus [LE]) despite normal workup.

A 44-year-old woman presented with a small, 2x 2 cm mass on her right inner thigh; that was erythematous, non-movable, and tender. She has had repeated admissions due to soft tissue infection, and persistent pleural effusions. She experienced eruption of numerous, nonpruritic, erythematous maculopapular rashes on her body. Initial laboratory workup showed persistent cytopenia (anemia and leukopenia), >10x elevated ferritin and LDH, and; elevated transaminases, and procalcitonin. Other work-ups such as bone marrow studies, pleural fluid flow cytometry (comprehensive lymphoma panel), CT-PET, and connective tissue disease (LE and antiphospholipid syndrome panels) were unremarkable. SPTL with LE profundus overlap was diagnosed upon review of the nodule biopsy report. The patient was treated with prednisone and cyclosporine with a good response, as seen on follow-up.

SPTL can arise in patients with LE or vice versa. The illness itself may also present initially as benign panniculitis, eczema, dermatitis, psoriasis, cellulitis, or other skin and soft tissue infections. The culprit behind this disease is a HAVCR2 (Hepatitis A Virus Cellular Receptor 2), mutation which occurs in 60% of SPTL cases and is more typically seen in Asian and Eastern patients. Its presence is related to HLH-refractory and severe disease course. For patients with indolent course, the suggested first-line remedies encompass immunosuppressive agents together with prednisone, cyclophosphamide, or methotrexate; extreme conditions with aggressive infiltrative lymphocytes may also require a combination of chemotherapy regimens. In patients resistant to chemotherapy, cyclosporine A alone or combined with steroids causes desirable, including in HPS patients.

SPTL is a diagnostic challenge. A high index of suspicion, and correlation of clinical, imaging, and histopathologic reports will increase the probability of detecting this disorder, consequently lessening the probability of the delay in treatment. Indolent courses have desirable overall survival, except for patients presenting with HLH/HPS upon diagnosis.

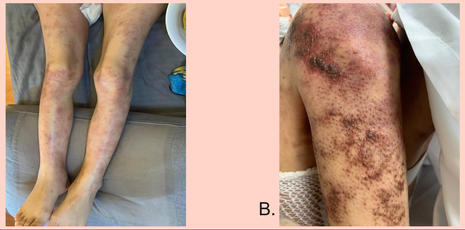
Keywords: SPTL, Lupus Panniculitis, Peripheral T-Cell Lymphoma

Objective

To present a diagnostic dilemma in a patient with skin manifestations, anemia and leukopenia, B symptoms, and unremarkable imaging and bone marrow studies.

Case

A 44-year-old woman presenting with multiple rashes associated with weight loss, fever, malaise. She had repeated admissions due to soft-tissue infection, and persistent pleural effusion. Lab works show anemia and leukopenia, >10x elevated ferritin and LDH; elevated transaminases, and procalcitonin. Her biopsy of the lesion was inconclusive, with unremarkable imaging and bone marrow studies.



Lesions noted in a 44-year-old patient. (A) Lesions during the beginning of the disease. (B) Lesions began to be darker in color and more erythematous as the disease progressed.

Discussion

Subcutaneous panniculitis-like T-cell lymphoma is a rare form of peripheral T-cell lymphoma, accounting for less than 1% of all non-Hodgkins lymphoma.

SPTL has a female predominance, occurring in both children and young adults. The average onset is 36 years. It can occur with autoimmune disease ~20%. SPTL can occur in patients with lupus erythematosus or vice versa. The overlapping features are called atypical lymphocytic lobular panniculitis.

The culprit behind the disease is a mutation in HAVCR2 (Hepatitis A Virus Cellular Receptor 2), which is present in 60% of SPTL cases. It abrogates TIM-3 (T-cell immunoglobulin domain and mucin domain 3) membrane expression leading to persistent immune activation and cytokine production.

Like other cases of SPTL, she presented as a diagnostic dilemma wherein bone marrow studies, autoimmune work-up, and imaging provided no clue as to the patient's diagnosis. Moreover, immunoglobulins were normal which led us to believe that B-cell is not the primary cause of the inflammation and stimulation of the cytokines.

Due to its histological characteristics, SPTL is frequently misdiagnosed as lupus panniculitis or a few different peripheral T-cell lymphoma (i.e., primary cutaneous gamma/delta T-cell lymphoma, extranodal NK T cell lymphoma).

For patients with a fairly indolent condition, the recommended first-line treatments consist of immunosuppressive agents consisting of prednisone, cyclophosphamide, or methotrexate; extreme conditions with aggressive infiltrative lymphocytes may also require a combination of chemotherapy regimens. Patients with chemotherapy resistance, cyclosporine A alone or combined with steroids result in good outcomes including patients with HPS . For cyclosporine, 15mg/kg/day is started with titration up to 200mg/day, while other studies start at 400mg/day with similar outcomes.

The overall five-year survival rate for SPTL is 90%, however, the rate falls to less than 50% if a patient presents with HPS.

Conclusion

Like other diseases, subcutaneous panniculitis-like T-cell lymphoma is also a diagnostic challenge. It can also occur in patients with an autoimmune disorder such as LEP. A high index of suspicion, and correlation of clinical, imaging, and histopathologic reports increases the likelihood of detecting such disease and therefore lessens the chances of delay in treatment. Indolent courses have good overall survival except for patients presenting with HLH/HPS upon diagnosis.

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