Atypical Lymphocytic Lobular Panniculitis: A Distinct Entity in the Spectrum of Cutaneous Lymphoid Dyscrasia

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Atypical Lymphocytic Lobular Panniculitis: a distinct entity in the spectrum of cutaneous lymphoid dyscrasia

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Abstract:
We report a case of atypical lymphocytic lobular panniculitis (ALLP) characterized by recurrent subcutaneous nodules with similar molecular and phenotypic profile of cutaneous T-cell lymphoma. A twenty-one year old female presented with a six-year history of recurrent erythema nodosum on the lower extremities. The histopathology revealed a lobular panniculitis composed of histiocytes and atypical CD3 T cells. The patient was treated with oral dapsone with resolution of the rash. However, the lesions recurred and most recent episode involved buttocks with subcutaneous panniculitis-like T-cell lymphoma and potential for malignant transformation, recognizing ALLP as a distinct entity is important in understanding the clinical and management spectrum of lymphocytic lobular panniculitis.

Case Presentation:
HPF: one year old female with a six-year history of recurrent “erythema nodosum”. Reports painful and pruritic lesions that usually appear spontaneously within one month and recur about one to two episodes per year. No prodromal symptoms.
ROS: Mild intermittent shortness of breath and joint pain
PMHx/Family Hx: Non-contributory
PE: Mobile, alert female without acute distress. Multiple indurated, erythematous, subcutaneous, tender nodules and plaques located on the forearms and lower extremities. No lower extremity edema.

Discussion:
ALLP as described by Magro et al. represents a spectrum of disorders with recurrent subcutaneous nodules without chronic and atypical lymphocytic infiltration. Other disorders in this spectrum include alopoeia mucinosa, pityriasis lichenoides chronicus, large plaque parapsoriasis, idiopathic follicular erythrodema, mucinosis, pigmented purpuric dermatoses, and atypical hypogamaglobulinemia. Some authors suggest that ALLP is a concomitant presentation or overlap of STCL and LEP. Wilenre et al. reported that 19% of STCL patients had an associated autoimmune disorder with lupus erythematosus LE being the most common. There are reported cases where diagnosis of earlier LEP has been changed to STCL. Pincus et al observed one STCL patient with a positive DIF test without histopathologic feature of LE. Monoclonal pattern has been described in 70 to 75% of ALLP patients. Therefore, the use of TCR gene rearrangement study in distinguishing the ALLP from malignant process should be made with caution. Initial screening should include hematologic work up including complete blood count, rheumatologic work up of cutaneous and extracutaneous manifestations, and erythrocyte sedimentation rate. Similarly, LEP patients should also be followed closely due to its close association with STCL. Perhaps ALLP may represent a continuum of inflammatory disarray from chronic antigenic stimulation. This spectrum then can prime the initial benign process to possible clonal selection and leading to malignant transformation (figure 3).

TREATMENT:
• Goal of treatment is to halt the progression to lymphoma
• Corticosteroids and nonsteroidal anti-inflammatories for pain
• Oral retinoid therapy for persistent lesions
• Case series by Magro et al. reported treatments with dapsone, hydroxychloroquine, interferon, and anti-CD25 monoclonal antibody alemtuzumab, which is currently approved for the treatment of chronic lymphocytic leukemia

PROGNOSIS:
• Exact prognosis remains unknown
• Reported cases of ALLP follow an indolent course and no reported cases of ALLP progressing to STCL
• However, few cases of STCL where history of waxing and waning patterns have been observed
• Persistent identical T-cell clone at different biopsy sites over time has been demonstrated in ALLP
• Therefore, ALLP may represent a preclinical stage of STCL and close monitoring and follow up is warranted

Differential Diagnosis:
• Subcutaneous panniculitis-like T-cell lymphoma (STCL)
• Lupus erythematous panniculitis (LEP)

Table 1. Comparison of clinical features of ALLP, STCL, and LEP

<table>
<thead>
<tr>
<th>Feature</th>
<th>ALLP</th>
<th>STCL</th>
<th>LEP</th>
</tr>
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<tbody>
<tr>
<td>Clinical</td>
<td>Indurated, erythematous, subcutaneous, tender nodules and plaques</td>
<td>Indurated, erythematous, subcutaneous, tender nodules and plaques</td>
<td>Indurated, erythematous, subcutaneous, tender nodules and plaques</td>
</tr>
<tr>
<td>Histopathologic</td>
<td>Lobular panniculitis composed of histiocytes and atypical CD3 T cells</td>
<td>Subcutaneous panniculitis-like T-cell lymphoma</td>
<td>Lupus erythematosus panniculitis</td>
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References:

Figure 1. Proposed schematic representation of Atypical lymphocytic lobular panniculitis: A clonal subcutaneous T-cell dyscrasia (figure 1).