

SKINmages

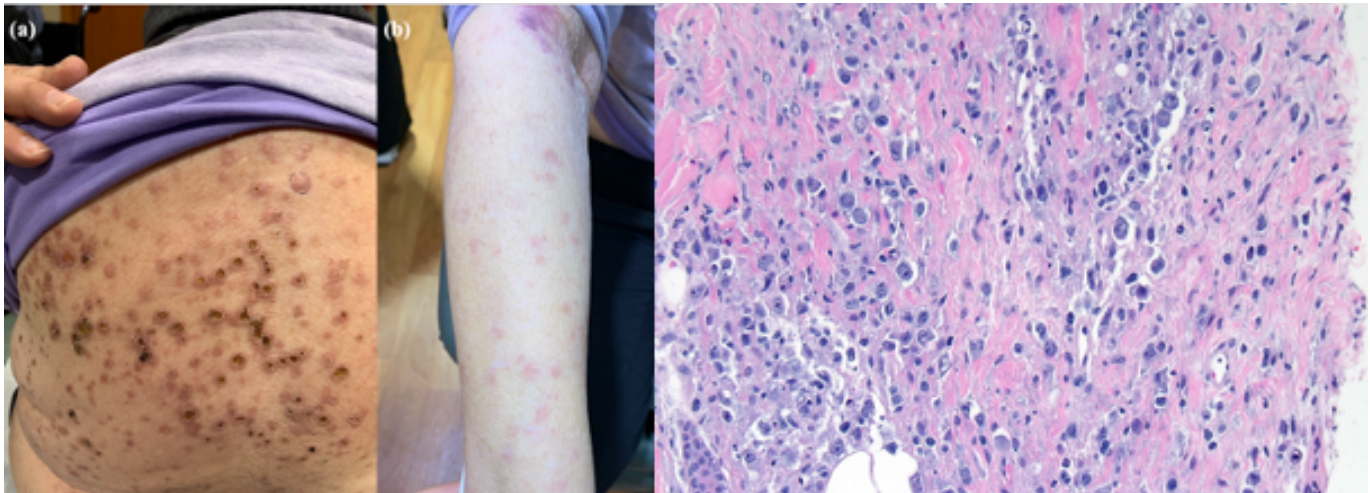
A Case of Systemic Anaplastic Large Cell Lymphoma with Secondary Cutaneous Involvement

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A 75-year-old female presented to the dermatology clinic with non-healing, occasionally pruritic bumps on her back and arms for the past three to four weeks. Physical examination revealed multiple crops of papules in different stages of healing with central areas of necrosis and ulcerations (Figure 1a). Bilateral forearms showed scattered, erythematous papules (Figure 1b). Cervical lymphadenopathy was also noted. The patient endorsed fatigue, but review of systems was otherwise negative.

The patient was followed concurrently by an oncologist due to the severe lymphadenopathy. Findings from the

oncologist included a CT of the head and neck, which showed high volume disease with bilateral axillary and mediastinal adenopathy. A bone marrow biopsy demonstrated cells staining positive for T-cell origin and negative for ALK.

A punch biopsy revealed large lymphocytes scattered throughout the dermis with a few eosinophils. Under higher magnification, atypical lymphocytes with horseshoe-shaped nuclei were seen (Figure 2). Immunohistochemical staining was positive for CD3 and CD30. In situ hybridization for Kappa and Lambda were negative. These findings were consistent with systemic

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anaplastic large cell lymphoma with secondary dissemination to the skin. The patient's oncologist was consulted and agreed with the final diagnosis. She was started on a chemotherapy regimen, but unfortunately, was lost to follow-up.

Systemic anaplastic large cell lymphoma (sALCL) is a rare T-cell lymphoma, representing 2% of non-Hodgkin lymphomas in adults. By definition, CD30 is expressed in at least 75% of T-lymphocytes. The appearance of neoplastic cells with horseshoe or kidney-shaped nuclei called "hallmark cells" under microscopic examination is pathognomonic.¹ The disease can be subclassified based upon the presence or absence of anaplastic lymphoma kinase (ALK) protein.² Both groups commonly present with signs of systemic disease, including fever, night sweats, and weight loss.³

Secondary dissemination to the skin in sALCL occurs in approximately 20% of cases and is often observed in advanced stages of the disease.³ Most reports describe limited spread to one area of the body, such as the groin.⁴ Involvement of multiple regions, as in our case, is seldom reported. The infrequency of these cutaneous manifestations can lead to misdiagnosis with other CD30+ lymphoproliferative disorders. These diseases differ from sALCL in terms of prognosis and management and it is important for the clinician to discriminate between these entities. For example, lymphomatoid papulosis presents as erythematous to brown papules or nodules in different stages of healing with a distinct waxing and waning course. Primary cutaneous anaplastic large cell lymphoma (pcALCL) is another disease with a more similar presentation to sALCL. Both diseases can manifest as solitary or localized, firm, red to brown non-healing ulcerative lesions.

However, pcALCL is not associated with B symptoms.⁵

In our case, the final diagnosis was made by linking our gross findings with the patient's reported symptoms and the results of histology. To arrive at the correct diagnosis and maximize the chance of remission, it is critical to consider all aspects of a case, from imaging to histopathologic findings to data from other medical specialties, in all patients.

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